Comprehensive review of typical and atypical pathology of the appendix on CT: cases with clinical implications

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ABSTRACT

Acute appendicitis is the most common abdominal surgical emergency in the United States with approximately 250,000 cases annually. Computed Tomography (CT) has emerged as the most accurate diagnostic test to triage these patients for emergent surgery. Although the radiology search pattern is prioritized to detect an inflamed appendix, not all appearances equate to a typical surgical appendicitis. There are a select set of atypical pathologies involving the appendix that have subtle differences on CT, but can have catastrophic complications if treated with emergent appendectomy. This paper will review the spectrum of CT appearances and clinical management for typical and atypical appendiceal pathologies.

1. Introduction

Acute Appendicitis is the most common abdominal surgical emergency in the United States (US) with approximately 250,000 cases annually since 1990 [1,2]. Advanced multi-detector Computed Tomography of the Abdomen and Pelvis (CTAP) has emerged as the most accurate diagnostic imaging test. Increased use of CTAP to evaluate right lower quadrant (RLQ) pain has reduced the negative laparoscopy rate (NLR) from 20 to 30% to as low as 1.7% in select institutions [2–6]. However, not all abnormal appendices seen on CT require emergent surgical resection [7,8]. Atypical appendiceal pathologies ranging from neoplasms to inflammatory conditions can mimic and even cause a superimposed acute appendicitis; making them difficult to differentiate from typical inflammation. While not always possible, pre-operative recognition of these entities can decrease complications from inadequate or unindicated emergent resections [8,9]. This paper reviews the spectrum of imaging and management features that differentiate typical acute appendicitis from atypical appendiceal pathologies.

2. CT use and technique

Non-focused CTAP is the gold standard and most cost-effective diagnostic test for appendicitis in non-pregnant adults with RLQ pain [3,10,11]. In 2011 the ACR gave it the highest diagnostic rating for this population, which is most frequently afflicted by acute appendicitis [3,11–14]. Current data shows the sensitivity (Sn) and specificity (Sp)

of CTAP for acute appendicitis ranges from 85 to 100% depending on the type of contrast enhancement [3,15]. Non-contrast CTAP in patients with adequate mesenteric fat has a Sn of 96% and Sp of 99% [16]. However, intravenous (IV) contrast significantly improves accuracy even with minimal fat; showing Sn of 100% and Sp of 97% due to mucosal and inflammatory tissue enhancement [16,17]. The use of positive oral or rectal contrast can be helpful to delineate bowel and opacify the normal appendix [17]. However, these contrast agents increase radiation, cost, scan time and risk of complications, particularly perforation [3,11,17]. In 2009 Anderson et al. showed no difference in diagnostic accuracy between IV only and IV with oral or rectal contrast for the diagnosis of appendicitis on CTAP [17]. Additionally, the ACR only recommends the routine use of IV contrast and leaves the addition of oral or rectal contrast to institutional preference [3]. Regardless of the contrast used all exams should cover from the diaphragm to the pubic symphysis with 5 mm thick slices reformatted in sagittal and coronal planes to better detect complications of appendicitis and alternate pathologies [3,18–20]. Recent studies also support the use of advanced low dose CT techniques for suspected appendicitis with randomized multi-center data showing non-inferiority in young adults [21–23]. Contrary to non-pregnant adults, Ultrasound (US) is recommended as the initial evaluation for children less than 14 years of age and pregnant women. A brief focused US in children is almost as accurate as CTAP due to their small body habitus allowing good visibility and compression of the appendix. This limits CT radiation exposure to only those children with equivocal US findings. US in

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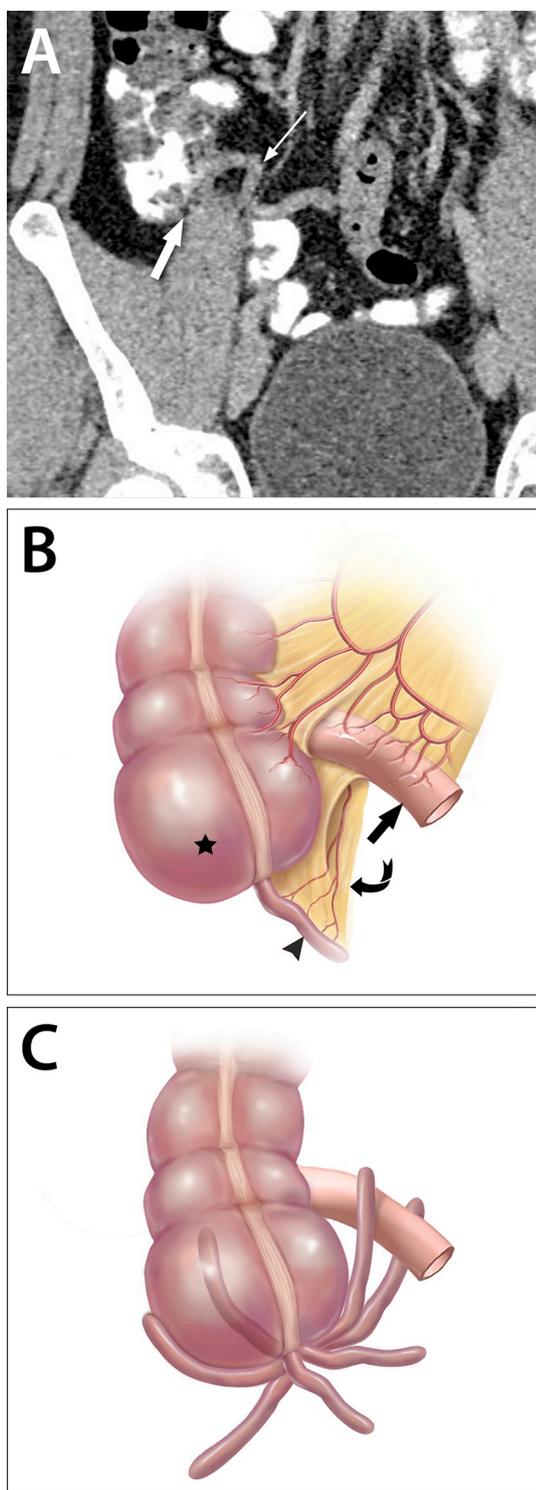 *E-mail address:* rehan.quadri@phhs.org (R. Quadri).

Fig. 1. Normal appendix.
Non enhanced coronal CT image (a) reveals a normal appendix (thin arrow) arising from the inferior tip of the cecum (thick arrow). Schematic of the normal ileocecal region (b) shows the normal appendix (black arrowhead), mesoappendix (black curved arrow), cecum (black asterisk) and terminal ileum (black arrow). (c) Schematic showing the different positions of the distal end of the appendix which always arises from the cecal tip.

pregnant women is very insensitive by itself, however when combined with focused magnetic resonance imaging the accuracy is similar to CTAP without any risk of radiation to the fetus [3,12].

