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Clinical Images

Castleman's disease presented as a rare unicentric pancreatic mass

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A 44-year-old male patient was initially presented with pain in the upper abdomen. Weight loss, jaundice and digestive disorders were not reported. Blood analysis showed white blood cell count 9.02×10^9, C-reactive protein (CRP) 3 mg/L, and gastrin 38 ng/L. Abdominal ultrasonography revealed tumor in the projection of duodenum and pancreatic head. Colonoscopy and tumor markers (CA19-9, CEA, CA72-4, and AFP) were all negative.

Additionally, abdominal contrast enhanced computed tomography (CT) examination was performed which demonstrated hypervascular tumor lesion, size 43 × 38 × 34 mm, in the lateral aspect of the pancreatic head (Fig. 1). No evidence of extra-pancreatic disease was found. Radical surgical resection of the tumor was conducted through medial laparotomy. Frozen section biopsy finding of well circumscribed nodular tumor mass was at that moment interpreted as nonmalignant hyperplastic peripancreatic lymph node (Fig. 2). Histology showed nodular architecture of lymphoid infiltrate, mostly composed of follicle-like structures with hyalinized and vascularized centers and onion skin-like distribution of surrounding small lymphoid cells (Fig. 3). Immunostaining showed B-cell phenotype ($CD20^+$, $CD5^-$, $CD10^-$, and $CD43^-$) with $CD21^+$ rich dendritic cell networks, focally mantle zone-like areas (focal expression of $CD10^+$ and $Bcl\text{-}6^+$) and a lack of Bcl-2 immunoexpression in germinal centers, but $CD3^+/CD5^+$ small T lymphocytes among polymorphous interfollicular lymphoid infiltrate (Fig. 3).

After an adequate recovery, without complications, the patient was discharged on postoperative day 11. He is now under diagnostic surveillance without the need for further treatment.

Castleman's disease is a rare atypical lymphoproliferative disorder that can easily be misdiagnosed. The etiology is still uncertain and it is thought that the cause is viral infection of the B cell pool and the lymphovascular compartment of lymph nodes [1]. It was first described as a single case by Benjamin Castleman, an American physician and pathologist [2]. Symptoms that can occur are fatigue, high fever, night sweats and loss of appetite. Castleman's disease can be clinically characterized by leucocytosis, increased erythrocyte sedimentation and CRP. In unicentric form of the disease symptoms can also be caused by focal compression. Angiofollicular hypertrophy of the lymph nodes is histological characteristic of Castleman's disease [3–5]. Diagnosis of these lesions can be challenging and surgery is an acceptable curative procedure.

Clinical forms of the disease are unicentric (localized), commonly seen as asymptomatic or with non-specific clinical presentation provoked by compression; and less frequent multicentric, often associated with systemic symptoms, polyadenopathy and worse outcome. Furthermore, there are four histological subtypes with various presentations and morphological features. The most common, hyaline-vascular type tends to present as localized and asymptomatic form, in comparison to plasma-cell type which is usually multicentric and symptomatic form with high fever, weight loss, anemia, night sweats, elevated sedimentation and hypergammaglobulinemia [3–5].

The more common form of the disease is unicentric, as in our patient, and median age of diagnosis is 35 years [6]. However, multicentric type of the disease is usually associated with adverse clinical outcome, generalized lymphadenopathy, sometimes splenomegaly and may be jointed with POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, M-protein and skin), paraneoplastic pemphigus and other paraneoplastic syndromes. Usually affected are male adults. It can also occur in children, more frequently in girls (75%) as plasma cell variant (63%), with good response to corticosteroid therapy. In multicentric disease (plasma cell or plasmablastic histologic type) neoplastic transformation to non-Hodgkin's lymphoma, angiomyoid, vascular or follicular dendritic cell neoplasms or Kaposi's sarcoma is also possible, such as in patients with HIV or other viral infections [3,6,7].

