Introduction

Castleman’s disease, also known as angiofollicular or giant lymph node hyperplasia, is a rare lymphoproliferative disorder that results in the unregulated growth of lymphoid tissue in a localized or multicentric form. Initially, Castleman’s disease was reported as an indolent disorder which was usually confined to a single lymph node group (1, 2). However, recent case reports have described a multicentric form of Castleman’s that often manifests a more malignant clinical course (3). Commonly, it is a unique tumour localized in the mediastinum (unicentric disease) (4). Extrathoracic localizations are however reported with an increasing frequency. The clinical presentation varies widely, but most commonly manifests as a solitary mediastinal mass, incidentally found on radiographic examination. It is classified histologically as two histopathological variants (hyaline vascular and plasma cell) and clinically two types (localized and multicentric) (4). Recently, a mixed type of plasma cell and hyaline vascular type has also been reported, but has rarely been found. The more common hyaline vascular variant has typically a benign clinical course with no constitutional symptoms other than localized pressure from the mass which is usually localized to the mediastinum or pulmonary hilum. The plasma cell pattern involves lymph nodes separately or in aggregations and often displays multicentricity with systemic symptoms including auto-immune phenomena and an aggressive course. Surgical removal of a unicentric mass of hyaline-vascular or hyaline-vascular/plasma cell type is curative (5). Patients with multicentric disease, either hyaline-vascular or plasma cell type, do not benefit from surgical management and should be candidates for multimodality therapy, the nature of which has yet to be defined. This case report concerns an unusual case of Castleman’s disease mimicking a pancreatic tumour with a rapidly fatal outcome.

Case report

A 56-year-old hypertensive woman was admitted to our hospital because of fatigue, weight loss and vague abdominal pain of one-month duration. The patient had no significant prior illnesses. On admission, physical examination and laboratory findings, including tumour markers and inflammatory mediators, revealed nothing in particular. Abdominal ultrasound demonstrated a well-defined isolated, 3.7 × 1.8 cm, hypoechoic mass in the pancreatic head. This appearance mimicked the mass lesion, which was isodense with the pancreatic paranchyma. The pancreatic duct was not dilated. No enlarged abdominal or retroperitoneal lymph nodes were detected (Fig. 1).

The initial diagnosis was pancreatic cancer and the case underwent surgical excision. Exploratory laparotomy revealed a smooth, solid mass, 3 cm in diameter, located in the pancreatic body. A Whipple procedure was performed. Gross examination showed a tumour 2.5 × 2.2 cm in size without adhesion to the surrounding tissue. Histopathologically, the tissue specimen revealed an angiofollicular lymph node hyperplasia (Castleman’s disease) of the hyaline vascular type (Figs. 2 and 3). The patient had an uneventful early postoperative course and was discharged 14 days postoperatively. Her symptoms relieved and healed postoperatively, but unfortunately she did not attend her follow-up appointments after the third month.
Pancreatic Castelman’s Tumour

Discussion

Castleman’s disease is an unusual disorder of immune regulation that results in abnormal proliferation of B-lymphocytes and plasma cells in lymphoid organs (6). It was first described in 1954 by CASTLEMAN et al. as giant lymph node hyperplasia (1, 2). KELLER et al. described two histological variants: the hyaline vascular type, which is the most common variant (90%) and the less common plasma cell variant (10%) (4). A mixed type of plasma cell and hyaline vascular Castleman’s disease has also been reported (7). The hyaline vascular variant is described histologically as having the onion-skin appearance of small lymphocytes circumferentially surrounding lymph node follicles (8).

Clinically, there are two distinctive types in Castleman disease of the abdomen: localized and multicentric. Patients with multicentric Castleman’s disease are generally older (8) and characterised by the appearance of multiple, multifocal adenopathies combined with systemic symptoms associated with hepatosplenomegaly (3). The localized Castleman’s disease, defined as a single, benign lesion, usually affects young people. The course of the multicentric disease is often fatal, caused by infectious complications or development of malignancies.

Seventy percent of cases present in the mediastinum and 20% occur in the axillary, cervical, inguinal and vulvar regions while approximately 12% occur in the abdomen; mostly located in the pelvis, mesentery and perinephric regions (4). No age or gender predominance has been evident (7). Two cases of retroperitoneal Castleman’s disease were reported in 1967 emphasizing Castleman’s disease in the differential diagnosis of retroperitoneal tumours (9). Presently, only seven cases of Castleman’s disease presenting as a pancreatic mass have been reported in the literature (3, 5, 10, 11, 12, 13, 14).

Patients with the hyaline vascular type are often asymptomatic. Those with the plasma cell type may have fever, night sweats, fatigue, anemia, hypergamma-globulinemia, hypo-albuminemia, elevated sedimentation rate and bone marrow plasmocytosis (4, 15). In the present patient, fatigue, weight loss and vague abdominal pain were all uncharacteristic symptoms.

Imaging techniques are commonly inconclusive for Castleman’s disease. Findings of ultrasonography and computed tomography revealing an isolated, hypoechoic mass or a fleshy structure adjacent to the pancreas with moderate enlargement of the intrahepatic bile ducts,
are not specific for the disease (5, 15). In our case, ultrasound and computed tomography seemed to be helpful in providing information about the presence of the tumour. Nevertheless, imaging procedures like dynamic computed tomographic scan, magnetic resonance imaging or angiography should be considered for the differential diagnosis of a pancreatic mass.

Recently, definitive diagnosis has been based on the postoperative pathological findings. The adhesion of the tumour to the surrounding tissue and hypervascularity in the mass are characteristic features of the hyaline vascular type (16, 17).

Our case revealed no specific finding to emphasize a suspicion for a pre-operative diagnosis for peripancreatic Castleman’s disease. The patient was indistinguishable radiographically and operatively.

**Conclusion**

Although it is rare, pancreatic Castleman’s disease should remain a consideration in the differential diagnosis of a pancreatic mass, as it is indistinguishable from a malignant tumour, radiographically and operatively. Therefore, imaging procedures like dynamic computed tomographic scan, magnetic resonance imaging or angiography, which can evaluate the hemodynamic aspects of the tumour, can be helpful in the diagnosis of the pancreatic Castleman’s tumour.

**References**