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CASE REPORT

Pancreatic Pseudopapillary Tumor in a Male Child

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ABSTRACT

Context Solid-pseudopapillary tumors are exceedingly rare in males. They are almost exclusively encountered in young females (mean age 26 years) and have a female predominance. It is most commonly detected incidentally, but may occasionally present with sudden pain or symptoms related to compression of adjacent organs.

Case report We report the case of a 12-year-old boy having a solid-pseudopapillary tumor of the pancreas presenting with a tender upper abdominal mass following a slight trauma. Radiological investigation showed the lesion to be a cystic mass arising from the body and the tail of the pancreas. The child underwent emergency distal pancreatectomy and has remained free of recurrence for 3 years.

Conclusion In the pediatric age group, solid-pseudopapillary tumors may present acutely with a tender abdominal mass following a slight trauma. Awareness of this fact will allow appropriate and prompt management to be undertaken.

INTRODUCTION

Solid-pseudopapillary tumors of the pancreas account for less than 4% of pancreatic cystic tumors [1]. They are composed of homogeneous, fleshy tissue separated by areas of hemorrhagic and necrotic cyst degeneration [2] and are believed to have an acinar origin [2]. Many tumors are detected incidentally, but may occasionally present with sudden pain due to bleeding within the tumor or with symptoms related to the compression of adjacent organs [3]. It is almost exclusively encountered in young females having a mean age of 26 years and has a male to female ratio of 1:9 [3, 4]. Therefore, solid-pseudopapillary tumors are exceedingly rare in males and only a few cases have been reported in children [5, 6, 7, 8, 9, 10, 11, 12, 13, 14, 15, 16, 17, 18]. We report a case of a young boy presenting with this rare pancreatic tumor and discuss the diagnostic dilemma and the malignant potential of such tumors.

CASE REPORT

A 12-year-old Saudi boy presented with a 2-day history of left upper quadrant pain, which started following a slight trauma to his abdomen and was associated with nausea and vomiting. Initially, he denied any history of trauma and there was no history of weight loss or preexisting abdominal swellings. There was neither history of sickle cell disease nor any other hemolytic anemias. On examination, he looked ill, dehydrated and pale but there was no jaundice or lymphadenopathy. His vital signs were stable, and chest and cardiovascular systems were normal. Abdominal examination revealed a