3. Normal appendix

The appendix is a true diverticulum of the cecum arising 3 cm below the ileocecal valve [20]. On CT it appears as a worm-like blind-ending air-filled loop of bowel with a length around 10 cm (Fig. 1). The walls are paper thin around 3 mm and are continuous with the cecal layers (mucosa, submucosa, muscularis and serosa), but have more submucosal lymph tissue [24]. The base of the appendix maintains a constant relationship with the cecum usually arising at the apex; however the body and distal tip are mobile laying in the retrocecal, subcecal, retroileal, preileal or pelvic regions based on the natural positioning of the cecal taeniae coli. Arterial supply and venolymphatic drainage runs through the triangular mesoappendix and is an important site of inflammation. Although no single intraluminal content is characteristic of a normal appendix, a non-dilated air-filled appendix is generally thought to be normal [20,24,25]. The average diameter of the appendix on CTAP from outer wall to outer wall ranges from 3 to 10 mm with most normal appendices measuring greater than 6 mm. Dedicated prospective CT studies have found the average to be around 8 mm [24–23]. Older literature reports 6 mm or less as a normal diameter, which is adapted from ultrasound [24,27,28]. However, on CTAP without associated inflammation larger sized appendices measuring between 6 and 10 mm are normal [15,24,26]. This is likely due to the lack of compression on CT, making the interpretation of luminal distension heavily dependent on concomitant inflammation rather than quantitative measure [24,26,27]. On CTAP the appendix is unseen more often in women than men with data showing it occurs about 24% of the time in women and only 13% of the time in men [24]. Fortunately, only 2% of these cases go on to develop an acute appendicitis, which also occurs more frequently in women. This is thought to be from women having lower amounts of fat resulting in less obvious inflammation. In general, however, a non-visualized appendix without inflammatory changes in the RLQ often excludes the diagnosis of acute appendicitis [29,30].

4. Typical acute appendicitis with potential complications

Typical acute appendicitis usually occurs in the 2nd decade of life with a second peak of incidence in elderly adults as well [1–3]. It is caused by obstruction of the appendiceal lumen by either appendicoliths from inspississated stool and salts or lymphoid hyperplasia from post-viral inflammation, which occurs more often in children. All other causes of obstruction are considered atypical. Patients present clinically and radiographically along a pathophysiologic spectrum that ranges from mild inflammation to perforation depending on the length of obstruction (Table 1) [20]. Various clinical assessment tools, such as the Alvarado score, exist to guide management and the use of imaging [31,32]. It is important to note that rapid triage and imaging in modern acute care facilities results in patients presenting earlier with milder inflammation. More severe presentations of prolonged obstruction are now less frequent and limited to the elderly and small children due to a lack of self-awareness [11,32].

4.1. Early and late presentation

Initial obstruction leads to accumulation of normal secretions that distend the appendix causing vague visceral periumbilical and

Table 1
Evolution of typical appendicitis.

|	**Early**	**Late**	**Delayed w/ Complications**
Pathology	Non-malignant luminal obstruction by appendicolith or lymphoid Hyperplasia (viral in children) causing distension	Increasing luminal distension impairs arterial perfusion & venous outflow causing wall ischemia, mucosal disruption and bacterial invasion	Prolonged high luminal pressure compromises perfusion causing necrosis from infarction leading to gangrene & perforation
Clinical Presentation	Vague periumbilical and epigastric visceral pain	Fever, elevated WBC, Anorexia, nausea, vomiting, RLQ pain with rebound & voluntary guarding	Severe sepsis, peritonitis with rigidity, ileus and hemodynamic instability
CT Findings	Luminal distension Mucosal enhancement Wall thickening Periappendiceal fat stranding	Early Appendicitis findings w/ **RLQ inflammation:** cecal apex inflammation (arrow sign), ileitis, lateral conalfascia thickening and mild free fluid **Severe luminal distension:** >15mm with appendicolith suggests impending rupture	**Late and Early findings w/ Gangrene:** thick shaggy non-enhancing wall +/-pneumatosis **Contained Perforation:** phlegmon, abscess, wall defects, moderate free fluid, fistula **Free perforation:** free air, large free fluid, multifocal abscesses, free appendicolith, Ileus, **peritonitis,** thrombophlebitis (portal vein), hepatic abscess
Treatment	Emergent Laparoscopic Appendectomy	Emergent Laparoscopic Appendectomy	Emergent appendectomy if no phlegmon or abscess If otherwise, antibiotics +/- percutaneous drainage (abscess > 4cm) and delayed surgery
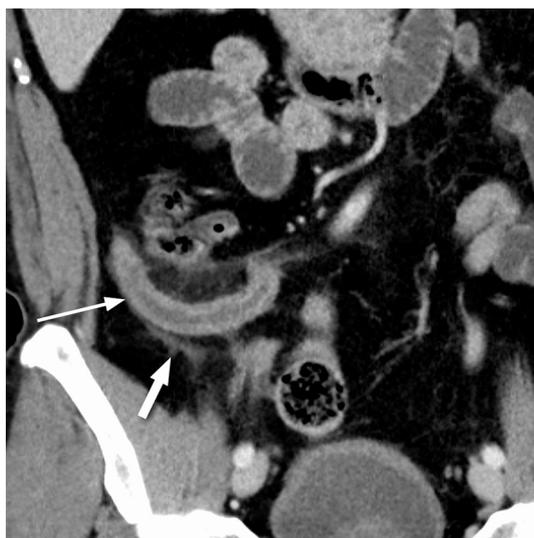
Fig. 2. Uncomplicated acute appendicitis.
53 yr old male with 16 h of initial periumbilical pain that then localized to the RLQ associated with tenderness on exam and elevated WBC. Coronal contrast enhanced CT image shows a dilated fluid filled appendix measuring 12 mm in diameter (thin arrow) with thickened homogeneously hyper enhancing wall. There is peri appendiceal fat strandying and thickening of the lateral conal fascia (thick arrow). There was no fluid collection or extra luminal air. At surgery an inflamed and unruptured appendix was identified.

epigastric pain. This progressively localizes to the RLQ with increasing inflammation [15,19,31]. On CTAP these patients show early findings of mild luminal distension to 6–10 mm in diameter (Sn 93%, Sp 92%), periappendiceal fat stranding (Sn 87% and Sp 74%), mucosal hyper-enhancement (Sn 75% and Sp 85%), edematous wall thickening greater than 3 mm (Sn 66% and Sp 96%) and rarely radiopaque appendicoliths (Fig. 2) [15,20,33–36]. Although luminal contents are nonspecific, an

entirely air filled appendix is usually normal whereas fluid content is more often seen with inflammation [15,25]. Eventually, continued obstruction without intervention results in worsening inflammatory changes that spread into the right lower quadrant. These late findings on CTAP include prominent luminal distension greater than 10 mm, increased wall thickening, thickening of the lateral conal fascia, right lower quadrant mesenteric fat stranding, reactive ileitis and a small amount of free fluid [34–38]. Inflammation also extends to the cecal base, which can cause oral contrast to pool in the shape of an "arrowhead" pointing towards the appendiceal orifice (Fig. 3) [39].

4.2. Delayed presentation with complications

Prolonged obstruction results in high intraluminal pressures that block blood flow causing ischemic infarction, gangrenous necrosis and eventually perforation [15,20]. Initial CTAP findings concerning for infarction and gangrene include a shaggy appendiceal wall with patchy areas of non-enhancement, discontinuity and pneumatosis (Fig. 4). Once luminal distension reaches 15 mm the appendix is at increased risk for perforation, which can be contained or uncontained [15,38]. On CTAP a contained perforation can result in a phlegmon of free fluid and/or air confined by the surrounding omentum and ileocecal loops. This phlegmon can undergo liquefactive necrosis and bacterial super-infection forming an abscess. On CTAP this appears as a separately organized thick-walled enhancing fluid collection with air (Fig. 5) [40]. However, if it is uncontained diffuse peritonitis ensues with abdominal rigidity, ileus and severe sepsis. CTAP can show scattered free air and moderate fluid, diffuse mesenteric and omental inflammatory haze and stranding, thickened enhancing peritoneum, ileus with reactive enterocolitis, multiple intraperitoneal and possibly intrahepatic abscesses, fistulas and portal venous thrombophlebitis (Fig. 6) [15,20,38,40].

