

# Inflammatory myofibroblastic tumors of the pancreas in children

## A case report and literature review

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### Abstract

**Introduction:** Inflammatory myofibroblastic tumors are an uncommon neoplasm, which are very rarely located in the pancreas. Clinically and radiologically, this rare pancreatic tumor presents as an abdominal mass lesion that mimics other pancreatic tumors, and should therefore be considered in the differential diagnosis of pancreatic tumors.

**Clinical Findings and diagnosis:** The 15-year-old boy complained of abdominal pain over the left upper quadrant with intermittent fever for 7 days. Abdominal sonography revealed one cystic lesion with a hyperechoic component in the left upper quadrant of the abdomen. Surgical excision was performed and postoperative findings indicated a pancreatic tail tumor. The pathology indicated inflammatory myofibroblastic tumors. To our knowledge, this patient is a unique case as the tumor was located in the pancreatic tail only, sparing the body.

**Interventions and outcomes:** The patient underwent tumor resection and segmental resection of the transverse colon with simple closure. The patient had no evidence of disease recurrence at 3 years follow-up.

**Conclusion:** Inflammatory myofibroblastic tumors of the pancreas in children are extremely rare. Surgical excision is the standard treatment, and corticosteroids use in children need more large-scale studies.

**Abbreviation:** IMT = inflammatory myofibroblastic tumor.

**Keywords:** case report, child, inflammatory pseudotumor, pancreas

## 1. Introduction

An inflammatory myofibroblastic tumor (IMT) is characterized histologically by proliferation of fibrous tissue with associated moderate or marked inflammation, also called inflammatory pseudotumors or plasma cell granulomas.<sup>[1]</sup> There are not many reports on pancreatic IMTs because of the rare nature of the condition, so the incidence is difficult to obtain. The most common gastrointestinal tract symptoms of this tumor are mass, pain, fever, weight loss, and malaise.<sup>[1]</sup> Diagnosis is based on the histological findings, which consists of variable quantities of plasma cells, lymphocytes, eosinophils, foamy histiocytes, and mast cells with an inflammatory component. IMTs of the

pancreas exhibit slow growth and do not undergo malignant transformation or exhibit distant metastasis, and most are surgically excised.<sup>[2]</sup> We report a 15-year-old male patient with an IMT and a literature review regarding the differential diagnostic, histopathological, and therapeutic features of this condition.

## 2. Case report

A 15-year-old boy was admitted to E-Da Hospital, Kaohsiung city, Taiwan, with an intraabdominal tumor. He complained of abdominal pain over the left upper quadrant with intermittent fever for 7 days. The results of laboratory examinations were as follows: white blood cell count,  $15,640 \times 10^6$  cells/L; neutrophils, 79.9%; hemoglobin, 9.7 g/dL (or 1.50 mmol/L); hematocrit, 31.1%; platelet count,  $432 \times 10^9$ /L; glucose, 93 mg/dL; alanine transaminase, 14 U/L (normal range 0–44 U/L); aspartate transaminase, 21 U/L (normal range 0–38 U/L); carcinoembryonic antigen (CEA), 1.0 ng/mL (normal <5.0 ng/mL); and carbohydrate antigen 19–9 (CA 19–9), 3.0 U/mL (normal <37.0 U/mL). Abdominal sonography revealed one 5.9 cm cystic lesion with a 3.7 cm hyperechoic component in the left upper quadrant of the abdomen (Fig. 1).

An ultrasound-guided aspiration biopsy was performed, with aspiration cytology showing numerous neutrophils and few spindle cells, and immunohistochemical studies of the biopsy specimen indicating positivity for vimentin and muscle actin antibody (HHF-35). These results indicate a tumor of a fibroblastic and muscular origin, respectively (Fig. 2).

Two weeks later, postoperative findings indicated a pancreatic tail tumor, about 5 cm × 5 cm × 4.3 cm in size, with adhesion and invasion of the transverse colon. Segmental resection of the

