Introduction

Benjamin Castelman in 1954 described for the first time an asymptomatic benign hyperplastic lymph node resembling a thymoma (1). This condition, known presently as Castelman’s disease, has many synonyms including giant lymph node hyperplasia, lymph node hamartoma, angiofollicular mediastinal lymph node hyperplasia and angiomatous lymphoid hyperplasia (2).

Most of the lesions are located in the thorax but extrathoracic involvement, including neck, axilla, mesentery and retroperitoneum, has also been described. We report a case of a 69 years old men with peripancreatic localisation, mimicking a pancreatic neoplasm treated with local excision. Only seven cases of pancreatic and peripancreatic localisation are described in the world-wide literature. This particular site of disease may give troublesome differential diagnosis due to non specific clinical signs and radiological features. Often surgical excision is both diagnostic and therapeutic.

Case report

A 69 years old patient with a brief history of left hypochondrium pain and fever was admitted to our hospital for further investigations. His past medical history showed type II diabetes mellitus and hypertension.

His physical examination showed abdomen soft, with a slight tenderness in the left hypochondrium on deep palpation and a palpable spleen due to splenomegaly. Laboratory tests were normal except for a mild hypobuminemia (2,5 g/dL) and hyperglycemia (270 g/dL). There was no serological indication of active HIV, B and C hepatitis, and tumoral markers as CA.19-9 and CEA were normal.

Upper gastrointestinal endoscopy with biopsy showed mild ch...
Chronic gastritis. A CT scan showed, posteriorly to the antro-piloric region near the trifurcation of the celiac axis and very close to the hepatic and gastroduodenal arteries, a 4 cm mass with a necrotic centre and a significative contrast enhancement (Fig. 1). This image had an equivocal interpretation, and was suspected to be a pancreatic mass. Multiple lymph node (maximum diameter 1 cm) were detected around the celiac axis and the aorta. The patient underwent endoscopic ultrasonography which showed near the celiac axis multiple anecogenic nodules, varying from 5 to 15 mm, compatible with lymph nodes, and a voluminous hypocogenic mass of the body of the pancreas with diffusely dishomogeneous parenchyma and faded margins compatible with neoplasm (Fig. 2). An endoscopic ultrasound-guided fine needle aspiration of the pancreatic mass showed bundles of small and medium-large sized lymphoid cells and some rare atypical cells with an altered nuclear-cytoplasm ratio; this result could not exclude a neoplastic process.

The patient underwent surgical operation for suspected pancreatic neoplasm. Operative exploration showed a well vascularized 4 cm mass on the superior edge of the body of the pancreas. The mass was soft and in direct contact with the hepatic and the splenic arteries. It was possible to dissect the mass completely from the pancreas and from celiac branches. Macroscopic examination showed a white-greyish 4x2.5x2.5 cm capsulated mass with hemorrhagic spots. Frozen section showed an atypical lymphoproliferative process and the final pathological report described an angiofollicular lymphoid hyperplasia compatible with mixed variety, hyaline-vascular plasma cell type of Castelman’s disease made of preserved lymphoid tissue with follicles at various degrees of maturation and diffuse hyaline involution and intervening sheets of plasma cells and capillary. Immunohistchemistry showed regular expression of CD3 in the follicles and CD79A in the para-cortical region, with a k/λ ratio monotypic for λ on plasma cells.

Postoperative course was uneventful and the patient was discharged on 14th postoperative day. He is alive and free of recurrence at one year follow-up.

Discussion

Castelman’s disease usually presents in young adult with median age of approximately 35 years, equally distributed between male and female (5). Little is known about the cause of this disorder. Most of the evidence point out a faulty immunoregulation which results in the excessive proliferation of B lymphocytes and plasma cells in lymphoid organs (6).

Keller et al. distinguished two basic histological types (hyaline-vascular and plasma cell types) and one mixed variant. The hyaline-vascular type, which is the most frequent (about 90% of all cases), is characterised by small hyaline-vascular follicles and interfollicular capillary proliferation. The plasma cell type is characterised by large follicles with intervening sheets of plasma cells (5). Clinically the former is almost always asymptomatic and may manifest itself only because of mass compression symptoms, while the latter is sometimes associated with systemic manifestation such as fever, anaemia, weight loss, night sweats, and polyclonal hypergammaglobulinemia (7).

