G Chir Vol. 29 - n. 4 - pp. 141-144
Aprile 2008

articoli originali

Unicentric Castelman's disease in peripancreatic tissue: case report and review of the literature

F. TALARICO, L. NEGRI, D. IUSCO, G.G. CORAZZA

SUMMARY: **Unicentric Castelman's disease in peripancreatic tissue: case report and review of the literature.**

F. TALARICO, L. NEGRI, D. IUSCO, G.G. CORAZZA

Castelman's disease is a rare disorder of the lymphoid tissue that usually occurs in the mediastinum even if 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.

RIASSUNTO: **Malattia di Castelman unicentrica nel tessuto peripancreatico: case report e revisione della letteratura.**

F. TALARICO, L. NEGRI, D. IUSCO, G.G. CORAZZA

La malattia di Castelman è un disordine linfoproliferativo che si osserva più frequentemente nel mediastino, anche se sono state descritte localizzazioni extratoraciche cervicali, ascellari, mesenteriche e retroperitoneali.

Descriviamo il caso di un uomo di 69 anni con una localizzazione peripancreatica della malattia che mimava la presenza di una neoplasia e che fu trattata con escissione locale.

Solo sette casi di localizzazione pancreatica e peripancreatica sono descritti in letteratura. In questi casi la diagnosi differenziale può essere problematica sia per i segni clinici aspecifici che per le caratteristiche radiologiche non dirimenti. L'escissione chirurgica è spesso sia diagnostica che terapeutica.

KEY WORDS: Pancreas - Giant lymph node hyperplasia - Castelman's disease.
Pancreas - Iperplasia linfonodale gigante - Malattia di Castelman.

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 involvment, including neck, axilla, mesentery and retroperitoneum, has also been reported (3). When the disease is localised in the retroperitoneum usually it has no distinctive clinical or radiological features, making it difficult to distinguish from other retroperitoneal tumours (4).

We describe a case of Castelman's disease in peripancratic tissue and review the main question concerning this rare disease.

Case report

A 69 years old patient with a brief history of left hypocondrium 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 hypocondrium on deep palpation and a palpable spleen due to splenomegaly. Laboratory tests were normal except for a mild hypoalbuminemia (2,5 g/dL) and hyperglicemia (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-

Azienda Unità Sanitaria Locale, Bologna
Ospedale "Santissimo Salvatore", San Giovanni in Persiceto (BO)
Dipartimento Chirurgico, Unità Operativa Chirurgia Generale
(Direttore: Dott. F. Talarico)

© Copyright 2008, CIC Edizioni Internazionali, Roma

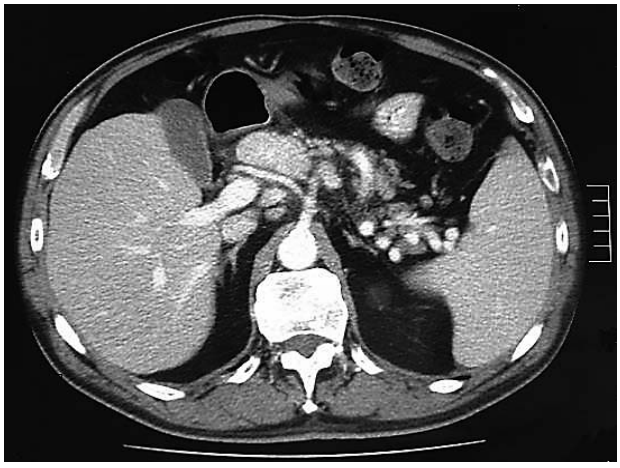

Fig. 1 - Computed tomography scan showing a 4 cm mass with a necrotic centre and a significative contrast enhancement near the trifurcation of the celiac axis and very close to the hepatic artery.

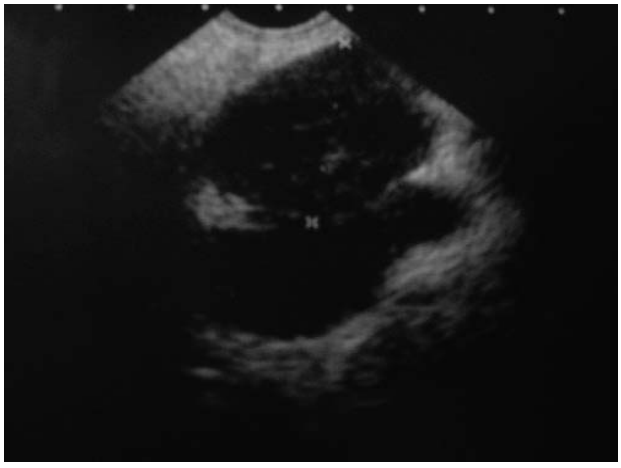

Fig. 2 - Endoscopic ultrasonography which showed a voluminous hypoecogenic mass in the body of the pancreas with diffusely dishomogeneous parenchyma and faded margins.

ronic gastritis. A CT scan showed, posteriously 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 hypoecogenic 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 alterated 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 hemoragic 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. Immunochemestry 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 plasmacells in lymphoid organs (6).

Keller et al. distinguished two basic hystological 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 intefollicular 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 tomograpy, magnetic resonance imaging have been proven to be helpful in diagnosing retroperitoneal tumour, it is almost always indistinguishable from other disease.

