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A rare case of Castleman disese mimicking a pancreatic tumor

Castleman's disease or angiofollicular lymph node hyperplasia is a rare clinical entity, and may involve any parts of the body. Unicentric presentation of the disease is the most common presentation. Castleman's disease of the pancreas that mimics a pancreatic neoplasm is more uncommon. We present a 38-year-old female patient with a one-month past history of abdominal pain. Imaging studies revealed hypervascular, 4 x 2 cm in size nodular mass localised in the pancreatic body. In FDG PET/CT, the pancreatic mass was FDG-avid. Laparoscopic enucleation of the lesion was performed. Histopathological study revealed unicentric form of Castleman's disease, a hyaline vascular variant. Since these lesions are rare and resembling malignant tumors on computed tomography and angiography, we discuss the problems of diagnosing Castleman's disease, together with the literature.

KEY WORDS: Castleman's disease, Lymphoid Follicular hyperplasia, Lymphoproliferative disorder, Peripancreatic tumor

Introduction

Angiofollicular lymph node hyperplasia, in other words, Castleman's disease (CD) was first described by Benjamin Castleman, as a mediastinal disease, in1956 [1]. Pancreatic CD is rarely seen with only a few reports in the literature, diagnosis is confusing because of resembling to pancreatic tumor [2]. In this case, the lesion was very close to the body of the pancreas, and laparoscopic resection of the mass was performed. Pathological examination revealed unicentric hyaline-vascular (HV) variant of CD.

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Case Report

A 38-year-old woman presented with abdominal pain. This pain had first started 1 month ago before admitted to our hospital. She had no jaundice, weight loss, change in appetite or general malaise. There is no disease in her past medical history. Physical examination of the patient and laboratory studies were normal. Abdominal ultrasound demonstrated a well circumscribed mass measuring 4,5x4x1,8 cm in size nodular mass localised very close to the anterio-superior border of pancreatic body (Fig. 1).
Tumor associated antigen levels (CEA, CA 19-9, and CA-125) were normal. Abdominopelvic CT scan showed a hypervascular, 4x2 cm in size nodular mass with cleavage point with pancreas. In FDG PET/CT, the pancreatic mass was FDG-avid (SUV-max 6.1). Enucleation of the mass was performed laparoscopically (Fig. 2).
Pathological findings were: The external surface of the mass was beige/tan in color, nodular shape, firm apperarance. Cut sections of the mass also revealed a few submilimetric sized, black, cystic components. According to lymph node microscopic examination; folicular patern was lost. Regression, atrophy and mantle zone dilatation

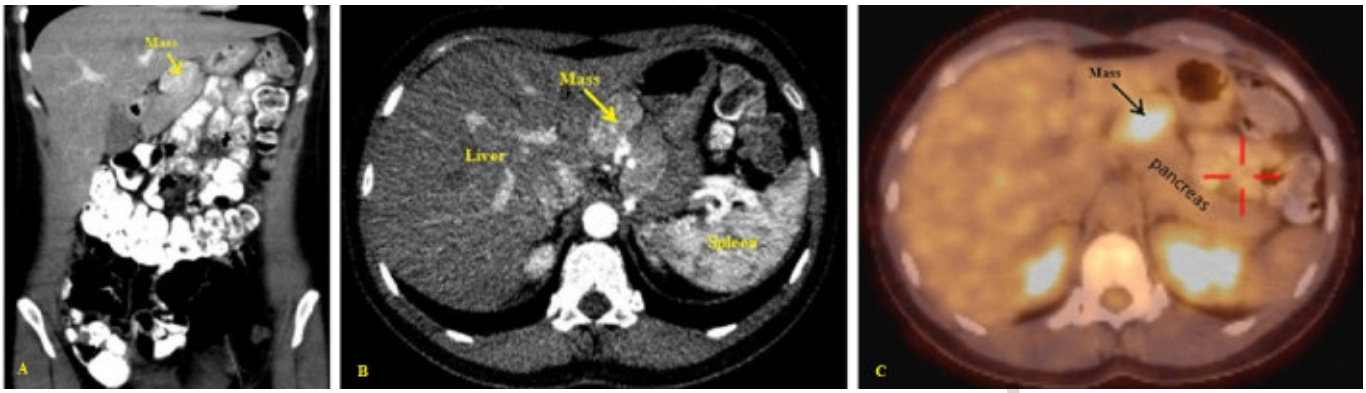

Fig. 1: Preoperative imaging of Castleman's disease. Coronal (A) and axial (B) images in abdomen CT shows well-defined mass and consistent with Castleman's disease (arrow). FDG PET-CT releaved increased FDG uptake (SUV-max:6.1) located exophytically in the antero-superior border of pancreas body (arrow).

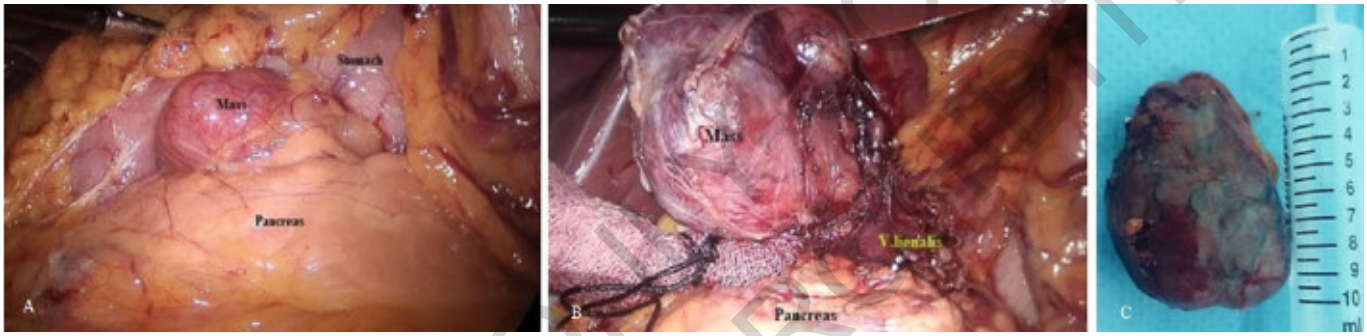

Fig. 2: Pictures during laparoscopy (A) and (B) showing the mass and postoperative appearance of mass (C) (4,5x4x1.8 cm).