Castelman’s disease is currently classified in two clinico-pathological groups: localised disease, with a typical benign course, and disseminated disease which has usually a malignant course (5, 7, 8). The multicentric variety, in fact, is frequently associated with AIDS, Kaposi sarcoma and the so-called POEMS syndrome which consists in polineuropathy, organomegaly, endocrinopathy, M protein and skin changes (6).

Peripancreatic localization of Castelman’s disease is an uncommon location. In a review of 315 cases, 70% occurred in the mediastinum, 20% in cervical, axillary, shoulder, inguinal or vulva areas, and 7% (21 cases) in retroperitoneum (9). To the best of our knowledge only seven cases of this disease originating from the pancreas and the peripancreatic tissue are reported in literature (10).

Castelman’s disease has no specific radiological fea-
tures, in fact even if ultrasonography, computerized to-

mography, magnetic resonance imaging have been pro-

ven to be helpful in diagnosing retroperitoneal tumour,
it is almost always indistinguishable from other disease.

The ultrasonographic picture, a uniform hypoe-

choic mass with enanched through transmission, is in

fact similar to that seen in lymphoma (11). Endosco-

pic ultrasonography is an excellent tool for characteri-

sing lesions of the pancreas due to its superior spatial

resolution, however the features of neoplastic and

non-neoplastic pancreatic lesions and those with and

without malignant potential overlap and interobserver

agreement is poor (12). Endoscopic ultrasonography

in our case has been used to study the peripancreatic

lesion and to perform fine needle biopsy; this kind of

approach is reported only by Goetze e coll. and their

finding - a well defined and hypocogentic mass - was

consistently similar to our finding (10).

Computed tomography examination shows solid

density and homogeneously enhanced masses with or

without calcification. The usual appearance of this le-

son on computed tomography is that of a non speci-

fic homogeneous mass which ranges from hypo to iso-
dense in relation to the liver on non-contrasted scan.

Dense enhancement immediately after the infusion of

iodinated material is observed (13). Castelman's disea-

se in the abdomen and in the pelvis may display a va-

riety of calcification patterns (punctuate, coarse, pe-


eripheral "arborizing") (10). In a recently published re-

trospective review of 16 cases of abdominal and pelvic

Castelman's disease localizations, 31% showed calcifi-
cation, one with an arborizing pattern and the remai-

ning lesions with punctuate calcifications (14). Hence,

possible differential diagnosis of this indeterminate tu-
mour containing calcification is with acinar cell carci-
noma, solid and epithelial neoplasm, solid-cystic tu-
mour, serous cystadenoma or cystic teratoma of the pan-

creas, which are all known as well defined tumours

with calcification patterns (15, 16).

Generally, the tumours of Castelman's disease are
described nearly isointense to muscle in signal inten-
sity on T1-weighted magnetic resonance imaging,

with heterogeneous signal characteristics within the

mass on T2-weighted imaging (17). The radiological
differential diagnosis includes various neoplastic, in-

flammatory (tubercolosis or sarcoidosis) and other mi-

sellaneous retroperitoneal diseases, so that often a

biopsy is the only way to have a definitive diagnosis.

In a retrospective study by Bowne e coll. surgical ex-
cision has been associated with the best chance of cure

for localised disease (8). Even partial excision of unre-
sectable lesions may be useful since recurrence or pro-
gression has not been reported (5, 18). In our case the

possibility to have a complete excision of the mass was

associated with no recurrence after one year follow-up.

For multicentric disease the treatment has not been

established due to its variable clinical course ranging

from chronic evolution with remission and exacerbation

requiring continuos steroids or chemotherapy or both,
to rapidly progressive and fatal course (6, 7).

In conclusion we can state that Castelman's disease is

a pathologic entity which, even if rarely, may involve

pancreatic and peripancreatic tissue and it is difficult to

make a differential diagnosis. Clinicians should therefo-

re be aware of this disease in which often surgical exci-
sion is both diagnostic and therapeutic.

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