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CASE REPORT
ΕΝΔΙΑΦΕΡΟΥΣΑ ΠΕΡΙΠΤΩΣΗ

Castleman disease mimicking pancreatic neoplasia

Castleman disease is a difficult diagnosis, even based on histopathological and immunohistochemical data from samples of the lesions. We report a case of a previously healthy male patient at the age of 57 years, who underwent the evaluation to clear the origin of a hypervascular mass with calcifications in the pancreatic area. Initial challenging interpretation of routine laboratory determinations and imaging studies, besides the histopathological and immunohistochemical data are commented on. The final diagnosis of Castleman disease was established after the procedure of corporal-caudal pancreatectomy associated with splenectomy performed by laparoscopy. Even single cases may enhance the awareness and suspicion index among non-specialist health care workers about the Castleman disease mimicking a primary neuroendocrine tumor.

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2025, 42(4):569–572

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Νόσος του Castleman μιμούμενη νεόπλασμα παγκρέατος

Περίληψη στο τέλος του άρθρου

Key words

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Castleman disease (CD) is a benign lymphoproliferative entity that evolves unicentric (single lymph node or nodal region) or multicentric (in multiple lymph node sites); although exceeding uncommon, the pancreatic location can mimic a neuroendocrine tumor (NET).[1–10] Multicentric CD can be idiopathic, or related to herpes virus-8 and HIV, or polyneuropathy, organomegaly, endocrinopathy, monoclonal plasma cell disorder, and skin changes (POEMS). Multicentric cases can evolve to malignant lymphoma, plasmacytoma or Kaposi's sarcoma.[5] CD more often evolves asymptomatic, and may be incidentally diagnosed affecting lymphoid tissues of the mediastinum (70%), neck (15%), abdomen and pelvis (12%), and axilla (3%).[4] The main differential diagnoses are thymoma, lymphoma, paraganglioma, adenocarcinoma, tuberculosis, HIV-related lymphadenopathy, autoimmune diseases, mycoses, and NETs.[1–10] The diagnostic clues include Bcl-2, Bcl-6, CD3, CD5, CD10, CD20, CD31, CD35, and CD138. Resection is the first choice for unicentric CD, with over than 90% relapse-free survival; chemo- and radiotherapy are options when the mass cannot be removed, and for multicentric CD including steroids due to higher recurrence rates and the risk of malignant transformation.[1–10] The objective of this report is to highlight a very uncommon presentation of abdominal CD involving the pancreas body and was treated by laparoscopic pancreatectomy and splenectomy.

CASE PRESENTATION

A 57-year-old previously healthy male patient sought the oncological surgery outpatient clinic because of a low back pain lasting 10 months, without weight loss or fever. On his physical examination, there was no peripheral lymph node enlargement or palpable abdominal masses. The routine laboratory determinations and endoscopic evaluations revealed no abnormalities. The abdominal computed tomography (CT) detected an expansive exophytic lesion (6.1×4.1 cm) in the posterior and inferior areas of the pancreatic body, which was hypervascular and containing coarse central calcifications, compatible with a primary tumor (fig. 1). Due to the hypervascularity, a positron emission tomography (PET) PET/CT with Gallium 68 (^{68}Ga-DOTATATE) was performed and showed the pancreatic mass with a maximum SUV of 13.94, and absence of distant lesions. The markers CA 19.9, CA 125, carcinoembryonic