4.3. Tip appendicitis

A rare variant of typical appendicitis is tip appendicitis. This is when a typical focus, such as an appendicolith or lymphoid hyperplasia,
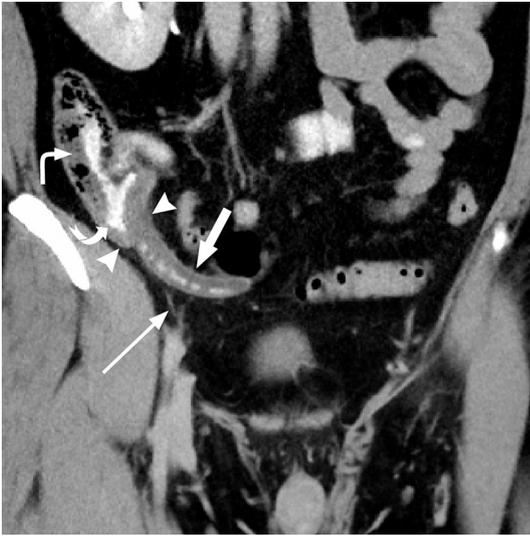
Fig. 3. Arrow sign.

38 yr old male presents with RLQ pain. Coronal contrast enhanced CT image shows diffusely thick walled appendix (thick arrow) with adjacent mild mesenteric inflammation (thin arrow) consistent with acute appendicitis. There is inflammation extending to the base of the cecum which is smoothly thickened (arrow heads), the funneled appearance of the cecal tip causes "arrowhead shape" of the oral contrast in the cecum (curved arrow) pointing towards the appendiceal orifice. Apparent wall thickening and pseudopneumatosis in the superior aspect of the cecum and adjacent ascending colon (bent arrow) due to incomplete mixing of oral contrast with bowel content.

causes an obstruction towards the mid to distal aspect of the appendiceal lumen. This results in the same characteristic inflammation and time course as outlined above, however it is limited to the distal end of the appendix (Fig. 7) [41]. It is a common radiological pitfall and missing the diagnosis can be avoided by tracing the entire appendix from the cecal base to its distal tip.

4.4. Management of typical acute appendicitis

Once the diagnosis of typical uncomplicated acute appendicitis has been established, the patient should be given preoperative fluid resuscitation and a single dose of intravenous antibiotics with anaerobic and gram negative coverage [42]. Emergent surgical appendectomy is the standard of care for uncomplicated appendicitis without perforation. The decision for laparoscopic versus open appendectomy should be made by the surgeon based upon experience, severity of disease, and patient factors including comorbidities and habitus. If there is no contraindication for abdominal insufflation, a laparoscopic approach is usually preferred given the lower rate of wound complications and deep surgical site infections associated with the laparoscopic approach compared to open surgery. In addition, laparoscopy is associated with a slight but significant reduction in postoperative pain and shorter recovery time. The conversion rate to open appendectomy is 1–2% [43–45].

Several European studies have evaluated the role of non-operative management to operative appendectomy in patients with uncomplicated, acute appendicitis. A randomized clinical trial was published in JAMA in 2015 comparing antibiotic therapy in 257 patients to laparoscopic appendectomy in 273 patients. Antibiotic therapy consisted of 3 days of IV ertapenem followed by 7 days of oral levofloxacin
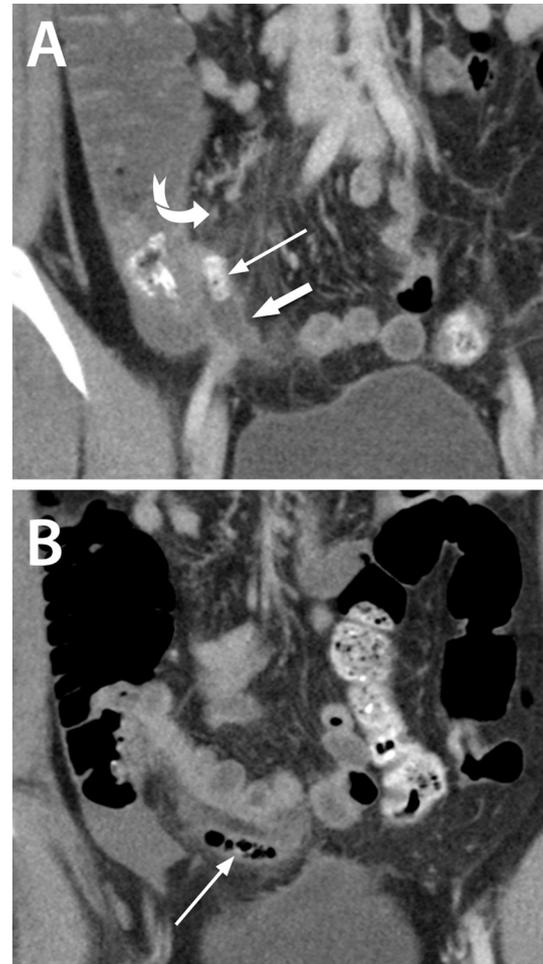

Fig. 4. Necrotizing appendicitis.

26 yr old female with two-day history of RLQ pain, nausea and emesis. Coronal contrast enhanced CT images show (a) an obstructing appendicolith (thin arrow) in the base of the appendix which is dilated and fluid filled (thick arrow) with adjacent mesenteric fat stranding (curved arrow) and (b) multiple bubbles of air in the mid and distal appendix (thin arrow) suspicious for intramural air. No pneumoperitoneum or abscess. Treated with appendectomy and pathology confirmed necrotizing appendicitis.

500 mg once per day and metronidazole 500 mg three times daily. There was a 15–25% rate of recurrent appendicitis in the medically managed patient cohort. The study was unable to prove non-inferiority of antibiotic treatment with a pre-specified non-inferiority margin of 24%. As such, the recommendation for management of acute, uncomplicated appendicitis remains surgical appendectomy in the United States [45,46].

More delayed presentations with gangrenous appendix or with early perforated appendix but without phlegmon or abscess are also treated with emergent appendectomy. These patients however usually present with severe systemic illness needing aggressive fluid resuscitation in addition. Therefore CT features of a gangrenous appendix or early perforation, such as a shaggy walls, patchy enhancement, moderate ascites and free air, should be emphasized in reports. The inflamed friable nature of these appendices results in up to 10–20% of patients developing a postoperative abscess. A laparoscopic approach is still preferred in delayed presentations due to fewer wound infections, but

Fig. 5. Perforated appendicitis with abscess.
18 yr old male presents with severe RLQ pain associated with fever. RLQ tenderness and guarding on exam with elevated WBC of 22.5. Coronal contrast enhanced CT image shows a dilated fluid filled appendix containing calcified appendicolith (thin arrow), and perforation along it's medial wall forming a walled off peri appendiceal abscess containing multiple bubbles of air and appendicolith (thick arrows). Treated with CT guided drainage and antibiotics.

also has a slightly higher risk of abscess formation when compared to open appendectomy. Conversely, patients who present with acute perforated appendicitis complicated by a phlegmon or abscess do not undergo emergent appendectomy. This is associated with increased morbidity from postoperative fistulas, abscess formation, and conversion to an ileocecectomy or right hemicolectomy. Due to the increased operative morbidity, non-operative management is considered standard of care. Patients with a phlegmon, small fluid collection, or collections not amenable to drainage are treated with intravenous antibiotics (Fig. 8) [38,48]. Formed abscesses and large fluid collections greater than 4 cm are treated with antibiotics and percutaneous drainage [15]. The method of drainage is dependent on the collection size and location; methods for drainage include percutaneous, transvaginal, transrectal, or laparoscopic. Interval appendectomy was previously recommended for patients treated non-operatively 6 to 12 weeks after resolution of symptoms, however the rate of recurrent appendicitis has proven to be small, approximately 8% at 8 years. As a result, interval appendectomy is only offered to patients with recurrent appendicitis or an appendicolith given it increases the rate of recurrent appendicitis [42–48]. There is also a higher incidence of underlying appendiceal neoplasms found in specimens at interval appendectomy and colonoscopies are often recommended in patients managed non-operatively [49].

