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CASE REPORT | PATHOLOGY

Castleman Disease Presenting as an Abdominal Mass

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ABSTRACT

Unicentric Castleman disease is a rare condition of lymphoid hyperplasia, of which only 15% of cases occur in the abdomen. We report a 66-year-old man who presented with complaints of abdominal pain. Computed tomography scans revealed nephrolithiasis and a homogeneous calcified mass between the pancreas and stomach and several para-pancreatic nodes. Direct visualization during exploratory laparotomy revealed a mass on the lesser curvature of the stomach. Pyloromyotomy and mass resection were performed. Biopsy showed reactive lymphoid hyperplasia consistent with the hyaline vascular variant of Castleman disease.

INTRODUCTION

Castleman disease (CD) is a rare cause of lymphoid hyperplasia.[1,2] Unicentric Castleman disease (UCD) is usually asymptomatic with a 95.3% survival rate.[1,3,4] Initial identification of CD usually occurs during physical exam or imaging.[5] Only 15% of UCD lesions are found in the abdomen.[3,6,7] Diagnosis requires excisional biopsy and pathological identification of lymphoid hyperplasia of the hyaline vascular variant or plasma cell variant.[1] UCD is best managed by surgical resection, although siltuximab has been shown to be effective. CD is associated with lymphoma, therefore close follow-up is imperative in these patients.[8]

CASE REPORT

A 66-year-old man presented with acute-onset right-sided abdominal pain. Computed tomography (CT) of the abdomen with contrast revealed nephrolithiasis and a 29-mm mass between the pancreatic and gastric bodies (Figure 1). The patient denied weight loss, diaphoresis, or lymphadenopathy, and his abdominal pain resolved by his next follow-up visit a month later. Other symptoms included a diffuse pruritic rash later defined as lichen simplex chronicus.

Endoscopic ultrasound fine-needle aspiration (EUS-FNA) was performed with an Olympus scope GF-UCT180 (Olympus, Center Valley, PA). The procedure revealed a hypoechoic 30 x 32 mm mass with calcifications and well-defined borders (Figure 2). The mass appeared to be attached to the nodular, fat-stranded pancreatic body and the lesser curvature of the muscularis propria, and it was identified as either a pancreatic mass or lymph node. Several round, well-defined hypoechoic peripancreatic lymph nodes were also noted, the largest being 20 x 10 mm. FNA of the mass was performed via a transgastric dry suction approach using a 22-gauge needle and stylet. Prior to aspiration, color Doppler confirmed the absence of vasculature in the needle path. Five passes were made, but biopsy of the aspirate was indeterminate, revealing only lymphocytes. Flow cytometric analysis was also performed, showing populations of polyclonal B lymphocytes and T lymphocytes that were immunophenotypically unremarkable other than a nonspecific increased CD4 to CD8 ratio. The blast gate was not increased.

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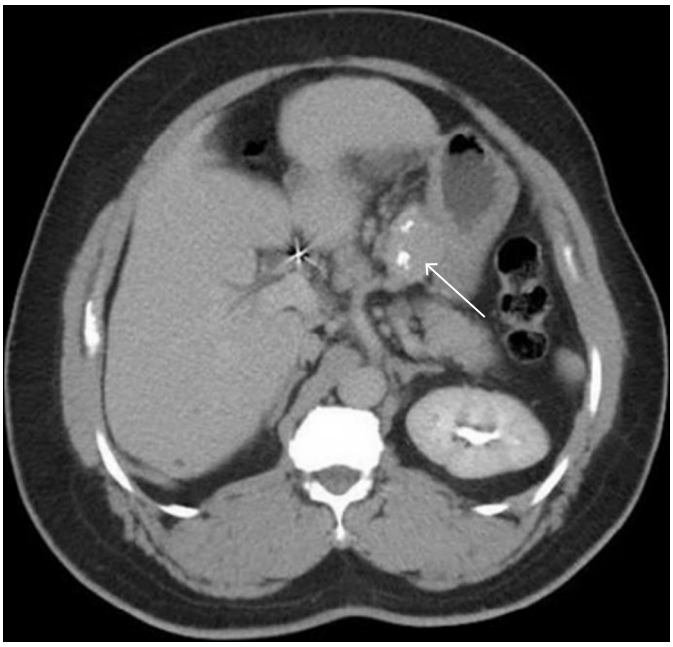

Figure 1. CT scan of the abdomen showing a calcified homogeneous mass measuring 3.7 cm between the pancreatic and gastric body (arrow).

At 1-month follow-up, EUS and core biopsy revealed an unchanged, well-defined, 30 x 32 mm hypoechoic calcific mass and enlarged lymph nodes on the pancreatic body. Calcification of the pancreatic duct was visualized. FNA of the pancreatic body mass was performed using the same technique and transgastric approach, and three passes were made with a 22-gauge needle. Core biopsy, performed using the SharkCore™ FNB Exchange System (Medtronic, Minneapolis, MN), revealed small fragments of fibroconnective tissue with chronic inflammation and blood clot without malignancy. FNA of peripancreatic lymph nodes revealed indeterminate findings of small fragments of benign lymphoid tissue.