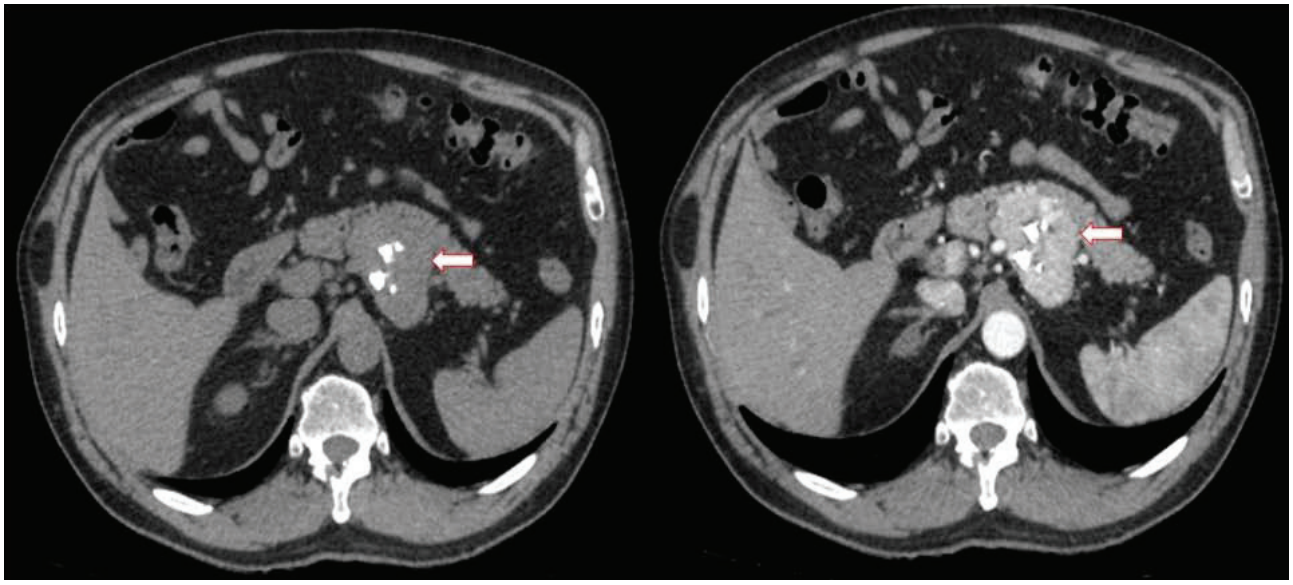

Figure 1. Axial views of the abdominal computed tomography (CT) images showing the hypervascular calcified lesion located in the body and tail of the pancreas (arrows).

antigen, and chromogranin A were normal. The multidisciplinary team evaluation raised the initial hypothesis of a PNET based on the characteristics revealed by imaging studies, and the prompt surgical management was indicated. Therefore, the corporal-caudal pancreatectomy (fig. 2) associated with splenectomy was performed via laparoscopy with success, and the patient had an uneventful postoperative course, being discharged asymptomatic from hospital on the fifth day. The immunohistochemistry panel was consistent with pancreatic CD showing positive CD3, CD10, CD15, CD20, CD23, CD30, CD138, Bcl-2, and Bcl-6 markers. With this confirmation, serological tests were carried out for HIV in addition to herpes virus-8, which resulted negative. The patient did not undergo any systemic therapy, and persisted without neither abdominal nor systemic signs or symptoms.

DISCUSSION

CD was first described in 1954 by Benjamin Castleman et al,[5,7,8,10] as a mediastinal lymph node hyperplasia resembling thymoma. The estimated incidence is 21 cases per one million population, mainly females at mean age of 35 years, and up to 12% are retroperitoneal.[7] The unicentric pancreatic CD here described was incidentally diagnosed in a healthy 57-year-old male, with the classical imaging features of a hypervascular calcified mass. Bhatia et al[1] reported a 26-year-old woman with abdominal pain and weight loss, who had a mass with calcification and arborising vessels in pancreatic body. The endosonographic ultrasound-based fine needle aspiration cytology yielded the diagnosis, and due to insufficient response to chemotherapy the mass was excised by laparotomy. The authors emphasized the utilization of fine needle aspiration cytology to establish the pre-operative diagnosis of CD.[1] Dev et al[2] described a 46-year-old diabetic woman with hypothyroidism, who had incidental evidence of a mass in the pancreatic neck and head, and surgical samples revealed a hyaline vascular CD.[2] The authors highlighted that if the histopathologic diagnosis NET is not previously confirmed, one must consider the pancreatic CD among the differential diagnoses of a suspected lesion.[2] Gunda et al[3] reported a 58-year-old female with incidental finding of a non-calcified mass at the posterior body of the pancreas presenting arterial enhancing, highly suspected of a NET. The patient underwent a distal pancreatectomy and splenectomy and the diagnosis was CD with positivity for CD3, CD20, CD21, CD23, and negative for Bcl-2, CD31, and herpes virus-8.[3] Huang et al[4] described a 44-year-old man with an incidentally found abdominal mass at the pancreatic head that was withdraw by laparoscopy; the CD3, CD5, CD20, CD23, Bcl-