5. Atypical appendiceal pathologies

Although typical acute appendicitis is very common, a unique set of less frequent, but clinically significant atypical pathologies can afflict the appendix. These entities are broadly categorized as neoplasms, chronic inflammatory diseases and special cases [8,9]. Standard appendectomy or percutaneous drainage of unrecognized atypical appendiceal pathology can result in catastrophic complications, such as tumor seeding and fistulas. CT, clinical characteristics and management that differentiate them from typical appendicitis are summarized below.
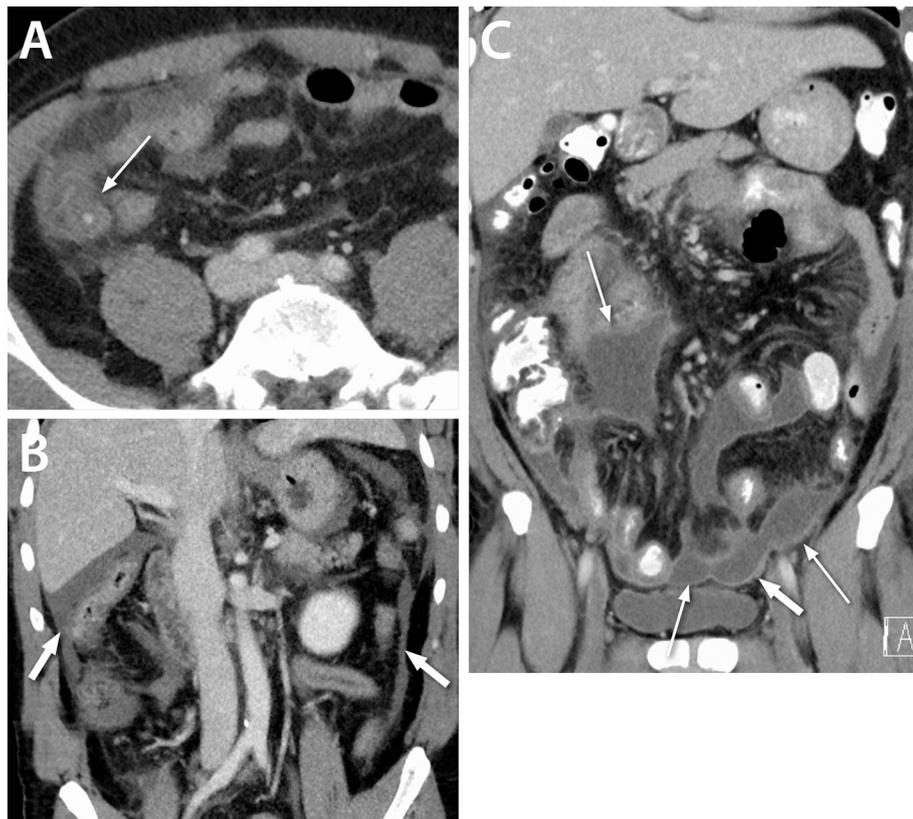

Fig. 6. Perforated appendicitis with peritonitis.
47 yr old male with 2-day history of mid abdominal pain, nausea, emesis, fever and chills. Contrast enhanced axial and coronal CT images show (a) dilated appendix with appendicolith and shaggy discontinuous wall (thin arrow) and (b) diffuse ascites (thick arrows) indicating perforation with peritonitis. Emergent surgery revealed gangrenous perforated appendix. Subsequent contrast enhanced coronal CT image (c) shows increase in peritoneal fluid, loculations (thin arrows) and peritoneal enhancement (thick arrow) consistent with developing abscesses which were treated with percutaneous drainage.
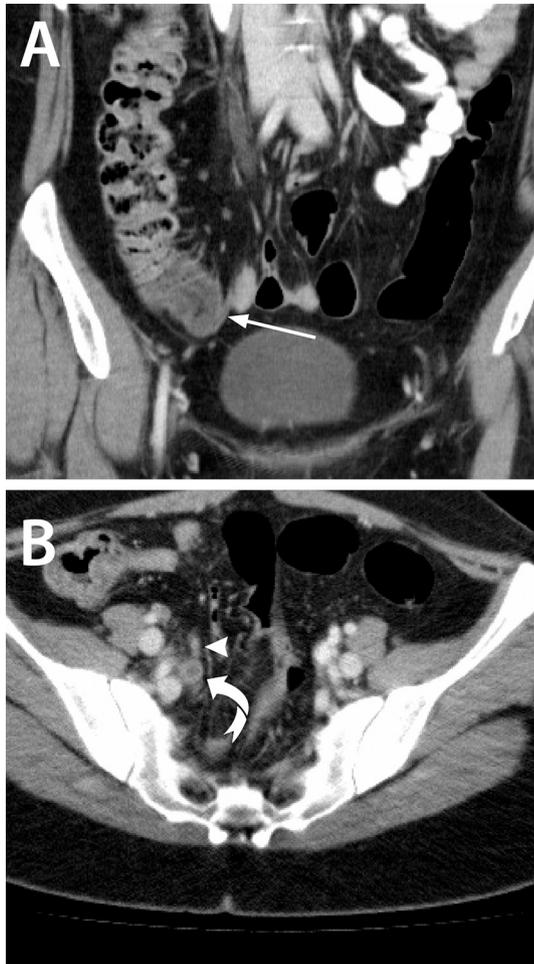
Fig. 7. Tip appendicitis.
5 yr old male presents with 3-day history of RLQ pain and anorexia. Contrast enhanced coronal CT image (a) shows a normal thin caliber appendix arising from the base of the cecum (thin arrow). Axial CT image shows a normal caliber mid appendix (arrow head) and a dilated, fluid filled and thick walled distal tip (curved arrow) consistent with tip appendicitis.

5.1. Neoplasms & management

Primary neoplasms of the appendix are rare and found in 0.5–1% of appendectomy specimens from older adults [50–52]. A 2002 study of patients with pathology confirmed primary neoplasms of the appendix showed that 40% presented with acute appendicitis from malignant obstruction [53]. Less frequent presentations include palpable masses, intussusception, GI bleeding, ureteral obstruction and hematuria or abdominal distension from pseudomyxoma peritonei. Primary appendiceal neoplasms are usually of epithelial, carcinoid or lymph cell origin with other very rare histologic types [54]. Secondary neoplasms of the appendix, particularly cecal carcinoma occur more often [7].