The ultrasonographic picture, a uniform hypoechoic mass with enanched through transmission, is in fact similar to that seen in lymphoma (11). Endoscopic ultrasonography is an excellent tool for characterising 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 hypoecogenic 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 lesion on computed tomography is that of a non specific homogeneous mass which ranges from hypo to isodense in relation to the liver on non-contrasted scan. Dense enhancement immediately after the infusion of iodinated material is observed (13). Castelman's disease in the abdomen and in the pelvis may display a variety of calcification patterns (punctuate, coarse, peripheral "arborizing") (10). In a recently published retrospective review of 16 cases of abdominal and pelvic Castelman's disease localizations, 31% showed calcification, one with an arborizing pattern and the remaining lesions with punctuate calcifications (14). Hence, possible differential diagnosis of this indeterminate tumour containing calcification is with acinar cell carcinoma, solid and epithelial neoplasm, solid-cystic tumour, serous cystoadenoma or cystic teratoma of the pancreas, 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 intensity 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, inflammatory (tubercolosis or sarcoidosis) and other miscellaneous 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 excision has been associated with the best chance of cure for localised disease (8). Even partial excision of unresectable lesions may be useful since recurrence or progression 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 therefore be aware of this disease in which often surgical excision is both diagnostic and therapeutic.

References

1. Castelman B, Towne VW. Case reports of Massachusetts General Hospital: weekly clinicopathological exercises: founded by Richard C. Cabot. Case 40011. N Engl J Med 1954;250:26-30.
2. Walter JF, Rottemberg RW, Cannon Wb, Sheridan LA, Pizzimenti J, Orr JT. Giant mediastinal lymph node hyperplasia (Castelman's disease):angiographic and clinical features. Am J Roentgenol 1978;130:447-50.
3. Frizzera G. Castelman's disease and related disorders. Sem Diagn Pathol 1988;5:346-364.
4. Sadamoto Y, Abe Y, Higuchi K, Kato K, Matsumoto S, Arima N, Nawata H Retroperitoneal Castelman's disease of the hyaline vascular type presenting arborizing calcification. Internal Medicine 1998;37(8):691-93.
5. Keller AL, Hochholzer L, Castelman B. Hyaline-vascular and plasma-cell types of giant lymph node hyperplasia of the mediastinum and other locations. Cancer 1972; 29:670-83.
6. Peterson Ba, Frizzera G. Multicentric Castelman's disease. Sem Oncol 1993; 20:636-47.
7. Herrada J, Cabanillas F, Rice L, Manning J, Pugh W. The clinical behavior of localized and multicentric Castelman disease. Ann Intern Med 1998;128:657-62.
8. Bowne WB, Lewis JJ, Filippa DA, Niesvizky R, Brooks AD, Burt ME , Brennan MF The managment of unicentric and multicentric Castelman disease: a report of 16 cases and a review of the litterature. Cancer 1999; 85:706-17.
9. Testas P, Pigne A, Voinesson A, Vieillefond A, Paillas J. Angiofollicular lymphoid hyperplasia (Castelman's disease). First case of meso-sigmoid localization (author's transl). Chirurgie 1980;106:156-60.
10. Goetze O, Banasch M, Junker K, Schmidt WE, Szymanski C Unicentric Castelman's disease of the pancreas with massive central calcification. World J Gastroenterol 2005:11(42):6725-27..
11. Barki Y, Shadked G, Levy I. Mesenteric Castelman disease: sonographic diagnosis. J Clin Ultrasound 1992;20:486-88.
12. Ahmad NA, Kochman ML, Brensinger C, Brugge WR, Faigel DO, Gress FG, Kimmey MB, Nickl NJ, Savides TJ, Wallace

MB, Wiersema MJ Ginsberg GG. Interobserver agreement among endosonographers for the diagnosis of neoplastic versus non-neoplastic pancreatic cystic lesions. Gastrointest Endosc 2003; 58:59-64.

13. Johkoh T, Muller NL, Ichikado K, Nishimoto N, Yoshizaki K, Honda O et al Intrathoracic multicentric Castelman disease: CT finding in 12 patients. Radiology 1998;209:477-81.
14. Meador Tl, McLarney JK. CT features of Castelman's disease of the abdomen and the pelvis. AJR Am J Roentgenol 2000:175:115-18.
15. Nakamura S,Okayama Y, Imai H, Aoki S, Kobayashi S, Hattori T, Shiraki S, Goto K, Sano H, Ohara H, Nomura T, Joh T, Yoshifumi Y, Itoh M. A solid cystic tumor of the pancreas with ossification and possible malignancy, coexisting nonfusion of the pancreatic ducts. J Clin Gastroenterol 2001;33:333-36.
16. Chiou YY, Chiang JH, Hwang JI, Yen CH, Tsay SH, Chang CY. Acinar cell carcinoma of the pancreas:clinical and computed tomography manifestations. J Comput Assist Tomogr 2004;28:180-86.
17. Debatin JF, Spritzer CE, Dunnick NR. Castelman disease of adrenal gland. MR imaging features. AJR Am J Roentgenol 1991;157:781-83.
18. Skolnik G, Wiklund LM, Risberg B. Castelman's tumour with retroperitoneal locations: a malignant-appearing benign tumor. J Surg Oncol 1985;28:153-55.