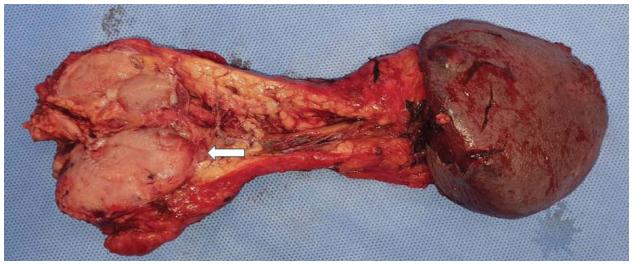

Figure 2. Aspect of the surgical specimen of the corpus-caudal pancreatectomy besides splenectomy performed to treat the pancreatic CD (arrow).

2, and Bcl-6 positivity confirmed the diagnosis of CD. The authors commented on diagnostic difficulties related to fine needle aspiration or core needle biopsy.[4] Liu et al[5] reported a 28-year-old female with a mass on pancreatic neck incidentally found by a routine ultrasound evaluation, and PET/CT showed increased ^{18}F-fluorodeoxyglucose uptake, and slightly increased somatostatin receptor expression on ^{68}Ga-DOTATATE images. As intraoperative histopathology study showed lymphatic hyperplasia, she underwent the tumor resection and the immunohistochemistry diagnosis was a hyaline vascular variant of CD.[5] The authors emphasized that 90% of the hyaline vascular CD are unicentric, and the best management is by complete removal of the affected lymph nodes.[5] Yazdi et al[6] described a 31-year-old female with abdominal pain and images of a mass at the head and uncinate process of the pancreas. The fine needle aspiration biopsy discarded malignancy, and the mass resected by laparotomy was diagnosed as hyaline vascular CD; the patient received no other therapy, and persists asymptomatic after 6 months of follow-up.[6] The authors commented diagnostic challenges of CD, besides the lack of consensual managing procedures for this condition.[6] Sawaya et al[7] reported a 34-year-old female with persistent hematuria after a vaginal delivery, and imaging studies detected a well-vascularized retroperitoneal sited mass between the liver, right kidney, and duodenum, and presenting enhancement after the intravenous contrast administration. The endoscopic ultrasound-guided biopsy showed multiple lymphoid cells, and the patient underwent the laparoscopic successful resection of the mass besides some large lymph nodes.[7] The authors emphasized the rare case of the unicentric para-duodenal retroperitoneal CD.[7] Wang et al[8] reviewed clinical and surgical data of 25 patients with retroperitoneal unicentric CD; the mean age was 43.80 (±12.79 years), 52.0% were men, the masses were in kidney or perinephric, adrenal, pancreas or peripancreatic, duodenal, periaortic, besides of iliac artery. All patients had total resection of masses, 96.0% had the hyaline-vascular CD, 92.0% had a long-term follow-up with a mean time of 35.4±33.9 months; there was no death or relapse.[8] Zhai et al[9] described an asymptomatic 44-year-old female with an incidental hypoechoic mass between the body of the pancreas, the liver and stomach showed by a routine ultrasound scan. CT and magnetic resonance images (MRI) suggested a solid pseudopapillary tumor or NET, but she underwent a surgery before confirmed diagnosis, that was established as a pancreatic CD with positive immunostaining for CD10, CD20, CD21, CD23, CD30, CD138, Bcl-2, and Bcl-6.[9] Zhou et al[10] reported a 33-year-old asymptomatic female who underwent a laparotomy due to enlarged peripancreatic lymph nodes, and a mass between the liver, stomach, and pancreas; the abnormal tissues were removed and she did not utilize adjuvant radiotherapy or chemotherapy. CD3, CD10, CD20, CD21, CD23, CD43, CD79α, Bcl-2, and Bcl-6 were the positive markers, considered consistent with the final diagnosis of a peripancreatic CD hyaline-vascular type.[10] The authors commented on the lack of consensus for treatment of unicentric CD, and the option of watch-and-wait in asymptomatic patients for whom the surgery is impossible or mutilating.[10]

In conclusion, the present case study of a middle-aged man with confirmed diagnosis of unicentric CD sited in the pancreas and managed with success by corpus-caudal pancreatectomy and splenectomy aims to enhance the interest about this uncommon and very challenging condition.

