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IMAGE OF THE MONTH

Adenocarcinoma is not always the diagnosis — colon neoplasia in patient with long-standing ulcerative colitis under long-term prednisone maintenance therapy

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Image of the month

We present the case of a 51-year-old non-smoking male with a 16-years history of left-sided UC (E2 of the Montreal Classification). Due to irregular follow-up and poor control of UC with oral Mesalazine he frequently self-medicated with oral prednisolone. He had never received other immunosuppressant drugs and denied family history of colonic cancer. A surveillance colonoscopy 1-year prior showed signs of active colitis (Mayo Score: 3) but random colon biopsies did not reveal any dysplasia. The patient was admitted to our department with abdominal pain and

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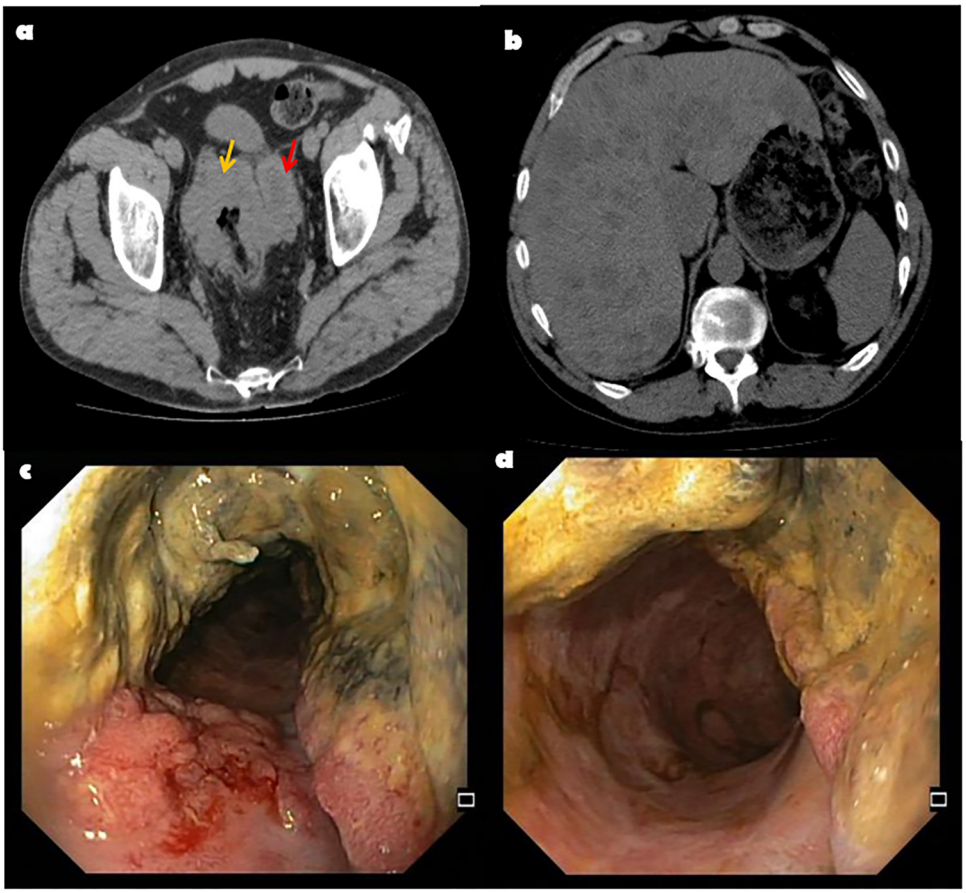

Figure 1 Abdominal CT image, in axial view, showed a marked irregular wall thickness in the rectum-sigmoid transition (yellow arrow) and several proeminent adjacent lymph nodes (red arrow), the largest one with 5.3 cm, compatible with neoformative lesion (a). CT image, in axial section, revealed multiple hypodense hepatic lesions in both hepatic lobes consistent with metastatic lesions (b). Colonoscopy view (c). Recto-sigmoidoscopy revealed a colonic substenosis caused by a large ulcerated and circular mass of about 4 cm of length, originating above the rectosigmoid junction (d).

tenesmus of 8 days' duration. Blood tests revealed elevated white blood cell count ($13.1 \times 10^3/\mu L$, normal range: $4.0-10.5 \times 10^3/\mu L$), C-reactive protein (20.8 mg/L, normal range: < 2.9 mg/dL] but normal haemoglobin level. Abdominal CT scan showed a large mass above the rectosigmoid junction with several prominent adjacent lymph nodes in the sigmoid mesocolon. Multiple hypoattenuating lesions in both liver lobes concerning for metastasis were also observed (Fig. 1a,b). Recto-sigmoidoscopy revealed a colonic substenosis caused by a large ulcerated and circular mass of about 4 cm of length, originating above the rectosigmoid junction (Fig. 1c,d).