Epithelial tumors of the appendix constitute about 30% of all primary neoplasms and occur more often in older adults. They can be mucinous or non-mucinous, which both range from benign to malignant. Mucinous epithelial tumors were recently reclassified as adenoma, low grade appendiceal mucinous neoplasm, high grade appendiceal neoplasm and mucinous adenocarcinoma [52]. They are the most common appendiceal neoplasm diagnosed on CT due to the formation
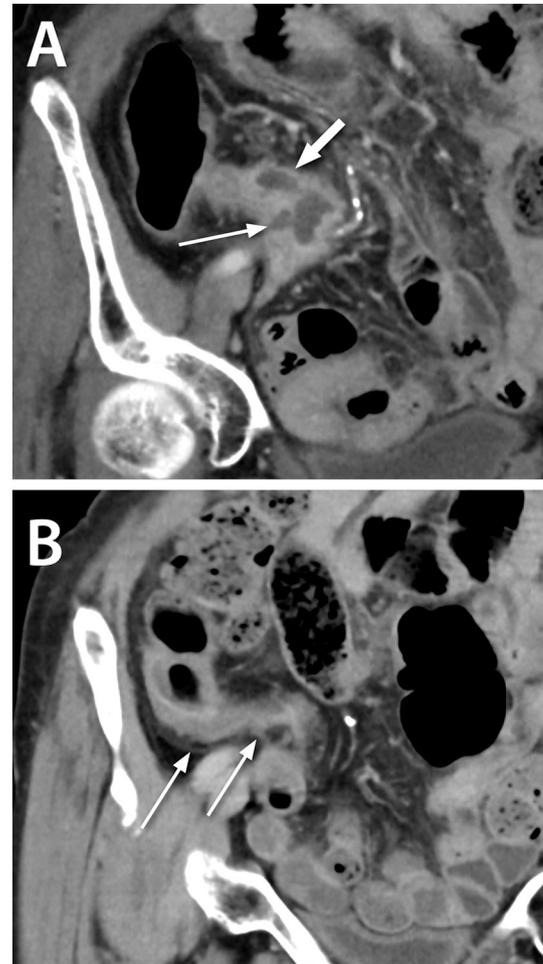

Fig. 8. Non operative management of complicated appendicitis.
82 yr old female presents with right lower quadrant pain. Contrast enhanced coronal CT images show (a) findings of appendicitis with abnormal thick walled appendix (thick arrow), adjacent phelgmon and micro abscess (thin arrow) which are too small to drain. Patient was managed non operatively with antibiotics and follow up CT in 4 weeks (b) reveals almost complete resolution with residual appendiceal thickening (thin arrows). Patient did not undergo interval appendectomy.

of a visible mucocele. It is important to note that a mucocele is not a neoplasm or pathologic entity, but rather a descriptive term of a mucus distended appendix that could be from an underlying chronic neoplasm, such as a mucinous epithelial tumor, or a non-neoplastic obstruction. This generally presents on CTAP as a well-circumscribed, dilated cystic appendix distended by low density intraluminal mucus contents [50–55]. Non-neoplastic mucoceles are essentially mucus retention cysts and are usually less than 2 cm in diameter on CTAP [56,57]. Mucoceles due to mucinous epithelial tumors are often greater than 2 cm in diameter and can contain curvilinear mural calcifications (Figs. 9 and 10). Characteristics such as mucocele size, wall calcifications or periappendiceal stranding cannot further stratify the type of mucinous neoplasms, but irregular wall thickening and enhancing nodules associated with a mucocele can favor a higher grade mucinous adenocarcinoma (Fig. 11). Higher grade malignant mucinous tumors may also have extra-appendiceal deposits of mucin confined to the periappendiceal space or disseminated as pseudomyxoma peritonei.
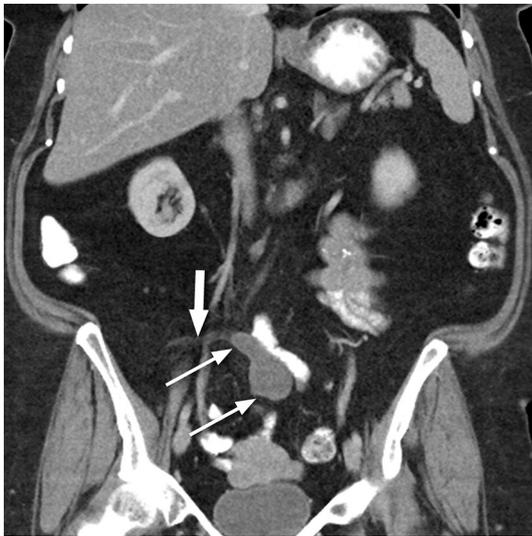
Fig. 9. Low grade appendiceal mucinous neoplasm.
62 yr old female with history of breast cancer and asymptomatic abdomen. Contrast enhanced coronal CT image shows normal caliber proximal appendix (thick arrow) and dilated fluid filled distal appendix (thin arrows) with lack of adjacent mesenteric inflammation. Patient underwent non emergent appendectomy with pathology revealing the distal appendix to be filled with white mucoid material caused by an obstructing low grade appendiceal mucinous neoplasm confined to the appendix.

Imaging features of pseudomyxoma peritonei include loculated peritoneal collections which may have linear or punctate calcification, malignant ascites, omental caking, and peritoneal implants on solid organs with scalloping of the organ margins due to mass effect (Fig. 12) [52,54]. Tumor implants tend to involve the ovaries and gastrointestinal tract which may cause bowel obstruction. Non-mucinous or colonic type epithelial tumors are less common compared to the mucinous type and range from polyps and adenomas to adenocarcinoma. They often cause obstruction and present as an acute inflammatory appendicitis. Unfortunately, non-mucinous tumors frequently exhibit advanced local invasion and have a poor prognosis, particularly signet ring cell types (Fig. 13) [50–54,58].

Primary carcinoid tumors have the highest prevalence in surgical specimens and make up approximately 66% of all appendiceal neoplasms [50,51,59]. However, they are difficult to detect on CT since 70% of the time they are often less than 1 cm and involve only the distal third of the appendix [60]. They usually occur in young adults and have a very indolent course with a 5-year survival of 90% [59–62]. Rarely carcinoids can be more aggressive and induce carcinoid syndrome as with goblet cell types [62]. These can be detected on CTAP due to their aggressive features including enhancing mural thickening and local invasion [60]. They can appear similar to small bowel carcinoids and present as a mesenteric mass in the ileocecal region with central calcification and spiculated margins due to a desmoplastic reaction. Golbet cell carcinoids also have a tendency to metastasize to the liver and peritoneum (Fig. 14) [62].

Primary lymphoma of the appendix is the least common of the three main subtypes despite the gastrointestinal tract being the most common site for extranodal disease [50,51,61,62]. All reported cases have been non-Hodgkin lymphoma. On CTAP appendiceal lymphoma presents as an enlarged appendix with a persistent vermiform appearance and a
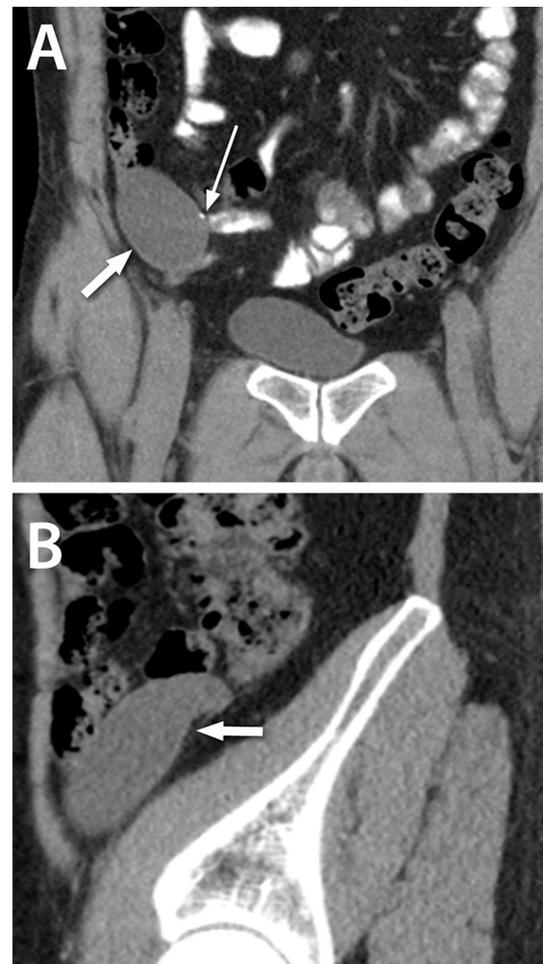