ΠΕΡΙΛΗΨΗ

Νόσος του Castleman μιμούμενη νεόπλασμα παγκρέατος

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Η νόσος Castleman είναι δύσκολη στη διάγνωση, ακόμη και με βάση τα ιστοπαθολογικά και ανοσοϊστοχημικά δεδομένα από τη βιοψία των βλαβών. Περιγράφεται μια περίπτωση ενός προηγουμένως υγιούς άνδρα, ηλικίας 35 ετών, ο οποίος παρουσίαζε μια υπεραγγειακή μάζα με ασβεστώσεις στην περιοχή του παγκρέατος. Σχολιάζεται η αρχική δύσκολη ερμηνεία εργαστηριακών εξετάσεων ρουτίνας και απεικονιστικών μελετών, εκτός από τα ιστοπαθολογικά και ανοσοϊστοχημικά δεδομένα. Η τελική διάγνωση της νόσου Castleman τέθηκε μετά τη διαδικασία της σωματικής-ουραίας παγκρεατεκτομής και σπληνεκτομής, που διενεργήθηκε επιτυχώς με λαπαροσκόπηση. Ακόμη και μεμονω-

μένες περιπτώσεις μπορεί να ενισχύσουν την υποψία μεταξύ των μη εξειδικευμένων σχετικά με τη νόσο Castleman που μιμείται έναν πρωτοπαθή νευροενδοκρινικό όγκο.

Λέξεις ευρετηρίου: Διάγνωση, Θεραπεία, Νόσος Castleman, Παγκρεατική νεοπλασία

References

1. BHATIA V, SINGH SP, MISHRA AK, RATH I, PRABHU M, BIHARI C. Cytological diagnosis of Castleman disease by endoscopic ultrasound guided fine needle aspiration. *J Cytol* 2023, 40:145–147
2. DEV S, POKHREL KM, POKHAREL P, BHANDARI RS, PRADHAN S. Castleman disease of the pancreas mimicking pancreatic neuroendocrine tumor: A rare case report. *Int J Surg Case Rep* 2023, 109:108524
3. GUNDA D, NAUGHTON J, STEVENS SG, PERINI MV. Castleman's disease masquerading as pancreatic neuroendocrine tumour. *BMJ Case Rep* 2021, 14:e242597
4. HUANG Z, JIN T, ZHANG X, WU Z. Pancreatic mass found to be Castleman disease, a rare case report. *Asian J Surg* 2020, 43:767–768
5. LIU SL, LUO M, GOU HX, YANG XL, HE K. Castleman disease of the pancreas mimicking pancreatic malignancy on ^{68}Ga-DOTATATE and ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography: A case report. *World J Gastrointest Surg* 2022, 14:514–520
6. YAZDI SAM, NAZAR E. Retroperitoneal unicentric Castleman's disease, a case report. *Ann Med Surg (Lond)* 2022, 79:104109
7. SAWAYA Z, SEMAAN DB, NICOLAS G, DIB A, TAYAR C. Unicentric Castleman's disease: Laparoscopic approach of a para-duodenal retroperitoneal mass. *Am J Case Rep* 2020, 21:e918444
8. WANG W, DONG D, WEN J, LI H. A 10-year observational single-center study of retroperitoneal unicentric Castleman disease. *Medicine (Baltimore)* 2021, 100:e25088
9. ZHAI HY, ZHU XY, ZHOU GM, ZHU L, GUO DD, ZHANG H. Unicentric Castleman disease was misdiagnosed as pancreatic mass: A case report. *World J Clin Cases* 2022, 10:1278–1285
10. ZHOU YX, JI Y, WU S. A CARE-compliant article: Unicentric Castleman disease presenting as a retroperitoneal mass of the upper edge of the pancreas: A case report. *Medicine (Baltimore)* 2020, 99:e19515

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