Castleman's disease is manifested by enlarged painless axillar, mediastinal or abdominal lymph nodes. It can mimic various other entities, as in our case where abdominal presentation of the disease is unusual and intraabdominal mass could be misdiagnosed with pancreatic cancer. CT is used to reveal localization, dimension of the mass and its relation to surrounding structures. It is essential to differentiate unicentric from multicentric forms of the

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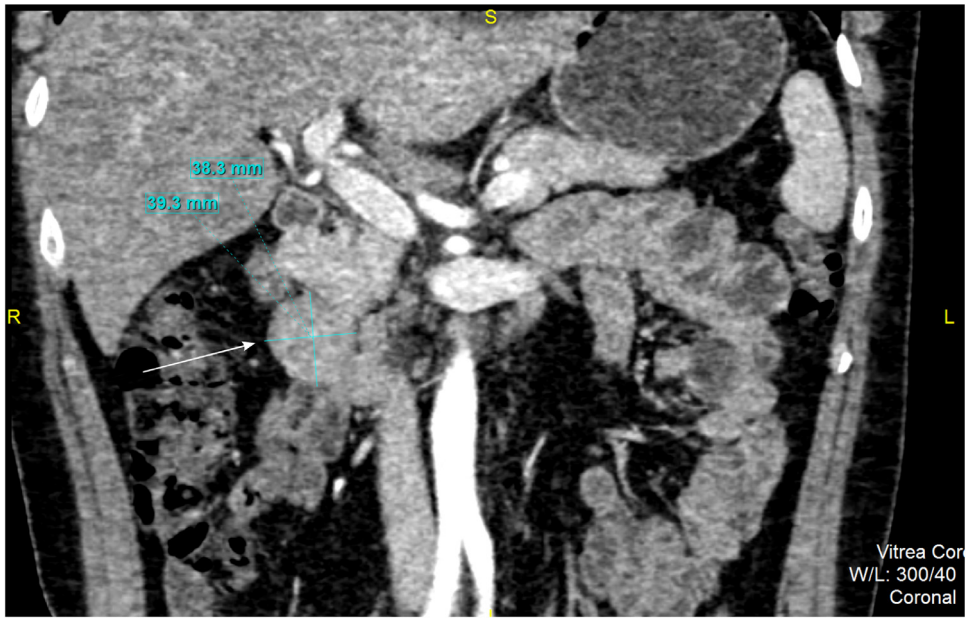

Fig. 1. Hypervascular tumor lesion localized in a projection of pancreatic head (arrow).

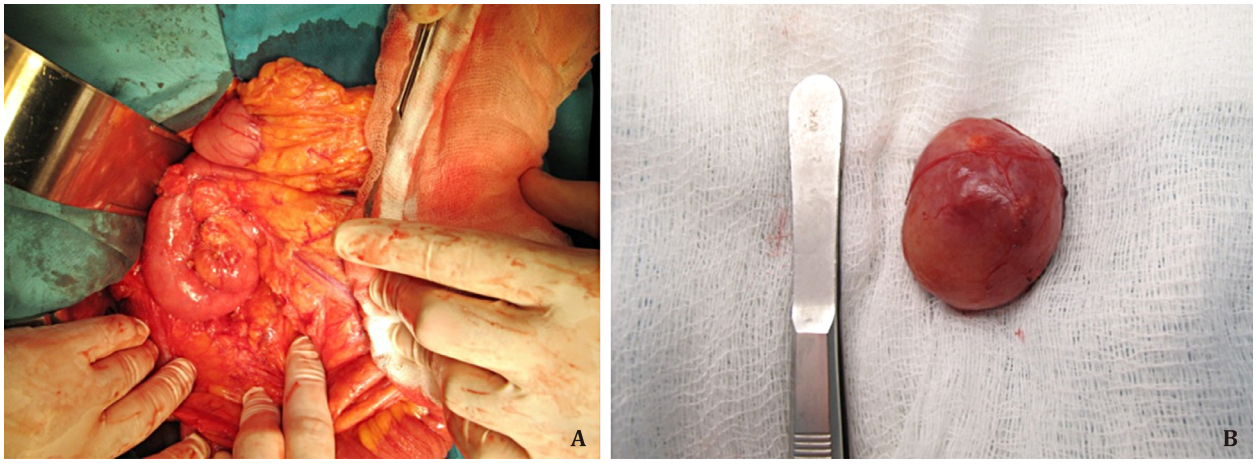

Fig. 2. Intraoperative finding of the mass (**A**) and pancreatic mass after excision (**B**).

disease, which is why CT examination should include neck, chest, abdomen and pelvis [5,7,8].

Surgical resection is the treatment choice for unicentric Castleman's disease [1,5]. Total mass resection should be performed since the recurrences have been reported in cases related to incomplete initial resection. Five-year disease-free survival rate was 81% [1,3,4].

