

Reactive nodular fibrous pseudotumor: A rare mesenchymal tumor

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ABSTRACT

Reactive nodular fibrous pseudotumor (RNFP) is a recently described non-neoplastic mesenchymal lesion arising from the gastrointestinal tract, mesentery, or retroperitoneum. It is a rare entity with only a few cases reported in literature so far and is less aggressive when compared to other mesenchymal lesions such as gastrointestinal stromal tumor (GIST) or fibromatosis. Here, we present the case of a 20-year-old female who presented with a huge abdominal mass. Contrast-enhanced computed tomography was suggestive of pseudopapillary tumor of the pancreas. The patient underwent exploratory laparotomy and excision of the mass in toto along with sleeve gastrectomy and splenectomy. Post-operative histopathology confirmed the entity to be RNFP. This case illustrates the need for keeping RNFP in mind as a differential diagnosis in a case of large abdominal mass as it has a fairly good prognosis with no reported recurrence after surgical excision.

Key words: *Fibromatosis, Gastrointestinal stromal tumor, Mesenchymal lesion, Reactive nodular fibrous pseudotumor*

Reactive nodular fibrous pseudotumor (RNFP) is a recently described rare differential diagnosis of mass occurring in the abdominal cavity. It is a non-neoplastic lesion which mimics malignancy preoperatively. Usually, it arises from the mesentery, may also involve colon or small bowel [1]. The pathogenesis of RNFP is possibly linked to endometriosis, ergotamine use, or prior abdominal surgery [2]. The exact prevalence of this condition is unknown and till date only around 20 cases has been reported in literature. We describe the first case in India in a young female aged 20 years with a huge abdominal mass.

CASE REPORT

A 20-year-old female with no significant medical or surgical history presented with complaints of mass per abdomen of 1-month duration which was progressively increasing in size. There were no other complaints associated with the mass. On examination, the general condition of the patient was fair and vitals were stable. The abdominal examination revealed a huge mass of 15 cm × 10 cm mainly occupying the left hypochondrium and the left lumbar region, extending into the epigastrium and umbilical region. It was firm to hard in consistency, non-tender with a smooth surface, and well-defined margins.

Blood investigations such as hemogram, renal function test, and liver function test were within normal limits and serum CA-19-9 was normal. Contrast-enhanced computed tomography of the abdomen and pelvis showed a well-defined heterogeneously enhancing soft tissue hypodense mass measuring 14.6 cm × 12.4 cm × 13.2 cm (CC × AP × TR) occupying the left

retroperitoneum region extending from D11-L4 vertebral level. It was causing breaking of the pancreas with splenic artery coursing through the lesion, most likely arising from the pancreas. On the basis of above-mentioned clinical features and investigation, a provisional diagnosis of solid pseudopapillary tumor of the pancreas was made (Figs. 1 and 2).

The patient underwent exploratory laparotomy. Intraoperatively, the mass was found adherent to the greater curvature of the stomach, distal pancreas, spleen, and splenic vessels. The mass was excised in toto along with sleeve gastrectomy and splenectomy (Figs. 3-5). Grossly, it was a well-circumscribed lesion measuring 20 cm × 14 cm with homogenous glistening gray-white areas and focal myxoid areas (Fig. 6). Histopathology of the mass showed a circumscribed unencapsulated paucicellular lesion composed of a spindle to stellate myofibroblast, foci of hyalinization, keloid-like collagen deposition with numerous interspersed mast cells, and an absence of atypical or bizarre forms, giving a final diagnosis of RNFP. The spleen, stomach, and lymph nodes were free. Immunohistochemical (IHC) markers CD 117, CD 34, S100, anaplastic lymphoma kinase-1 (ALK-1), and B-catenin were negative.

The patient came for a follow-up at 1, 6, and 12 months. After 1 year, the patient was asymptomatic with no evidence of recurrence of the tumor.