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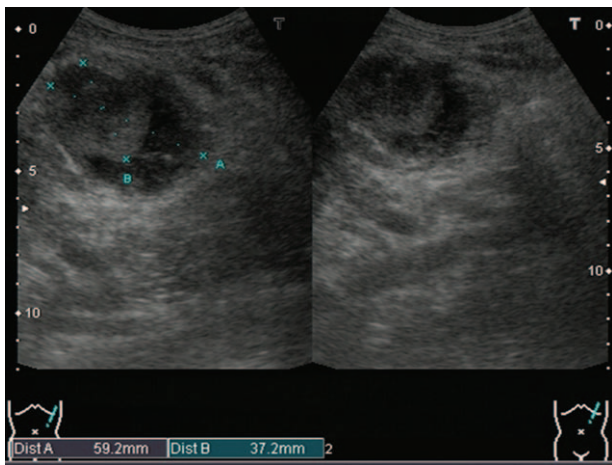
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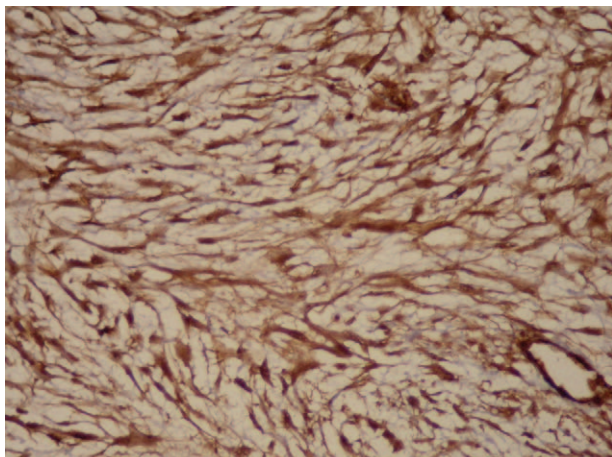


**Figure 1.** Abdominal ultrasound reveals one 5.9 cm cystic lesion with a 3.7 cm hyperechoic component in the left-upper quadrant of the abdomen.

transverse colon with simple closure and tumor resection were performed. The pathology of the specimen revealed a spindle cell tumor with myogenic differentiation. External to the main tumor mass, there was an inflammatory lesion with aggregates of foamy histiocytes. Immunohistochemical studies showed spindle cells that stained positive for actin, thereby matching the histopathological features of the biopsy specimen. Based on these findings, a diagnosis of IMT was made. The patient had an uneventful hospital course, and was discharged 6 days after the operation. After surgery, the patient had complete resolution of his abdominal pain, and there was no evidence of disease recurrence at 3 years follow-up.

### 3. Discussion

IMTs, also known as inflammatory pseudotumors or plasma cell granulomas, have been described in a wide variety of sites, including the orbit, head and neck region, salivary glands, lung and liver, spleen, retrorectal space, soft tissues, and peripheral nerves.<sup>[1,3–10]</sup> The mass may compress adjacent structures, leading to a variety of symptoms, depending on the organs



**Figure 2.** Immunohistochemical studies show positivity for muscle actin antibody. (400×).

involved, such as local pain, malaise, loss of appetite, wasting, and subfebrile elevation of temperature.<sup>[1]</sup> Histologically, IMT is composed of myofibroblastic spindle cells and inflammatory components of variable quantities of plasma cells, lymphocytes, eosinophils, foamy histiocytes, and mast cells.<sup>[2]</sup> The definitive etiology of IMT is not clear; however, some possible causes such as genetic predisposition and infection have been proposed.<sup>[11–13]</sup> Anaplastic lymphoma kinase and p80 expression as well as chromosomal rearrangements involving 2p23 have been reported to be related to IMT.<sup>[12,14]</sup> The differential diagnoses of IMC include low grade myofibroblastic sarcomas and some benign, neoplastic spindle cell lesions, such as leiomyoma and solitary fibrous tumor.<sup>[15]</sup> The concept of IMT being a benign reactive lesion is doubtful owing to its high recurrence (as high as 37%), the presence of regional metastases, and the evidence of acquired clonal chromosomal abnormality. However, the issue of reactive or neoplastic pathogenesis of this lesion remains unsolved.<sup>[16]</sup> An IMT of the pancreas is not common, and clinically and radiologically, this rare pancreatic myofibroblastic tumor presents as an abdominal mass lesion that mimics a malignancy such as pancreatoblastoma, solid-pseudopapillary tumor, or insulinoma.

Pancreatoblastoma, the most common pancreatic neoplasm in young children, should be considered in the differential diagnosis. On computed tomography scan, pancreatoblastomas are heterogeneous and often multilocular with hyperechoic and enhancing septa, as opposed to IMTs, which usually appear as homogenous and well-defined solid masses.<sup>[17]</sup>

Solid-pseudopapillary tumors, which are slow-growing tumors usually located in the pancreatic tail, are often large and encapsulated or circumscribed, with marked degenerative and hemorrhagic components. Solid-pseudopapillary tumors are most commonly diagnosed in adolescent girls and young women (83%–98.5%), especially in blacks and East Asians.<sup>[17]</sup> On ultrasonography, the tumors are usually visualized as an echogenic cyst.