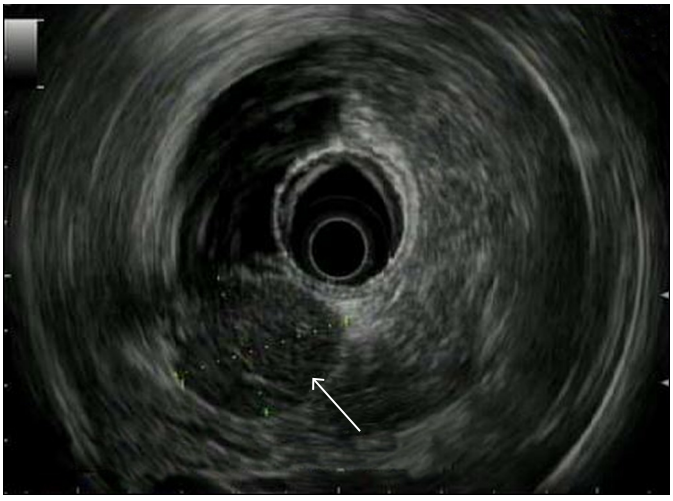

Figure 2. EUS revealing a round, hypoechoic, calcified mass (arrow) with well-defined borders in the pancreatic body invading the lesser curvature of the muscularis propria. Several abnormal lymph nodes were also visualized in the peripancreatic region.

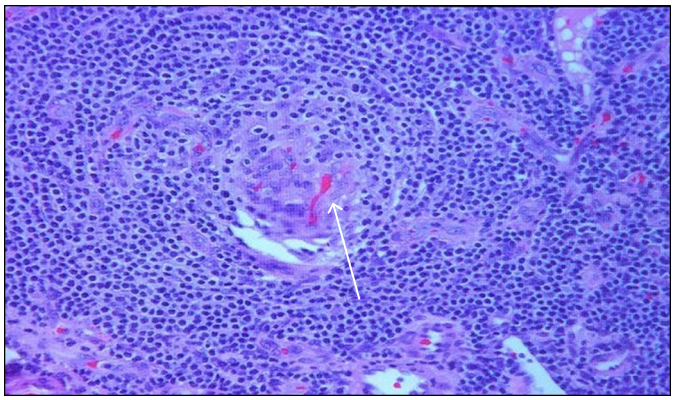

Figure 3. Pathology of the mass showing a small follicle with a partially lymphocyte-depleted germinal center with hyaline deposits (arrow). Capillaries and small vessels are present in the germinal center within the interfollicular space, showing the unique lollipop configuration.

Three months after the first presentation, the patient presented to an outside hospital with lightheadedness and hypoglycemia. Their laboratory tests revealed elevated insulin and C peptide. His worsening symptoms, rash, abnormal laboratory values, and inconclusive cytological diagnosis were concerning for neuroendocrine tumor. Glucagon levels (180 pg/mL), neuron specific enolase (25 ng/mL), and prolactin (23.3 ng/mL) were elevated. Chromogranin A, pancreatic polypeptide, and serum serotonin were negative.

Due to uncertainly regarding the location of the lesion and malignancy status, the patient was referred for exploratory laparotomy. Direct surgical visualization revealed a 5-cm hypervascular mass adherent to the lesser curvature of the stomach with regional lymphadenopathy. Resection of the lesser curvature of the stomach and pyloromyotomy was completed. Pathological evaluation showed a lymph node with reactive lymphoid hyperplasia characterized by regressively transformed germinal centers with increased interfollicular vascularity, consistent with the hyaline vascular variant of Castleman disease (Figure 3). Repeat flow cytometric analysis of tissue showed populations of polyclonal B lymphocytes and T lymphocytes that were immunophenotypically unremarkable. Viral serology, including hepatitis and HIV, was negative. Postsurgical recovery was complicated by delayed wound healing. He remained active with no gastrointestinal symptoms or development of new lymph nodes, and he was scheduled for future lymphoma screening.

DISCUSSION

Our patient presented with vague abdominal pain secondary to nephrolithiasis. The abdominal mass was only found

incidentally, which is typical of UCD.[9] Identification of CD was delayed, despite two EUS with FNA and biopsy, and definitive diagnosis was obtained only after surgical resection. While EUS is a powerful and noninvasive diagnostic tool used to identify abnormal cytology in the pancreatobiliary region, it does not allow for accurate visualization of anatomic location or histological evaluation, which are crucial diagnostic steps when considering CD. One should consider following up with histological evaluation when EUS is inconclusive. Though we were unable to obtain a definitive diagnosis using biopsy alone, cases have reported diagnosing CD using EUS-FNA and CT-guided biopsy.[10,11] Real-time tissue elastography has also been indicated in CD diagnosis; however, elastography is not recommended by the National Comprehensive Cancer Network (NCCN) current guidelines.[12,13] Flow cytometry showed no abnormal cell populations, thus ruling out other concerning lesions; this does not, however, confirm CD diagnosis.[1]

Diagnosis was further complicated because CD rarely presents in a primary organ.[4] Our patient's mass was initially identified as a pancreatic mass, but surgical removal of the mass revealed lymphoid tissue on the lesser curvature of the stomach. Unicentric lesions are most commonly found in the chest, although they have also been found in the pelvis, retroperitoneum, neck, and axilla. Some CD cases have been reported in the peripancreatic and pancreatic duct region.[7,10,11,14] A systematic review of 404 published cases of CD found that UCD is best managed by surgical resection.[4] Following this practice, surgical resection was performed. NCCN recommends postsurgical observation with positron emission tomography or CT only if there are signs of recurrence.[13] We emphasize the importance of including rare diseases as differentials for an abdominal mass. If recognized early, it can streamline treatment and lymphoma screening.

DISCLOSURES

Author contributions: F. Shariati, E. Verter, and W. Chang wrote the manuscript. L. Huang interpreted the pathology. J. Virendra reviewed the manuscript for important intellectual content and is the article guarantor.

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