DISCUSSION

Mesenchymal tumors arising in the abdominal cavity represent a heterogeneous group of entities that are a diagnostic challenge

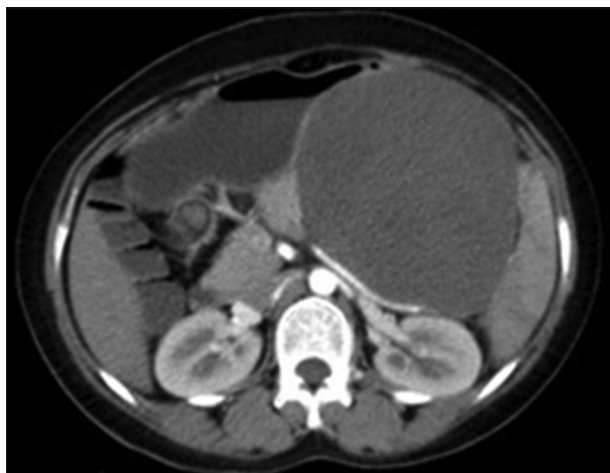


Figure 1: Axial computed tomography image with contrast showing well-defined heterogeneously enhancing soft tissue hypodense mass measuring 14.6 cm × 12.4 cm × 13.2 cm in the left retroperitoneum with splenic artery coursing through the lesion

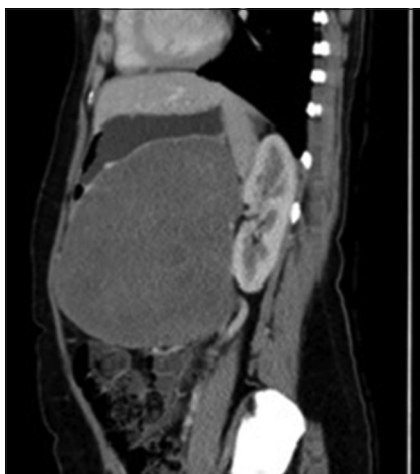


Figure 2: Sagittal computed tomography image showing the mass to be extending from the D11-L4 vertebral level causing breaking of the pancreas

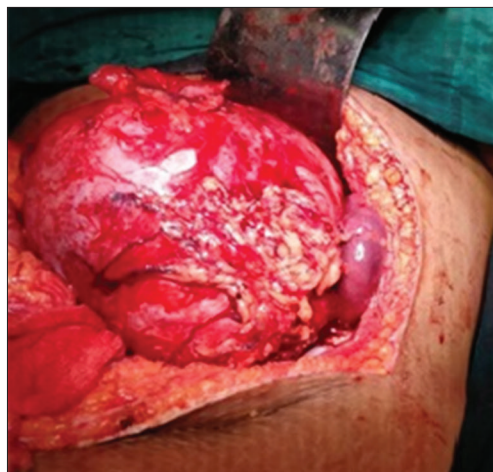


Figure 3: Intraoperative picture showing the mass being adherent to greater curvature of stomach, distal body, and tail of pancreas and spleen