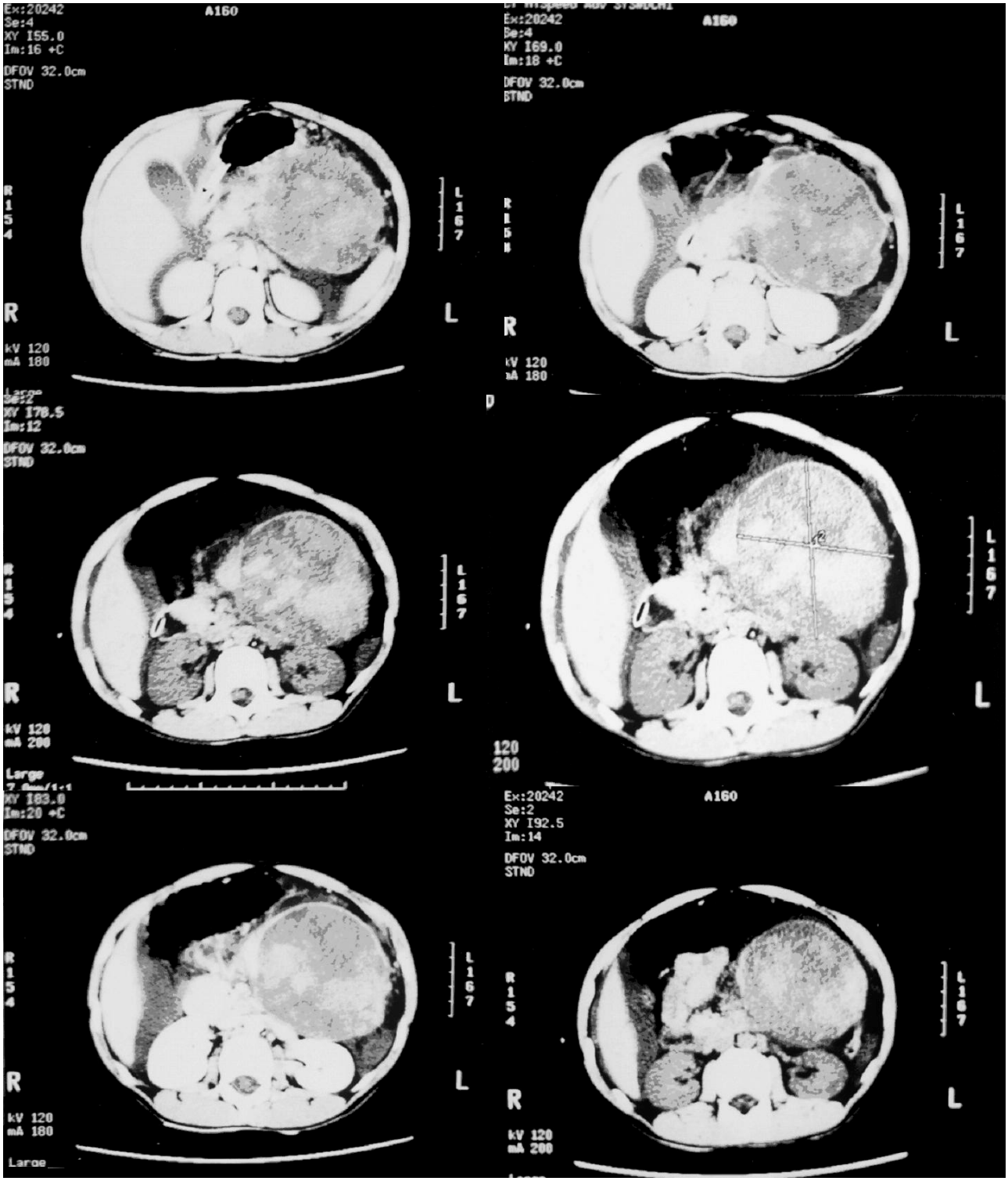

Figure 1. The ACT scan of the abdomen showing the cystic lesion arising from the tail of the pancreas with a normal pancreatic parenchyma.

tender abdominal mass occupying the epigastrium and left hypochondrium, with guarding and rigidity. Blood investigations revealed a hemoglobin of 9.6 g/dL (reference range: 12-16 g/dL), a leukocytosis of 13.6 $x10^9$/L (reference range: 3.5-10.0 $x10^9$/L) and normal amylase, lipase and liver function tests. Emergency abdominal ultrasound and computerized tomography (CT) scans revealed a mixed attenuated round mass (9.2x9.3 cm) at the body and the tail of the pancreas, possibly a complicated pseudocyst with a normal liver and spleen (Figure 1). Soon after admission, he became pyrexial

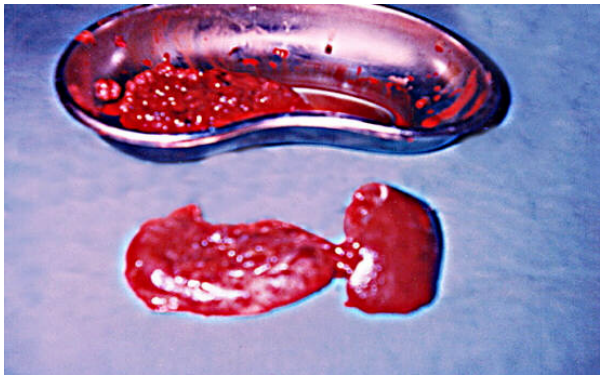

Figure 2. A macrograph of the resected operative specimen showing the tumor at the tail of the pancreas and the spleen. The hemorrhagic contents are also shown.

with persistent tenderness and rigidity. Repeated investigation revealed a leukocytosis of 30,000 mm-^{3} and a normal amylase level. An emergency laparotomy was performed using a roof-top incision. There was free blood in the peritoneal cavity and a huge retroperitoneal mass arising from the body and the tail of the pancreas with bleeding coming from a small laceration in its wall. Distal pancreatectomy and splenectomy were performed (Figure 2). His postoperative recovery was uneventful and he was given pneumococcal and *Haemophilus influenzae* vaccines before he was discharged a week later. The histopathologic examination revealed a solid-pseudopapillary tumor of the pancreas (Figures 3 and 4). He has remained well after a 3-year follow-up with no evidence of recurrence on repeated abdominal CT scan.