Fig. 10. High grade appendiceal mucinous neoplasm.
45 yr old asymptomatic male. Non enhanced coronal (a) and sagittal (b) CT images show a cystic dilated appendix measuring 3.5 cm in diameter (thick arrows) and containing minimal peripheral rim calcification (thin arrow). Patient was treated with appendectomy. Pathology revealed high grade appendiceal mucinous neoplasm.

blind-ending loop. It is important to not confuse this for an adjacent bowel loop or hydrosalpinx [63,64].

Several extremely rare primary neoplasms have also been seen to afflict the appendix including kaposi's sarcoma, mesenchymal tumors, GIST and ganglioneuromas or paragangliomas. They generally appear on CTAP as heterogeneous soft tissue nodularity or masses and can only be differentiated with history and pathology [50–54,61,62].

Secondary malignancies of the ileocolic region particularly cecal carcinoma can involve the appendix or result in acute appendicitis from obstruction. Approximately 95% of cecal cancers are adenocarcinoma that usually present with anemia from bleeding, weight loss and/or RLQ mass. Adenocarcinoma is notorious for obstructing and causing perforation of the colon or appendix. On CTAP this appears as enhancing irregular soft tissue density focal wall thickening of the cecum (Fig. 15) [65]. In contrast the inflammatory wall thickening in the base of the cecum due to typical acute appendicitis will be smooth resulting in the "arrowhead" sign (Fig. 3) [39].

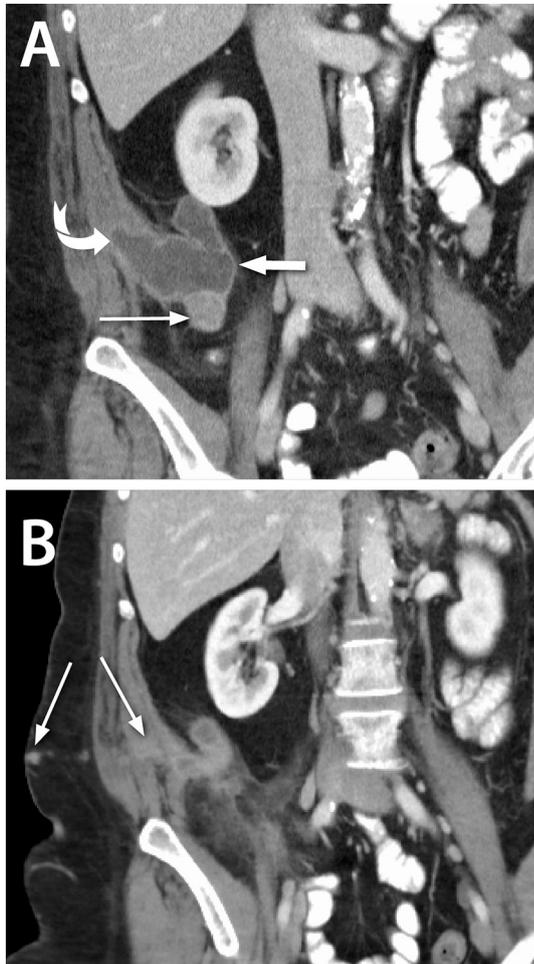Regardless of the pathology, key CT findings for primary

Fig. 11. Mucinous adenocarcinoma of appendix.

65 yr old F presented with right lower quadrant pain. Contrast enhanced coronal CT image (a) shows a markedly dilated appendix (thick arrow), that invades the adjacent abdominal wall (curved arrow) and has an enhancing mural nodule (thin arrow). Misdiagnosed as perforated appendicitis with abscess and treated with drain placement. Subsequent contrast enhanced coronal CT image (b) shows development of tumor implants along the drain tract (thin arrows). Treated with resection of the appendiceal mass and abdominal wall resection.

appendiceal neoplasms include enhancing soft-tissue density (intraluminal or intramural), substantial dilation with less than expected inflammation for an appendicitis, intramural calcifications, surrounding invasion or mass effect, intussusception, periappendiceal low attenuation mucin deposits and pseudomyxoma peritonei [52–58].

The above CT findings along with an indolent clinical presentation should raise suspicion for a neoplastic etiology. Alerting the surgeon is necessary for appropriate treatment planning and prevention of complications. Patients with tumors recognized on preoperative imaging will require staging evaluation for the appropriate oncologic surgery [54]. Tumors less than 1 cm that do not involve the base or the mesoappendix are managed with appendectomy. However, tumors that are larger than 2 cm, involve the base of the appendix, or extend into the mesoappendix are treated with a right hemicolectomy and regional

lymphadenectomy. Adenocarcinoma of the appendix is treated as cecal adenocarcinoma and requires right hemicolectomy, regional lymphadenectomy and adjuvant chemotherapy for patients at risk for disease recurrence. Percutaneous drainage of a mucinous appendiceal or colonic origin carcinoma masquerading as appendicitis with an abscess can result in chronic fistulas, tumor seeding of the drain tract and carcinomatosis with or without pseudomyxoma peritonei. In general, the lesions should be managed surgically unless the patient is under palliative care [54,66,67]. Treatment of simple appearing mucoceles isolated to the appendix is appendectomy. Careful dissection of the appendix is necessary to avoid rupture. Complex appearing mucoceles with ileal or cecal involvement require right hemicolectomy, and often adjacent organ resection if locally advanced [68]. The treatment of pseudomyxoma peritonei is extensive cytoreductive surgery and intraperitoneal chemotherapy [69].

5.2. Chronic inflammatory diseases & management

Inflammatory bowel disease (IBD) can afflict the appendix in the setting of both Ulcerative Colitis (UC) and Crohn's disease [70,71]. CT can differentiate UC or Crohns ileocolitis with appendiceal involvement from simple appendicitis by identifying inflammatory wall thickening of the terminal ileum and other involved bowel (Fig. 16). Without additional findings of IBD it is difficult to differentiate the inflammatory appearance from a typical appendicitis. It is important to suggest the possibility of IBD since standard appendectomy can result in complications such as stump leakage or fistula formation. Rather these patients are usually managed medically [72–78].

Xanthogranulomatous inflammation is a well described entity involving other organs especially the kidney and gallbladder, however it is very rare in the appendix (only 2% of all surgical specimens). It has a chronic indolent course and can mimic malignancy both clinically and radiologically, warranting excision with diagnosis made on pathology. It is usually found retrospectively on surgical pathology and has no unique features on CTAP. It also does not have the same risk of fistualization as IBD (Fig. 17) [65,70,79].

Endometriosis is another inflammatory condition that can involve the appendix. It is usually asymptomatic and occurs in less than 1% of pelvic endometriosis. When symptoms do occur they are usually melena with colicky pain that cycle during menstruation. On emergent CTAP the findings are very similar to acute appendicitis and pre-operative diagnosis is heavily dependent on clinical history. MRI may reveal other findings of pelvic endometriosis to increase specificity [70,80,81].