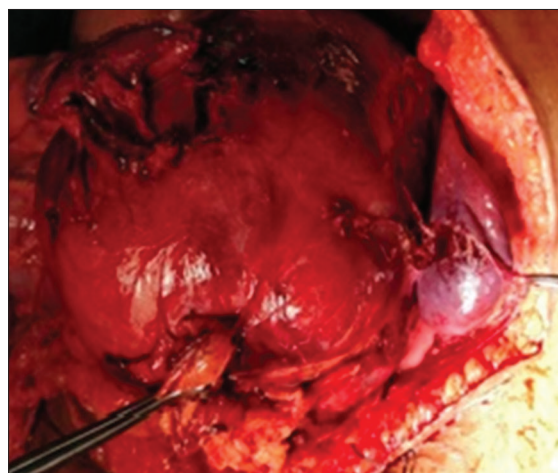


Figure 4: Splenic artery seen traversing through the tumor

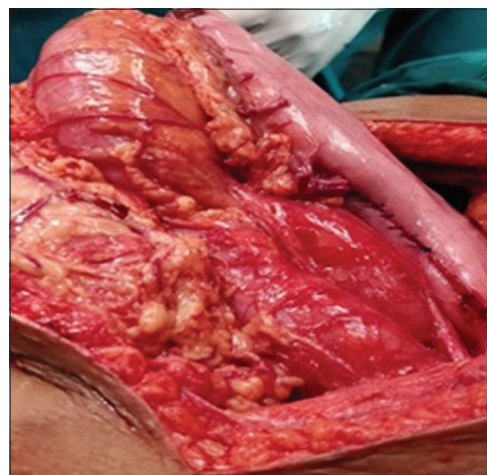


Figure 5: Intraoperative picture showing sleeve gastrectomy post in toto excision of tumor with splenectomy



Figure 6: Resected specimen of the tumor in toto with the spleen and the greater curvature of the stomach

both to surgeons and pathologists. Distinguishing between them is essential as their treatment and prognosis vary widely. RNFP is

one such mesenchymal tumor, first described by Yantiss *et al.*, in 2003, in a series of five cases arising in the gastrointestinal tract and mesentery [1]. Since 2003, around 20 such cases have been recognized. The target population of RNFP appears to be adults between the age group of 40–60 years, with a male preponderance

of the disease. However, this case is reported in a young female of 20 years.

The exact etiopathogenesis of RNFP remains unclear. It is considered a non-neoplastic lesion, related to the proliferation of multipotent subserosal cells as an overwhelming inflammatory response to an injury [2]. The various kinds of injury that has been proposed are prior abdominal surgery, endometriosis, ergotamine use, peptic ulcer disease, perforated diverticulitis, and foreign body ingestion [2]. In our case, we could not recognize any of these predisposing risk factors, implying that there could be more risk factors which are still obscure or it can be idiopathic.

Macroscopically, it may present as a tan-white, firm, well-circumscribed solitary tumor or multiple masses arising from the bowel wall or mesentery mimicking malignancy [3,4]. Microscopically, it is composed of a spindle to stellate myofibroblast embedded in a collagenous matrix with foci of hyalinization and lack of atypical or bizarre forms [1,5]. All cases of RNFP described in literature have stained positive for vimentin and most have stained positive for smooth muscle actin [6]. It may also stain positive for muscle-specific actin, CD117, and AE1/AE3 and stains negative for CD34, S100, and ALK-1.

Analyzing the clinical and morphological features of this case, the close differentials of RNFP are gastrointestinal stromal tumor (GIST), fibromatosis, and inflammatory myofibroblastic tumor (IMT). However, the immunohistochemistry study (IHC markers - CD 117, CD 34, S100, ALK-1, and B-catenin were negative) suggested that it is a result of another reactive process (RNFP) which helped us in arriving at the diagnosis of RNFP. One of the important differential diagnoses of RNFP is GIST as both show rapid clinical progression. In contrast to RNFP which originates in subserosa, GIST mostly arises from muscularis propria [7]. Grossly, GIST is more brown, fleshy with hemorrhage and necrosis, and histologically, it is more cellular than RNFP, being composed of spindle and/or epithelioid cells [8-10]. GIST is strongly positive for CD117 and approximately 70% also stains positive for CD34.

IMTs, unlike RNFP, are seen in children and younger adults with a high propensity of local recurrence and rarely distant metastases [11]. Histologically, they are hypercellular composed of spindle cells that stain positive for ALK-1 with abundant eosinophilic cytoplasm and some cytological atypia and mitoses [12]. The negativity of ALK-1 in RNFP helps in distinguishing it from IMT. Another mesenchymal lesion that needs to be distinguished from RNFP is fibromatosis. It is usually solitary, gray tanned, ill-defined mass with irregular borders, unlike RNFP which is well-circumscribed. It stains positive for B-catenin [13].

CONCLUSION

This case illustrates the need for keeping RNFP in mind as a differential diagnosis in case of a large abdominal mass. The exact etiopathogenesis remains unclear with various predisposing risk factors described. In this case, we could not recognize any of these risk factors, implying the possibility of other unidentified risk factors or it is idiopathic. It is curable with surgical resection with no known recurrence reported so far.

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