DISCUSSION

Solid-pseudopapillary tumors of the pancreas are very rare and almost exclusively encountered in young females (mean age 26 years) as genetic and hormonal factors may play an important role in their development [3, 4, 18, 19]. The tumor is exceedingly rare in males [3, 20, 21, 22]. In one review, there was only one male among 31 patients [3]. Review of the literature revealed some 24 cases of solid-pseudopapillary tumors of the pancreas reported in children (Table 1) with an average age of 10.8 years (range 8-16 years) and a male:female ratio of 1:4.75 [5, 6, 7, 8, 9, 10, 11, 12, 13, 14, 15, 16, 17, 18]. There were 4 cases (3 girls and a boy) 13-16 years of age presented acutely following blunt abdominal trauma in a fashion similar to that of our present case [5, 11, 14, 15] An emergency Whipple procedure was performed in 3 cases [11, 13, 15]. Bombi *et al*. reported another 2 cases of pseudopapillary tumors in older female patients 22 and 23 years of age [2]; one presented with an acute abdomen and pneumoperitoneum. This presentation was also somewhat similar to that of our patient whose tumor was the result of a slight trauma which had already been forgotten and was initially denied causing a diagnostic dilemma. Patients are often asymptomatic and the cyst is discovered incidentally on physical or radiological examination [3]. Patients may also occasionally present with an increasing abdominal mass associated with vague abdominal discomfort or may rarely present with an acute abdomen due to tumor rupture and hemoperitoneum as happened in our patient. Jung *et al*. reported a series of 6 pediatric cases (4 girls and 2 boys) with a mean age of 11.2 years (range 8-13 years); 5 of the lesions were located in the head

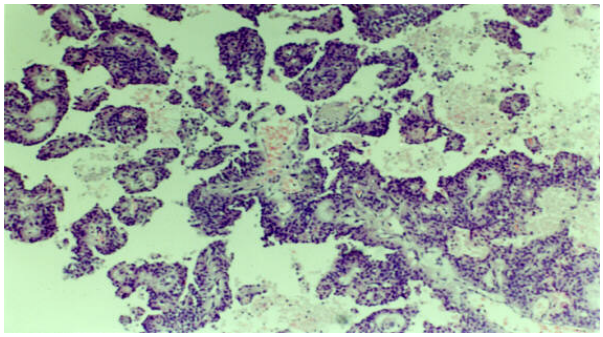

Figure 3. A micrograph showing cystic degeneration with solid and pseudopapillary formations and red blood cells in the cystic space. (H&E stain. x100)

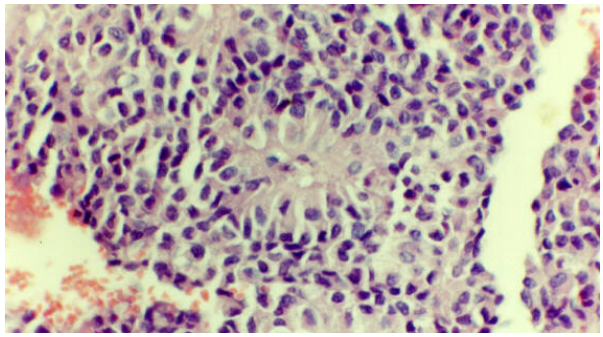

Figure 4. Tumor cells radially arranged around fibro-vascular stalks forming rosette-like patterns. (H&E stain. x400)

Table 1. Reported cases of solid pseudopapillary tumors of the pancreas in children.

Reference	Year	Number of cases	Sex	Age (years)
Persson *et al*. [5]	1996	1	Girl	16
Wang *et al*. [6]	1998	3	1 boy, 2 girls	10, 11, 14
Herskovits *et al*. [7]	1999	1	Boy	13
Jung *et al*. [8]	1999	6	2 boys, 4 girls	8-13
Rebhandl *et al*. [9]	2001	4	Girls	12-16
Akiyama *et al*. [10]	2002	1	Girl	15
Cervantes-Monteil *et al*. [11]	2002	1	Girl	15
Sabatino *et al*. [12]	2003	1	Girl	13
Carrincaburu *et al*. [13]	2003	1	Girl	9
Portc *et al*. [14]	2003	1	Boy	14
Jiang etal [15]	2003	1	Girl	13
Andronikou *et al*. [16]	2003	1	Girl	15
Saw *et al*. [17]	2004	1	Girl	12
Bardales *et al*. [18]	2004	1	Girl	13
Total		**24**	**5 boys, 19 girls (Ratio 1:4.75)**	**Average: 10.8 years**

