Castleman's Disease Presenting as Peri - Pancreatic Neoplasm

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Abstract Castleman's disease (CD) is a relatively singular disorder. Usually presenting as a mediastinal mass, peri pancreatic localization of CD is even rarer. Diagnosis is seldom established pre-operatively. We report two such cases of CD in which peri pancreatic region was involved and were diagnosed on biopsy, hence avoiding complex and high risk surgery.

Keywords Castleman's disease · Pancreatic neoplasm

Introduction

Castelman's disease (CD) or angiofollicular lymph node hyperplasia, first described by Benjamin Castleman, in 1956, in patients with mediastinal localized disease, is a rare heterogenic lymphoproliferative disorder [1]. Its presentation as peripankreatic mass results in diagnostic dilemma as preoperative degree of suspicion of the same is less. We present two cases which were diagnosed by biopsy and treated conservatively.

Case Reports

Case 1: 34 year old lady presented with complaints of abdominal pain of 5 months duration and weight loss of 5 Kg. She had prior history of appendectomy. Physical examination

did not reveal any abdominal mass or generalized lymphadenopathy. Tumor markers CA 19-9 and CEA were within normal range. Contrast enhanced CT (CECT) abdomen revealed a mass lesion anterior to the neck and body of pancreas with foci of calcification (Fig. 1). Ultrasonography guided biopsy revealed lymphoplasmacytic lesion (plasma cells with CD 138 positivity) but did not rule out neoplasm. With a working diagnosis of unclear pancreatic neoplasm, she was scheduled for pancreaticoduodenectomy (PD). Operative exploration revealed a large nodal mass in gastro-hepatic ligament with multiple conglomerate nodes adjacent to the celiac trunk. Excision biopsy of one of the nodal masses was done which on frozen section showed plasma cells with lymphocytes and no evidence of malignancy. Hence PD was deferred. Histopathology report revealed the mass to be atypical lymphoid proliferation showing prominent follicular pattern consistent with features of CD (Fig. 2). She was treated conservatively and is currently asymptomatic for the disease at 2 year follow up.

Case 2: 51 year old lady presented with incidentally detected retroperitoneal (RP) mass on CT on routine examination. She had prior complaints of hypertension, nephrotic syndrome and hypothyroidism. Physical examination did not reveal any abdominal pathology. Serum CA 19-9 was 27 ng/mL. CECT abdomen showed a 6 cm intensely non-homogenously enhancing mass lesion in RP posterior to duodenum. Fat plane with inferior vena cava (IVC) was maintained (Fig. 3). Endoscopic ultrasound (EUS) showed well defined RP mass free from pancreas and duodenum. EUS biopsy, however, was not conclusive. CT guided biopsy revealed atypical cells. She was lost to follow up for 8 months but continued to remain asymptomatic. Recent CT guided biopsy revealed reactive lymph node with CD like features. Immunohistochemistry (IHC) revealed CD20 reactive B cells and CD3 reactive T cells (Fig. 4). Staining for CD30, CD56, chromogranin, CD10 and cyclin D1 were negative. There has been no interval change in the size of lesion on CT. She is on

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Fig. 1 CECT of patient 1 showing lesion anterior to pancreatic body

regular follow up since then and has remained asymptomatic for the lesion since last 6 months.

Discussion

CD is a rare lymphoproliferative disorder mostly affecting the mediastinum. Peri pancreatic location is unusual [2]. There is no consensus on underlying etiology of CD. Theories suggesting reactive lymphoid hyperplasia and vascular lymphoid hamartoma have been proposed [3]. Although in the first case etiology is unknown, in the second case CD may have arisen in a background of chronic immunosuppression by steroids. Clinically, there are two variations. Localized Castleman's disease (LCD) only affects a single set of lymph nodes and more common among younger adults. The other clinical form is multicentric Castleman's disease (MCD) which is more debilitating than LCD and seen in elderly and those with HIV infection [4]. LCD was the mode of presentation in our two patients.

Histopathologically, CD presents with three distinct histological variants—the hyaline vascular (HV) type, the plasma cell type (PC) and the mixed type (MV) [3]. Both our patients were found to have HV type on histology. The initial challenge in CD remains in establishing the diagnosis and differentiating the disease mimicking CD which includes infectious and inflammatory lesions, and CD disease involving the peri-pancreatic region from pancreatic carcinoma. IHC thus plays an important role in confirming these lesions. IHC would

Fig. 3 CECT of patient 2 showing lesion posterior to duodenum

Fig. 2 Histopathology slide showing characteristic follicular pattern

show characteristic follicular dendritic network of residual germinal centers with CD 20 and CD 3 reactivity and negative for CD30, CD56, CD10 and BCL-2 which are reactive in lymphoid malignancies.

Radiological diagnosis of CD is usually not possible pre-operatively as it mimics many common disorders. Retrospective review by Meador et al. showed calcification in 31 %, one with an arborizing pattern and the remaining lesions with punctuate calcifications [5]. EUS is an excellent tool for characterizing 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 inter-observer agreement is poor [6]. The decision to biopsy retroperitoneal masses pre-operatively remains controversial. Most surgeons agree that surgical resection in the appropriate clinical setting without biopsy is acceptable [7]. In all but two of the reported pancreatic and peri pancreatic CD cases (Table 1) the diagnosis was established after histopathological examination of surgically resected specimens [10, 12]. Our second case was diagnosed purely based on the CT guided biopsy. Hence we would suggest preoperative biopsy especially in a setting of peri pancreatic lesions as differential of these lesions could range from lymphoma to pancreatic adenocarcinoma. Diagnosis of CD in such cases would alleviate patient anxiety and avoid unwarranted neoadjuvant therapy.

Fig. 4 Immunohistochemistry picture of CD 20 positive germinal centers

Table 1 Summary of reported cases of peri pancreatic CD

Study	Location	Symptoms	Sub type	Treatment	Preoperative diagnosis	Follow up
Baikovas [8]	Peripancreatic	None	HV	Excision	None	NA
Erkan [9]	Peripancreatic	Abdominal pain	PC	Enucleation	None	NER—1 year
Rhee [10]	Peripancreatic	None	HV	Excision	CD by EUS-trucut biopsy	NER
Talarico [2]	Peripancreatic	Abdominal pain	HV	Excision	None	NER—1 year
Charalabopoulos [11]	Peripancreatic	Abdominal pain	PC	Distal pancreatectomy	None	NER—2 year
Khasab [12]	Peripancreatic	None	HV	Excision	CD by EUS-FNAC	NA

CD Castelman's disease, BOP body of pancreas, TOP tail of pancreas, HV hyaline-vascular type, PC plasma cell type, DP distal pancreatectomy, EUS FNAC endoscopic ultrasound fine needle aspiration cytology, NER no evidence of disease recurrence, NA not available

Surgical excision has been the mainstay of treatment for LCD especially those localized to pancreas as suggested by Bowne et al. [13]. In the absence of progression of the disease, conservative management of our two cases, provide some evidence to prove the contrary. This would thus avoid a morbid surgery. Regular follow up with CECT, of such conservatively managed patients, is recommended.

Conclusion

CD being a rarity and its ability to mimic other common diseases, establishing a preoperative diagnosis of the same has been found to be challenging. While offering a PD remains an option for all pancreatic and peri-pancreatic solid masses in the absence of a tissue diagnosis, a high index of suspicion ought to be retained by the clinician in unusual situations. Furthermore, our experience shows that these cases can be managed conservatively especially in asymptomatic patients.

Conflict of Interest None

Financial Disclosures None

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