Islet cell tumors are either insulinomas or gastrinomas. Insulinomas are composed of beta-cells and cause fasting hyperinsulinemic hypoglycemia, and are most common in the body and tail of the pancreas (65%). In contrast, the vast majority of gastrinomas involve the head of the pancreas (71%).<sup>[17]</sup>

Evidence supporting an autoimmune etiology includes the association of IMTs with other autoimmune diseases such as Sjögren syndrome and idiopathic thrombocytopenic purpura.<sup>[18,19]</sup> Nevertheless, there is no evidence that IMTs are associated with a preceding inflammatory or traumatic process.<sup>[20]</sup>

IMTs of the pancreas exhibit slow growth and do not undergo malignant transformation or exhibit distant metastasis.<sup>[3]</sup> Theoretically, resection of the mass lesion would lead to the prompt resolution of symptoms. However, this rare pancreatic lesion may recur in some instances, such as after incomplete resection. One adult patient was reported to experience local recurrence of a pancreatic myofibroblastic tumor about 1 year after the Whipple procedure.<sup>[20]</sup>

IMTs of the pancreas are extremely rare in children, and only 11 cases have been reported in the literature (Table 1).<sup>[2,21–28]</sup> Most of the reported pediatric cases were girls (8/11, 73%), with only 3 boys (3/11, 27%). Our patient is therefore the 4th reported male child to have this kind of pancreatic tumor. There was no predominant location of the pediatric pancreatic IMTs in the literature review (head: 7 cases, body with or without the tail: 4 cases). To the best of our knowledge, this boy is a unique case as

**Table 1****Literature review of published cases of inflammatory myofibroblastic tumor of the pancreas.**

Age, years	Gender	Symptoms/signs	Location	Size	Operative procedures	Follow-up	Author
2.5	Female	Fever, anemia, abdominal mass	Body	Not available	Distal pancreatectomy	Not available	Scott et al <sup>[21]</sup>
4	Female	Malaise, lethargy, a vesicular skin rash	Head	3 × 3 cm	Whipple procedure	Disease-free at 48 months	Slavotinek et al <sup>[22]</sup>
5	Female	Vomiting	Head	Not available	Whipple procedure	Disease-free at 9 months	Stringer et al <sup>[23]</sup>
8	Female	Abdominal pain and mass	Body and tail	10.7 × 9.9 × 9.4 cm	Distal pancreatectomy	Disease-free at 24 months	Shankar et al <sup>[24]</sup>
8	Female	Jaundice, anemia, weight loss	Head	Not available	Whipple procedure	Disease-free at 12 months	Uzoaru et al <sup>[25]</sup>
11	Female	Jaundice, pruritis, decreased appetite, weight loss, epigastric pain, tea-colored urine	Head	3.2 × 1.6 × 3.4 cm	Whipple procedure	Not available	McClain et al <sup>[2]</sup>
12	Female	Abdominal mass	Body	12 × 10 × 6 cm	Pancreatectomy, unspecified	Not available	Abrebanel et al <sup>[27]</sup>
13	Female	Jaundice, Vomiting, anorexia, weight loss	Head	4 × 3 × 2.5 cm	Whipple procedure	Disease-free at 6 years	Dagash et al <sup>[25]</sup>
11	Male	Abdominal mass, lethargy	Body and tail	10 × 10 × 7 cm	Distal pancreatectomy	Disease-free at 36 months	Morris-Stiff et al <sup>[26]</sup>
10	Male	Abdominal pain, jaundice, anorexia	Head	2.2 cm	Medical treatment (Prednisolone and prophylactic cefuroxime)	Disease-free at 6 years	Dagash et al <sup>[25]</sup>
0.5	Male	Jaundice and pruritus	Head to uncinuate process	3.7 cm	Whipple procedure	Disease-free at 3.5 years	Ales Tomazic et al <sup>[28]</sup>
15	Male	Abdominal pain, fever	Tail	5 × 5 × 4.3 cm	Distal pancreatectomy	Disease-free at 3 years	This article

the tumor was located in the pancreatic tail only, sparing the body. Compared to pediatric patients, in almost all adult cases, the mass lesions have been found in the pancreatic head.<sup>[29,30]</sup> In addition, unlike in other pediatric patients, the tumor in this patient invaded the transverse colon.

Most patients encountering IMTs of the pancreas undergo surgical excision. Since it is a benign lesion, the administration of corticosteroids instead of an operation has been reported in 4 adults and 1 child (tumor size ranging from 2.2–4 cm), and all 5 cases showed remission or regression of the tumors.<sup>[25,31,32]</sup> Although there is only 1 reported case of treatment with corticosteroids in a child to date, such treatment could be considered in the future.

#### 4. Conclusion

In summary, IMT is an uncommon mass rarely located in the pancreas, with only 11 cases reported in the literature.<sup>[2,21–28]</sup> However, it must be included in the differential diagnosis, along with other pancreatic tumors. Most patients encountering IMTs of the pancreas undergo surgical excision, and only 4 adults and 1 child have been reported to receive medical treatment with corticosteroids. Such treatment should consider in pediatric cases; however, more studies are needed to assess corticosteroids as a treatment modality where complete surgical resection cannot be performed.

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