necessitating pancreaticoduodenectomy and one was located in the tail which was treated by distal pancreatectomy [8]. All were alive with no recurrence at an average follow-up of 5.5 years. Wang *et al*. reported a solid-pseudopapillary tumor in 3 children (2 girls 11 and 14 years of age and a boy 10 years of age) and concluded that the tumor arises early in life, grows slowly and rarely metastasizes [6]. Another presentation of a solid-pseudopapillary tumor is acute pancreatitis with an abdominal mass [23]. Rebhandl *et al*. reported the cases of 4 girls 12-16 years of age presenting with abdominal pain and masses (diameter 7-15 cm); located in the tail (n=2), the body and tail (n=1) and the head (n=1). Only one patient developed two recurrences and metastases after surgical resection despite receiving chemotherapy [9].

In our case, a CT scan raised the possibility of a complicated pancreatic pseudocyst (either infected or bleeding within), but amylase and lipase levels were within normal limits. Furthermore, the rest of the pancreas looked normal with no evidence of pancreatitis in the CT scan. It was noted that magnetic resonance imaging (MRI) is superior to CT for diagnosing these tumors [20, 24, 25]. An MRI was not requested in this case but, at laparotomy, the mass had a mature thick true wall, easily separable from the posterior wall of the stomach and was therefore thought to be a cystadenoma or a cystadenocarcinoma rather than a pseudocyst of the pancreas. Drainage of this cystic tumor in the stomach or jejunum would have resulted in disastrous consequences of local invasion and possible future metastases. The option of distal pancreatectomy and splenectomy offered a complete cure and settled this diagnostic dilemma. This procedure can be conducted laparoscopically and laparoscopic spleen-preserving distal pancreatectomy for solid-pseudopapillary tumor has been reported [13].

The role of endoscopic ultrasound-guided fine needle aspiration in accurately diagnosing solid-pseudopapillary tumors is now well-established [18, 26, 27, 28, 29]. This usually demonstrates low levels of carcinoembryonic antigen and a moderate elevation in cyst fluid carbohydrate antigen 19-9 and lipase and the cyst fluid cytology may be diagnostic [29]. Extensive necrosis and rare mitotic figures may be present. Solid-pseudopapillary tumors of the pancreas show strong cellular immunoreactivity for vimentin and focal weak keratin reactivity. Neuron-specific enolase, alpha1-antitrypsin, and alpha1-antichymotrypsin stains, if carried out, may be strongly positive [18, 29]. US-guided FNA

was not carried out in our case due to the acute presentation.

Solid-pseudopapillary tumors possess a malignant potential risk of 5-10% and must therefore be resected completely and aggressively as there are no prognostic factors to distinguish between pseudopapillary tumors with or without malignant potential [3, 30]. Unlike pancreatic ductal adenocarcinoma, surgical resection often results in cure and long disease-free periods even in patients who have recurrences or metastases [31]. One series reported a 100% survival after an average 10-year follow-up [32]. However, anything short of surgical resection (e.g. internal or external drainage) is associated with tumor progression locally and invasion of the surrounding structures and distant metastases [33]. Even in the presence of advanced local invasion, palliative resection is advised and offers an excellent prognosis and survival benefits [4, 31, 34]. After resection, only a small number recur or develop metastases. However, subsequent visceral metastases after incomplete resection of a pseudopapillary tumor following a prolonged period of observation have been reported [4, 16, 35, 36]. Nevertheless, the growth of a recurrent tumor is very slow. Our patient has had a relatively short follow-up period (just over 3 years); until now there has been no evidence of recurrence or distant metastases. It seems that tumors arising in children are low grade, grow very slowly, rarely metastasize and have a good prognosis [6, 8]. This low-grade malignant potential manifests itself by invasion of the capsule and neighboring structures [3]. Macroscopically, they are well-circumscribed tumors which contain solid and cystic areas consisting of hemorrhagic and central cystic necrosis. This often gives a characteristic CT appearance which aids diagnosis and allows differentiation from islet cell tumors [32, 37]. Our case was erroneously diagnosed as a complicated pseudocyst based on CT scan findings.

In conclusion, this case report emphasizes the fact that solid-pseudopapillary tumors of the pancreas may arise in male children, and that it may cause diagnostic confusion especially in children with asymptomatic lesions who may present acutely following trauma. Increased awareness of this tumor allows appropriate emergency management to be undertaken.

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