Anatomopathological examination of biopsies performed from described lesion showed cells with scant cytoplasm and round to oval hyperchromatic nuclei. Immunohistochemical staining was positive for pancytoqueratin MNF116 and neuroendocrine markers, chromogranin A (focally) and synaptophysin and CD56 (diffusely) (Fig. 2a,b,c,d,e). These findings were consistent with diagnosis of the high grade small cell neuroendocrine carcinoma. A somatostatin receptor scintigraphy confirmed the presence of local lymph node metastasis and multiple liver metastasis (Fig. 2f). The definitive TNM stage was compatible with IV stage and the patient was scheduled for palliative radiotherapy and chemotherapy with cisplatin and etoposide.

Long-standing, extensive and severe inflammation activity UC are known risk factors for the development of colorectal tumors [1]. NEC in IBD setting are exceptionally rare (less than 1% of colorectal tumors) with just over 15 cases reported in the literature [1,2]. However, whether IBD increases the risk of developing NEC is not clear [2]. According to Siegel et al, neuroendocrine differentiation might evolve from multipotential cells in dysplastic epithelium, suggesting that long-standing inflammation might be involved in its pathogenesis [3].

Furthermore, NEC in IBD patients seems to be extremely aggressive and carry a dismal prognosis, with about 70% of patients presenting with metastatic disease at diagnosis and a reported median survival rate of 10 months [2].

Despite of their rarity, we highlight the importance to recognize the NEC in differential diagnosis of colonic tumors in UC patients given the aggressive behavior and poor outcomes of this entity. It is also important to clarify the pathogenesis of the NEC and to identify risk factors for the

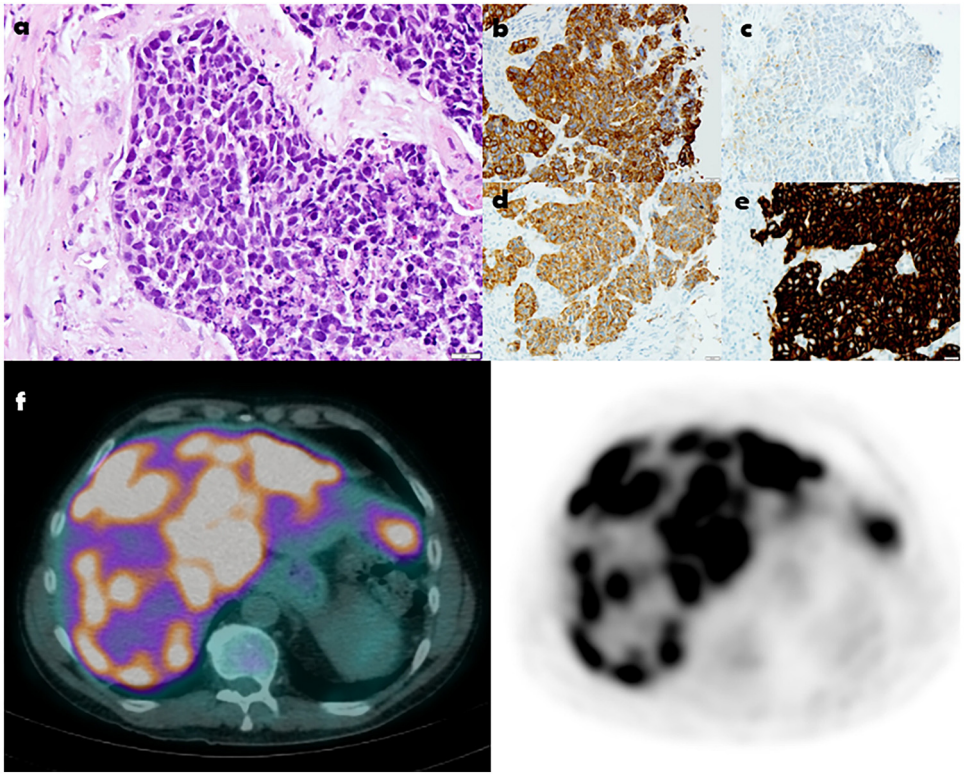

Figure 2 Histological examination showed cells with scant cytoplasm and round to oval hyperchromatic nuclei (H&E 400x) (a). Immunohistochemically, neoplastic cells express pancytoqueratin MNF116 and neuroendocrine markers, focally chromogranin A and diffusely synaptophysin and CD56 (images b,c,d,e, respectively, 400x). A staging somatostatin receptors imaging showed positive marking on local lymph nodes and hepatic parenchyma (f).

development of NEC in patients with IBD, which not only could improve surveillance but might also help with the management of these patients.

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Disclosure of interest

The authors declare that they have no competing interest.

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