Vasculitis from polyarteritis nodosa rarely can involve small and medium-size muscular arteries that supply the appendix. About one third of patients have abdominal pain, but complications requiring surgery are rare with gangrenous appendicitis reported in less than a dozen patients. This is difficult to identify on CT and is usually diagnosed on surgically pathology [82,83].

5.3. Special conditions & management

Stump appendicitis occurs when simple ligation of the appendix is performed and the stump is inadequately invaginated or there is an incomplete resection at the cecal base. This leaves a remnant stump of the appendiceal lumen that can form a diverticulum and incur recurrent inflammation. Visualization of the base of the appendix intraoperatively to assure a stump length no greater than 3 mm is

Fig. 12. Pseudomyxoma peritonei.

49 yr old male presents with weight loss, abdominal pain and bloating. Coronal contrast enhanced CT image shows a fluid filled appendix engulfed by mucinous ascites with punctate foci of calcification (thick arrow), peritoneal soft tissue implants and omental caking (thin arrows) and scalloping of the liver surface by mucinous ascites (arrow head) consistent with pseudomyxoma peritonei. CT guided biopsy of the omentum revealed mucinous adenocarcinoma of appendix.

considered optimal to avoid stump appendicitis. However, when it occurs it presents clinically as acute or subacute abdominal pain and can occur anywhere from 2 months to 50 years after appendectomy. Stump appendicitis is associated with a high rate of perforation (70%). Specific findings on CTAP include pericecal fat stranding, fluid in the right paracolic gutter, focal thickening of the cecal apex causing an arrow sign, an ileocecal mass, abscess formation and occasionally an appendicolith (Fig. 18). Recognition of stump appendicitis on CT is heavily dependent on knowing the prior history and recognizing that the history of a prior appendectomy does not always exclude appendicitis [84–88]. Treatment can be targeted endoscopically with snare electrocautery or in more complex cases with ileocecectomy or right hemicolectomy. If there is an associated abscess, the patient should be managed medically with drainage if with subsequent endoscopic or surgical resection [44].

Acquired pseudodiverticula can occur even without surgical intervention. These outpouchings can become acutely or chronically inflamed and present with appendiceal diverticulitis similar to elsewhere in the bowel. CTAP shows periappendiceal inflammation surrounding a single diverticulum or diffusely throughout the appendix. Appendiceal diverticulitis is associated with a higher mortality than traditional appendicitis given the weak wall perforates easily. These are treated similar to uncomplicated and complicated typical appendicitis [89–91].
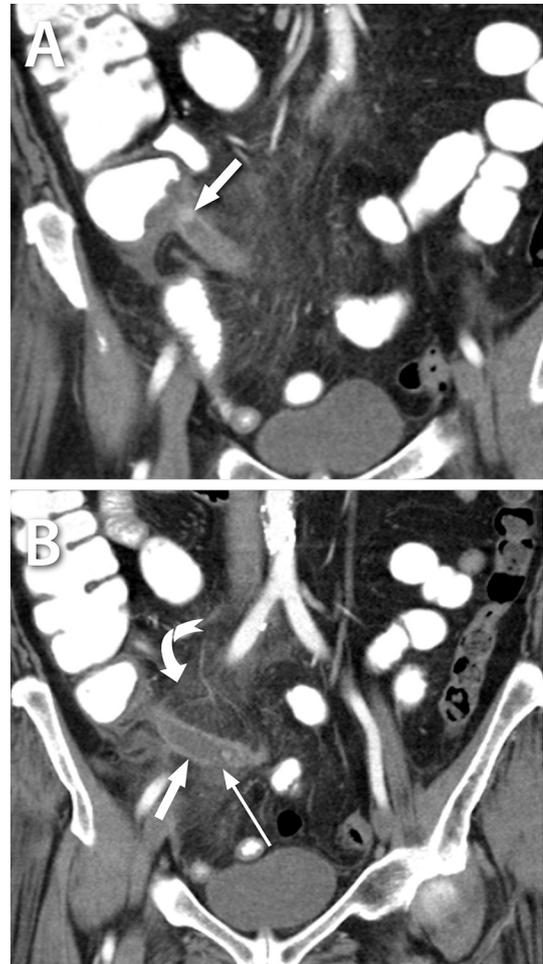

Fig. 13. Adenocarcinoma of the appendix.

76 yr old female presents with acute right lower quadrant pain. Contrast enhanced coronal CT images reveal (a) small enhancing nodule in the base of the appendix (thick arrow) causing obstruction, (b) shows a dilated appendix with wall enhancement (thick arrow), peri appendiceal inflammation (curved arrow) and focal discontinuity of the appendiceal wall (thin arrow) indicating acute appendicitis with perforation. Patient treated with appendectomy. Pathology reveals poorly differentiated adenocarcinoma of signet ring type. Patient was subsequently treated with neoadjuvent chemotherapy followed by right hemicolectomy.

Amyand's hernia is defined as the presence of the appendix within an inguinal hernia sac and may present as an incarcerated hernia on CTAP (Fig. 19). A retrospective study published in the Journal of Pediatric Surgery reviewed 46 Amyand's hernias; 33 hernias contained normal appendices, 9 inflamed, 1 perforated, and 3 adherent to the hernia sac. If appendiceal inflammation is present in the Amyand's hernia, an appendectomy and tissue hernia repair is recommended. Normal appendices in the setting of Amyand's hernia may be approached with hernia reduction and mesh repair [92–97].
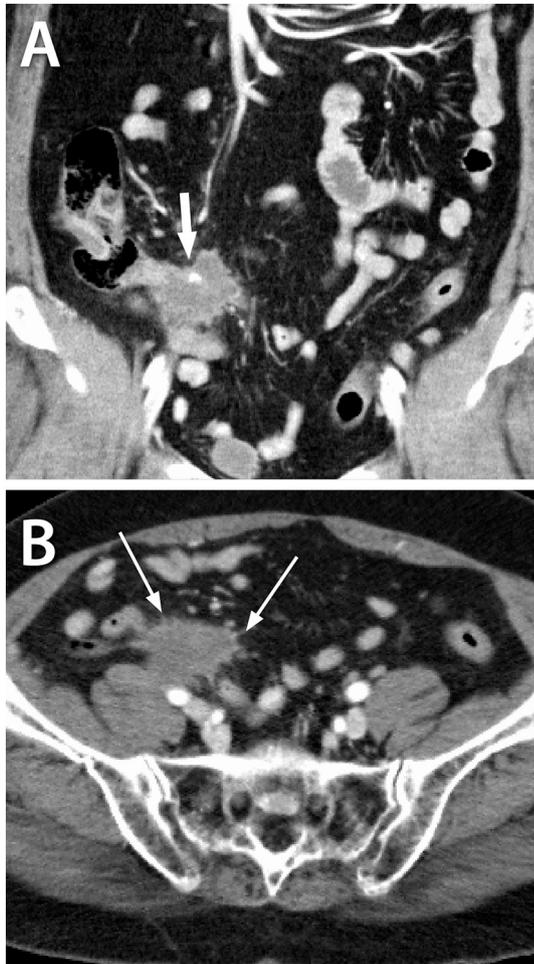
Fig. 14. High grade neuroendocrine tumor of the appendix.
60 yr old female presents with intermittent RLQ pain of two-month duration. Contrast enhanced coronal (a) and axial (b) CT images show an irregular soft tissue mass arising from the distal appendix that has invaded the adjacent mesentery (thick arrow) and has spiculated margins (thin arrows). Patient had peritoneal carcinomatosis (not well depicted on these images) and was offered cytoreductive surgery and intraperitoneal chemotherapy.
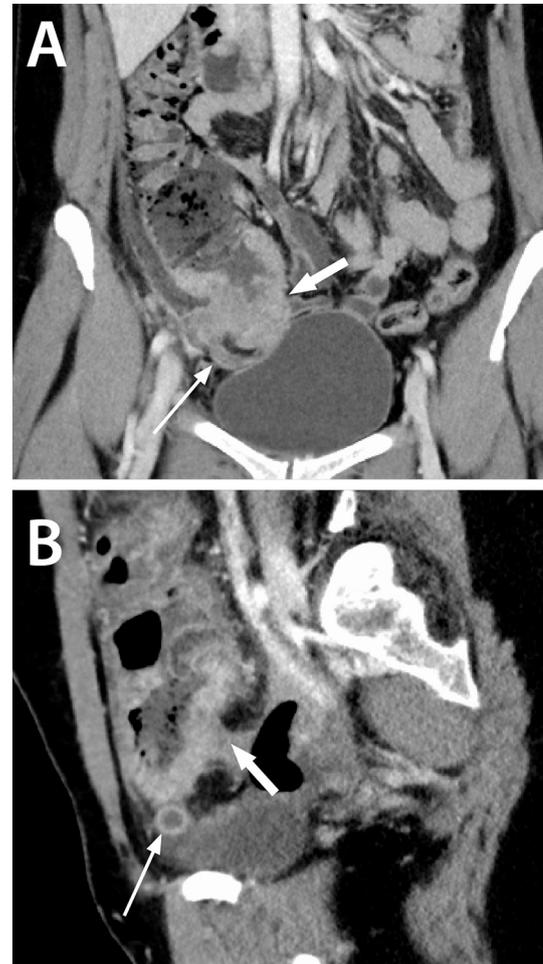
Fig. 15. Adenocarcinoma of the cecum.
48 yr old female presents with right lower quadrant pain. Contrast enhanced coronal (a) and sagittal (b) CT images show irregular wall thickening of the base of the cecum concerning for neoplasm (thick arrows) which causes secondary inflammation of the appendix which is fluid filled with thin enhancing wall (thin arrows). Colonoscopy confirmed cecal mass, adenocarcinoma on pathology and was treated with right hemicolectomy.