In conclusion, Castleman's disease can be presented as a profound ingrowth of the lymphoid proliferation within the head of the pancreas, as a very unusual localized form, which was initially clinically misdiagnosed as pancreatic cancer. This case is one of very few reported as hyaline-vascular type of localized/unicentric form in the pancreatic head which broadens differential diagnosis of pancreatic lesions. Although resection of this lesion seems curative, there is a need for careful follow-up and exclusion of more controversial and aggressive multicentric forms, including the possibility of neoplastic progression, especially lymphomatous transformation. Complete resection of the mass was curative.

Contributors

MV, SD and KZ performed surgery of the patient and proposed the study, MM did the histopathological analysis; SD was involved in diagnostic and imaging part, KS collected and analyzed the data. All authors contributed to the design and interpretation of the study and to further drafts. KZ is the guarantor.

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None.

Ethical approval

This study was approved by the Hospital Ethics Committee. Consent was obtained from the patient for publication of this report and any accompanying images.

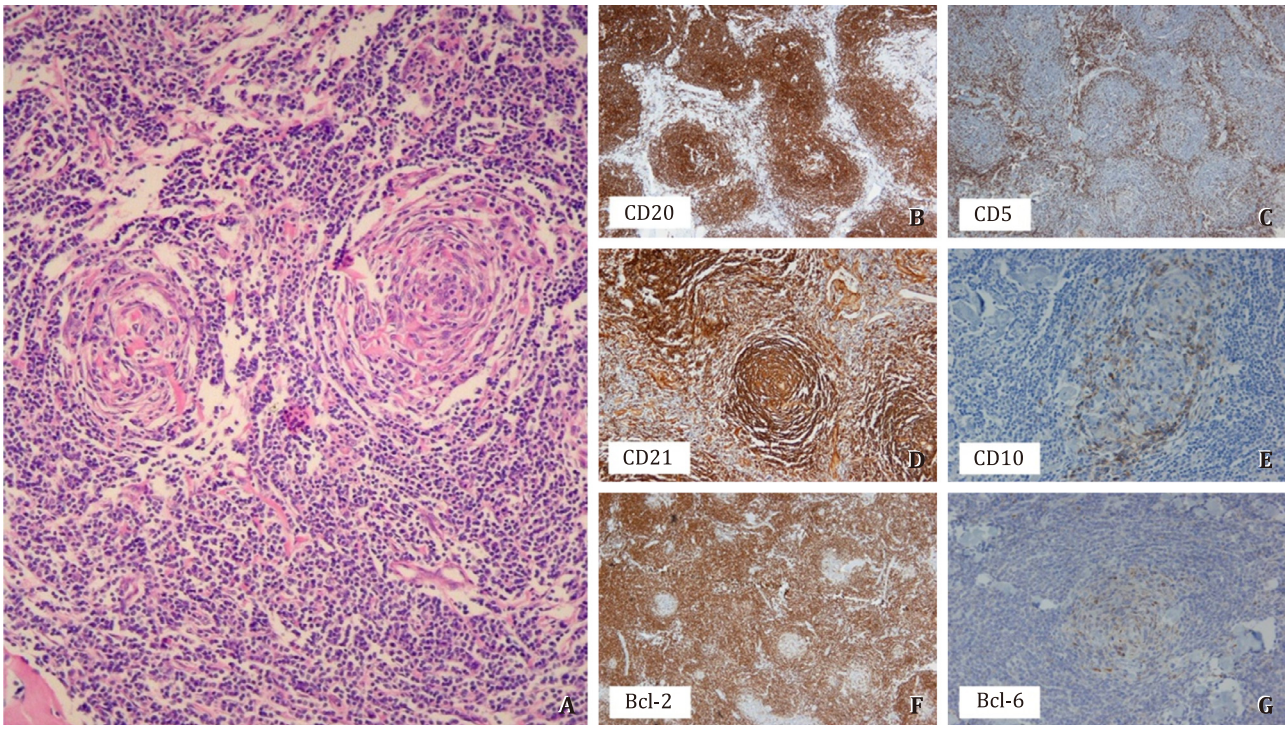

Fig. 3. Distinctive hyalino-vascular lesion within lymphoid nodules which, although not pathognomonic, presents the hallmark of this type of the disease (**A**, HE staining, original magnification × 20) with immunostaining characterizing (**B-G**).

Competing interest

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

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