A Retroperitoneal Inflammatory Pseudotumor involving Duodenum: A Case Report and Review of literature.

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Abstract
Inflammatory pseudotumor or (IPT) is a rare lesion of unclear etiology reported in various organs. Although mostly benign, these tumors may pose a therapeutic challenge in cases of recurrence. We report the case of a 65-years-old male who presented with a mass in the left upper abdomen and upon evaluation was noted to have IPT in the retro-peritoneum involving the duodenum. Complete surgical resection was done with primary repair of the 3rd part of the duodenum with no evidence of tumor recurrence on 6 months follow-up. We review the literature and discern the epidemiological, clinical, pathophysiological and management aspects of IPTs.

Introduction
Inflammatory pseudotumors (IPTs) are rare, well-circumscribed, unencapsulated, quasineoplastic tumors of unregulated growth of inflammatory cell, first recognized by Umiker and Iverson¹. IPTs are rare benign process mimicking malignant process and most commonly involving the lung and orbit but found in nearly every site in the body. Various terms have been used to describe them based on the predominance of the particular type of cytology, such as plasma cell granuloma, plasma cell pseudotumor, inflammatory myofibroblastic proliferation, omental-mesenteric hamartoma, histocytoma and fibroxanthoma. It can roughly be categorized into an organizing pneumonia pattern, a fibrous histiocytic pattern (most common) and a lympho-histocytic pattern (least common)². It is not yet established if these lesions represent a primary inflammatory process or a low-grade malignancy with a prominent inflammatory response³.

Case report:
We report a case of a 65-years-old male who presented with a lump in the left upper abdomen with mild pain, recurrent low grade fever, generalized weakness, anorexia and weight loss of 7 months duration. He was a farmer and smoker. He had no history of chest pain, cough, hemoptysis, jaundice, bone pain. On examination, he was average built, and had pallor. A lump was found in left hypochondric region about 8x7cm, mild tender, ill define margin, not mobile, hard in consistence. Lab investigations revealed high erythrocyte sedimentation rate (ESR, 128mm/hour), anaemia (9gm/dl), and thrombocytosis. Ultrasonography of the abdomen revealed one irregular solid inhomogenous structure (about 7.2 x 6.3cm) having internal 'ring down' shadow in the left hypochondric region, anterior to the left kidney (suggestive of bowel mass). USG guided Fine needle aspiration cytology (FNAC) was done...
which was suggestive of chronic granulomatous inflammation consistent with tuberculosis. Full video colonoscopy showed normal colon. Contrast-enhanced computed tomography (CT) abdomen, revealed a large heterogenous soft tissue mass having areas of necrosis with air fluid level measuring about 9.5 x 8.0 x 7.0 cm was noted in the left upper abdomen, attached to the bowel loop compressing the tail of the pancreas. The mass was separated from the spleen, kidney and stomach. The mass was suggestive of small bowel GIST in left upper abdomen. Patient was taken up for laparotomy and a large mass, measuring about 8x7cm, found in the retroperitoneal region, attached to the 3rd part of the duodenum. The mass was resected completely with repair of the 3rd part of duodenum. Grossly, the specimen was globular grey brown, cut surface was solid, in some areas small cystic spaces were found. Histopathology showed extensive areas of fibroblastic proliferation with multiple focal areas containing polymorphs, lymphocytes, plasma cells and eosinophils, with areas of abscess formation. No granuloma or malignancy was seen (Figure-1). Final diagnosis of inflammatory pseudotumor of retroperitoneum involving duodenum was made.

**Figure-1:** Histopathology of inflammatory pseudotumor.

On immediate post-operative period patient developed duodenal fistula that responded subsequently by conservative management and discharged on 25th post operative day. The patient had been doing well and a follow-up of 6 months showed no evidence of tumor recurrence. There was a feeling of well-being with acceptable weight gain was reported.

**Discussion**

Though retroperitoneal inflammatory pseudotumors (IPTs) are rare, they pose a diagnostic and/or therapeutic challenge to the managing team of clinicians, pathologists, and surgeons. IPT is a distinctive pseudosarcomatous lesion occuring primarily in the viscera and soft tissues. It is seen mostly in children and young adults (half of the patients are less than 40 years) with an equal sex distribution. Although initially reported and is more common in the lung, it has a propensity to involve the orbit, stomach, testis, oesophagus, liver, spleen, pancreas, kidney, adrenal gland, retroperitoneum, diaphragm, mesentery, bladder, heart, thyroid, tonsil, fourth ventricle, spinal cord meninges, central nervous system, maxillary sinus, nasopharynx, larynx and trachea. IPT can present as a single mass or multiple masses with polymorphous inflammatory cell infiltrate and variable amounts of fibrosis, necrosis, granulomatous reaction and myofibroblastic spindle cells. Patient may present with fever, weight loss, growth retardation, thrombocytosis, iron deficiency anaemia, hypergammaglobulinemia, symptoms related to mass effect or a combination of these signs and symptoms. Elevated ESR, anaemia, thrombocytosis and hypergamma globulinemia, which characteristically resolve after the lesion is excised. The cause of IPT is unknown. Some of the postulated pathophysiological mechanism include trauma or surgery, immune- autoimmune mechanism, associated with other malignancy or secondary to infection. Various organisms have been implicated in pathologic specimens, including mycoplasmata and nocardiae in lung pseudotumors, actinomycetes in liver pseudotumors, Epstein-Barr virus in splenic and nodol pseudotumors, and mycobacteria in spindle cell tumors.

The radiologic features of IPT are variable and nonspecific, possible because of the amount of fibrosis and cellular infiltration. On ultrasound images, lesions can be hypoechoic or hyperechoic with either ill-defined or well-circumscribed
borders\textsuperscript{11}. These lesions often have increased vascularity during color or power Doppler examinations. CT also shows varying appearances; lesions can have low, equal, or high attenuation compared with the surrounding tissue\textsuperscript{11,12}. Contrast-enhanced CT may show a homogenous or heterogenous lesion. The imaging features of gastrointestinal inflammatory pseudotumor include ulceration, wall infiltration, and extra luminal extention, findings similarly seen malignant disease\textsuperscript{13}.

Proliferation of spindle cells, arranged in short fascicles with a focal storiform (whorled or cartwheel-like) architecture, associated with a variable dense polymorphic infiltrate of mononuclear inflammatory cells is the most consistent histologic finding\textsuperscript{14}. Cytologic atypia, ganglion-like cells, \textit{p53} expression and DNA aneuploidy along with focal and/or vascular invasion and nuclear pleiomorphism are suggestive of more malignant behavior and a worse prognosis\textsuperscript{15}.

Complete surgical resection is the definitive treatment. If the patient are poor surgical candidates or have multiple nodules or unresectable disease, nonsurgical modalities like treatment with glucocorticoids, radiotherapy, and chemotherapy may be tried. Complete tumor excision and tumor size ≤ 3cm are factors associated with decreased risk of recurrence. Malignant transformation, although rare, has been reported.

\textbf{Referances:}
\begin{enumerate}
\item Dehner LP. The enigmatic inflammatory pseudotumors: The current state of our understanding or misunderstanding. J Pathol. 2000; 192: 277-9.
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