6. Right lower quadrant mimickers

It is important to note, that other non-appendiceal right lower quadrant pathologies can mimic the presentation of acute appendicitis, including tubo-ovarian abscesses, ruptured corpus luteal cysts, epiploic appendagitis, omental infarcts, inflammatory or infectious terminal ileitis, typhlitis, infectious or ischemic colitis, Meckel's diverticulitis, mesenteric adenitis or enteritis and cecal diverticulitis. The best way to differentiate these entities on CTAP from an acute appendicitis is to first identify a normal appendix and then determine where the focus of inflammation is greatest based on the largest burden of fat stranding, free fluid or bowel wall edema. However, in the setting of a non-visualized appendix with perforation of the above entities it can be very difficult to localize the etiology and exclude an acute perforated appendicitis. Often times this scenario requires a differential diagnosis that can be narrowed down by identifying normal non-appendiceal organs, localizing free air or a fluid collection in or near a specific organ and correlating with history of prior appendectomy, gynecologic diseases and other surgical procedures [82].

7. Conclusion

CTAP is increasingly used for the diagnosis of acute appendicitis. While the vast majority of acute typical appendicitis do not present a diagnostic challenge on CT, less common atypical appendiceal pathologies, have significant clinical implications in patient management. The

Fig. 16. Crohn's disease involving appendix.

30 yr old male with history of Crohn's disease presents with right lower quadrant pain. Contrast enhanced coronal CT image shows a markedly thickened appendiceal wall with mucosal hyperenhancement (thick arrows) and similar findings involving the terminal ileum (thin arrows) consistent with active Crohn's disease. Patient was medically managed without surgery.
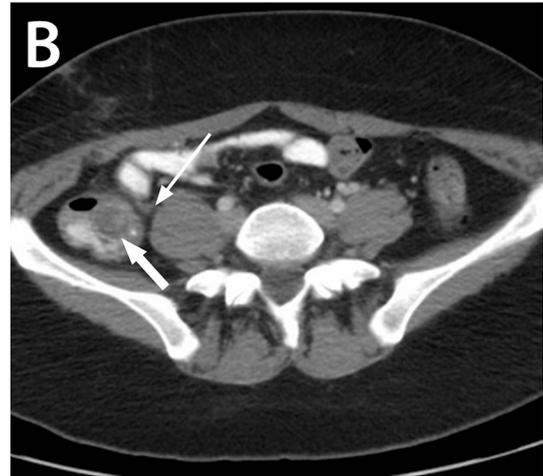

Fig. 17. Xanthogranulomatous appendicitis.

64 yr old male presents with RLQ pain and palpable mass. Non contrast coronal CT image shows an ill-defined soft tissue lesion adjacent to the base of the cecum (thick arrow) with non-visualization of a normal appendix and adjacent mesenteric strandying (thin arrow). Patient underwent right hemicolectomy with diagnosis of Xanthogranulomatous appendicitis made on histopathological examination.

most important suspicious CT findings for atypical pathologies include disproportionate dilation of the appendix relative to the degree of inflammation, enhancing soft tissue density, mural calcifications and

Fig. 18. Stump appendicitis.

36 yr old female with prior history of appendectomy presents with right lower quadrant pain. Contrast enhanced coronal and axial CT images (a and b) show focal mass like thickening of the cecal apex (thick arrows) with minimal adjacent mesenteric inflammation (thin arrow). Managed with partial cecetomy and pathology confirmed stump appendicitis.
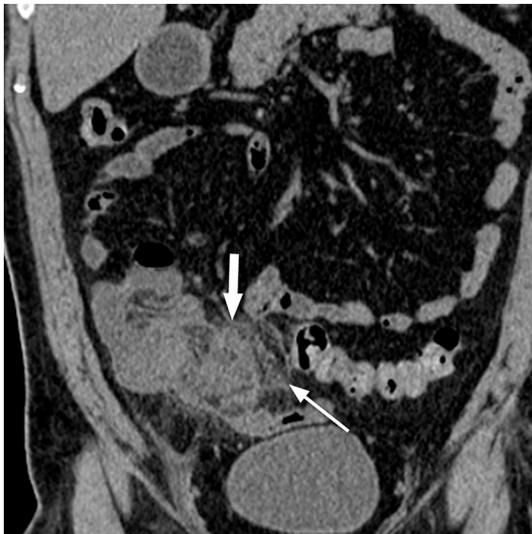
secondary findings of systemic diseases including inflammatory bowel disease, vasculitis and endometriosis. Although rare, these findings in the setting of an abnormal appendix on CT should raise concern for an underlying atypical process, especially when there is chronic symptomatology. The most difficult scenario is when patients present with acute symptoms due to appendicitis from an underlying neoplasm. While it is not always possible to make a preoperative diagnosis in such cases, the presence of certain features as discussed should prompt the radiologist to include an underlying neoplastic etiology in the differential diagnosis and alert the surgeon.

Fig. 19. Amyand hernia.
70 yr old male presents with right inguinal bulge. Contrast enhanced coronal CT image shows a right inguinal hernia (arrow heads) containing thick walled appendix (thick arrow) and mesenteric fat with inflammation (thin arrow). Patient treated surgically with repair of the hernia.

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