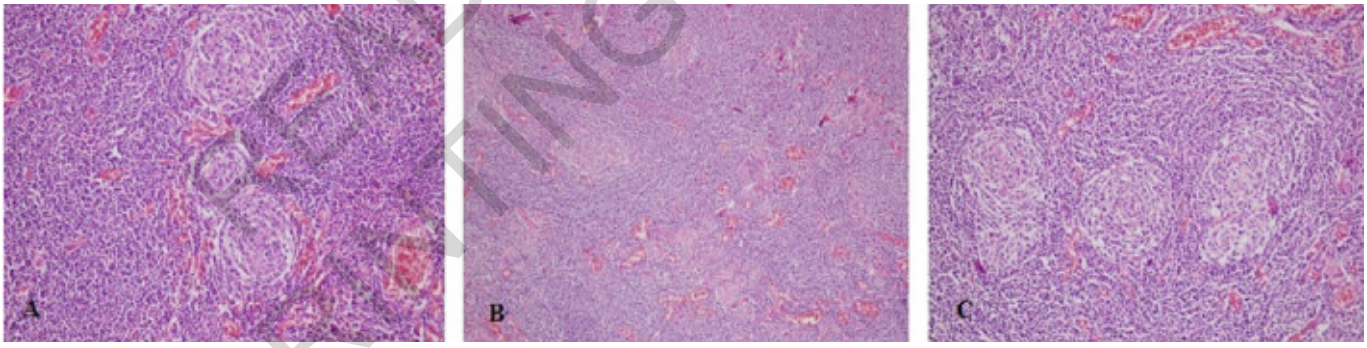

Fig. 3: Histolopathological examination of mass (H&Ex40) A) duplicated germinal centers and regressed follicles B) hyalinised foci in stroma C) concentric layering of mantle zone cells.

were seen in germinal centers of the folicules. There are tendency of fusion of regressed folicules and collagenised-hyalinised band formations. And these features are consistent with localized hyaline-vascular type of Castleman's disease (Fig. 3).

Castleman's disease mimics a number of diseases such as mantle cell lymphoma, so immunohistochemistry must rule out malignancies. In our case, CD 3, CD 5, CD 20, CD 21, CD 23, CD 68 were positive, CD 123 was negative, HHV-8 was negative, Kappa, Lambda, S-100 were rarely positive.

These morphologic and immunohistochemical findings are most consistent with a diagnosis of hyaline-vascular type of Castleman's disease.

Discussion

Castleman's disease is one of the rarest lymphoproliferative disorders. The main clinical manifestation of unicentric Castleman's disease (UCD) is a single lymph node enlargement with few symptoms or no pain, so CD is diagnosed by imaging modalities. On the other hand, multicentric CD affects multiple lymph nodes, so systemic symptoms like fatigue, fever, weight loss and night sweats are seen more often. Mediastinum was the most common lymph node site (70%) and the neck and abdomen the least common (15%) sites for CD was reported by Van Rhee et al [3]. For pancreatic UCD there are no distinctive radiologic features and this raises concerns for pancreatic tumors, requiring pathologic evaluation. Histologically, CD is classified into 3 variants: The hyaline vascular (HV-CD), plasma cell (PC-CD) and mixed type (MT-CD) variants. HV-CD (80%-90% of cases) is the most common clinical variant with follicular hyperplasia, marked vascular proliferation and regressed germinal centers as in our case. PC-CD is characterized by Russell bodies, larger lympho-reticular nodules and fewer hyalinized blood vessels. MT-CD is the rarest variant [4,5]. The case presented here a hyaline-vascular type of CD. That can be treated with surgery, steroids, radiotherapy and immunotherapy or by a combination. Treatment and prognosis depend on the form of CD. To Chen at all total resection is the most common curative treatment option for the UCD and prognosis after total surgical resection is excellent [6].

Conclusion

Due to unclear etiology and pathogenesis, and clinical manifestations, diagnosis and treatment of CD have not been standardised. Differential diagnosis is difficult and very important because of the clinical complexity of the disease, a clinical and pathological correlation is necessary for definitive diagnosis. As a result, surgery may be the best therapeutic alternative for CD.

Riassunto

La malattia di Castleman o iperplasia angiofollicolare dei linfonodi è un'entità clinica rara e può coinvolgere qualsiasi parte del corpo. La presentazione a localizzazione unica della malattia è l'osservazione clinica più comune. La malattia di Castleman del pancreas che imita una neoplasia pancreatica è più rara. Presentiamo una paziente di 38 anni con una storia pregressa di dolore addominale di un mese. Gli studi di imaging hanno rivelato massa nodulare ipervascolare di dimensioni 4 x 2 cm localizzata nel corpo pancreatico. In FDG PET / CT, la massa pancreatica captava avidamente il FDG. Sull'enucleazione laparoscopica della lesione, lo studio istopatologico ha portato alla diagnosi di una forma unicentrica della malattia di Castleman, variante vascolare ialina. Poiché queste lesioni sono rare e assomigliano a tumori maligni nella tomografia computerizzata e nell'angiografia, vengono discussi i problemi diagnostici della malattia di Castleman alla luce della letteratura.

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