Inflammatory Myofibroblastic Tumour of Pancreas- A Case Report

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INTRODUCTION

Inflammatory Myofibroblastic Tumour (IMT) of pancreas is a rare benign mesenchymal neoplasm with a predilection for lung and abdomino-pelvic region. Other sites of occurrence are orbit, peritoneum, mesentery, and pancreas. In children about one-third cases were seen in lung and two-thirds extrapulmonary sites.[1] These tumours were previously known as inflammatory pseudotumour because on histopathological examination consists of myofibroblastic and fibroblastic proliferation with an inflammatory infiltrate.[2] This tumour was first described in the lung, and very rare in pancreas.[3] It is mostly seen in children and young adults with a worldwide prevalence rate of 0.04%-0.7%.[4,5]

PRESENTATION OF CASE

A 31 year old male presented to the Department of Oncology, Government Medical College, Thrissur, with complaints of abdominal pain and vomiting. History of weight loss and malaise was present for the last six months. No history of any co-morbidities or no history of malignancy in the family. On physical examination there was a mass palpable in the epigastrium. Basic laboratory investigations including complete blood count, renal function test, liver function test, serum amylase, serum lipase, CEA, CA -19-9 were with in normal limits.

Radiological Findings

Contrast Enhancing Computed tomography of abdomen shows large solid enhancing lesion with cystic areas arising from the head of pancreas with a size of 8.5 x 11 x 13.6 cms., suggestive of pseudocyst (Figure 1).

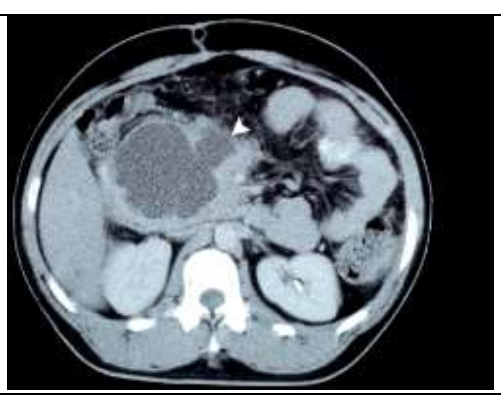

Figure 1

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DOI: 10.18410/jebmh/2020/52

Financial or Other Competing Interests: None.

How to Cite This Article:
Archana S, Prema KR, Balakrishnan S, et al. Inflammatory myofibroblastic tumour of pancreas- a case report. J. Evid. Based Med. Healthc. 2020; 7(5), 246-248. DOI: 10.18410/jebmh/2020/52

Submission 26-12-2019,
Peer Review 29-12-2019,
Acceptance 30-01-2020,
Published 03-02-2020.

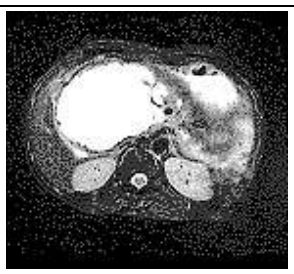

Figure 2

MRCP shows well-defined cystic lesion in the abdomen in the right hypochondrium. Head and uncinate process of pancreas seen separately from lesion. Lesion encases superior mesenteric vessels, compression of portal vein and inferior vena cava with a small portion of bowel within the tumour (Figure. 2). He underwent laparotomy, and tumour was inoperable due to local infiltration into major vessels. Patient has features of intestinal obstruction, so palliative gastrojejunostomy was done. Also, debulking and biopsy was taken from the tumour.

CLINICAL DIAGNOSIS

Pseudocyst of Pancreas.

DIFFERENTIAL DIAGNOSIS

Gastrointestinal Stromal Tumour.
Fibrosarcoma.
Inflammatory Myofibroblastic Tumour.

PATHOLOGICAL DISCUSSION

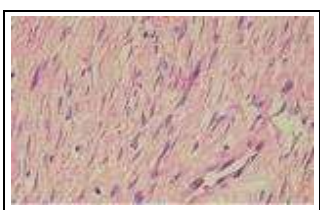

Figure 3

Figure 4

Figure 5

Histological examination shows loosely and diffusely arranged spindle cells. Some of them are stellate shaped and having myofibroblastic morphology, with scattered inflammatory cells composed of lymphocytes and occasional plasma cells. No mitosis and neoplastic cells show mild pleomorphism. No necrosis seen (Figure 3). Immunohistochemistry shows spindle cell neoplasm, positive for Vimentin (Figure 4), and positive for SMA (40%) (Figure 5). Negative for ALK, CD 117, S100, DOG1 and desmin. Ki 67 index is low (<3%).

DISCUSSION OF MANAGEMENT

Inflammatory myofibroblastic tumour is a rare benign tumour affecting the pancreas which is difficult to differentiate from pancreatic cancer clinically and radiologically as in our case. Differential diagnosis were adenocarcinoma pancreas, fibrosarcoma, and leiomyosarcoma of pancreas. It was only after the immunohistochemical study of the resected specimen that the diagnosis of inflammatory myofibroblastic tumour could be established.

Aetiology of inflammatory myofibroblastic tumour is unknown, an inflammatory reaction in an underlying low grade malignancy was considered as the cause.[6] But there is only limited information available, and development of IMT is still unknown, association has been reported with infections, previous abdominal surgery or steroid use.[7] On pathological examination there is mainly myofibroblasts with less fibroblastic component with an inflammatory infiltrate. Immunohistochemistry show smooth muscle actin (SMA) positivity in 80-90% of inflammatory myofibroblastic tumours.[8] Also vimentin is positive in 50% of tumours.[9] About 50% of tumours show ALK (Anaplastic Lymphoma Kinase) gene translocation on chromosome 2p23, and this ALK expression is more seen in younger age group.[10,11] Mitotic activity is low, about 0-2 mitoses per 10 high power field.[12,13] Depending upon the site of the tumour symptoms vary and some of them are asymptomatic. Common symptoms of pancreatic IMT are abdominal pain, back pain and vomiting. Fever, weight loss and malaise, and raised ESR is seen in 15-20% of cases. IMTs involving the head of pancreas presenting with obstructive jaundice due to extrinsic compression of biliary system.[14] Surgical resection is the treatment of choice.[6] But in aggressive type of inflammatory myofibroblastic tumours the complete resection is not possible due to adherence and local infiltration to adjacent structures and blood vessels as in our case.[4] There is high chance of local recurrence after incomplete resection.[14,15] Metastasis to other parts of the body is seen in < 5% cases.[9] Distant metastasis commonly occurs in lung, brain, liver and bone.[16,17] Medical management with corticosteroids and nonsteroidal antiinflammatory drugs are given for inoperable cases.[18] Chemotherapy may be benefited in aggressive type of inflammatory myofibroblastic tumours after incomplete resection, tumours with distant metastasis, and those

tumours recurred after resection.[19] Our case is inoperable due to vessel invasion and treated with steroids and nonsteroidal anti-inflammatory drug (celecoxib) for the last six months. He became asymptomatic and improved his general condition after two months of starting treatment, so we are continuing same medicines. Because of the rare incidence and difficulty in diagnosis we presented this case. It may be misdiagnosed as pancreatic cancer, and the management is totally different in these two cases.

FINAL DIAGNOSIS

Inflammatory Myofibroblastic Tumour

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