

## Tumor of the Pancreas in Children: Pseudopapillary Pancreatic Tumor in Children: A Report of Two Cases

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### Abstract

### Case Report

**Introduction:** The solid pseudopapillary tumor of the pancreas is a tumor very rarely seen in pediatrics. It primarily affects young women and accounts for 2–3% of primary pancreatic tumors. It has a low potential for malignancy and has a good prognosis when completely resected. **Observations:** We report two cases of solid pseudopapillary tumors operated on in 2019 and 2024 in the Pediatric Visceral Surgery Department of the Abderrahim HAROUCHI Mother-Child Hospital, IBN ROCHD University Hospital, Casablanca. We analyzed the mode of presentation, the initial clinical examination, paraclinical tests, as well as the type of surgical procedure and postoperative course. **Case 1:** A 12-year-old girl was admitted with acute abdominal pain while her general condition remained stable. The clinical examination revealed isolated tenderness in the epigastrium without a palpable mass. Ultrasound combined with CT scanning revealed a tissue mass in the tail and body measuring 10.4 x 8.3 x 6.8 cm. Laboratory tests showed elevated amylase and lipase levels. An ultrasound-guided biopsy was performed, with histopathological findings consistent with a solid pseudopapillary neoplasm of the pancreas. A corpus-caudal pancreatectomy with spleen preservation was performed, with a clear margin of resection as confirmed by histopathological examination of the surgical specimen. The postoperative course was uneventful. **Case 2:** A 14-year-old girl was admitted with acute abdominal pain associated with abdominal distension, occurring in the context of afebrile status and preserved general condition. Physical examination revealed a hard, fixed, painful abdominal mass straddling the epigastrium and right hypochondrium. An abdominal CT scan revealed a 99x76x95 mm tissue mass at the head of the pancreas with two cystic formations in the body and tail, associated with dilation of the Wirsung duct. Serum amylase and lipase levels were elevated. An ultrasound-guided biopsy was performed. Pathology concluded that the lesion was a solid pseudopapillary neoplasm of the pancreas. A cephalic duodenopancreatectomy was therefore indicated and performed with a healthy resection margin, as confirmed by the pathological examination of the surgical specimen. The postoperative course was uneventful; currently, at 2 months post-op, normal eating has resumed and is well tolerated. The two patients were referred to the cancer treatment department for regular and specialized follow-up. Both girls were referred to a pediatric endocrinologist for follow-up to screen for possible diabetes. **Discussion:** The solid pseudopapillary tumor of the pancreas is a rare tumor, with fewer than 500 cases reported in the literature. The average age at diagnosis is between 20 and 40 years in 90% of cases, with a clear female predominance; a few cases have been described in the literature in adolescents. Indeed, in our study, the patients involved were two girls aged 12 and 14, respectively. The clinical signs of the tumor are nonspecific; it is often revealed by isolated suprapubic abdominal pain or in association with an abdominal mass, which was the case in one of our patients. Sometimes, it is discovered incidentally or due to a complication. Ultrasound, CT, and MRI constitute the arsenal of radiological diagnostic tools. The tumor presents as solid or mixed. Radiology will help determine the tumor's location and relationships to surrounding structures. Preoperative biopsy is not always indicated and should only be performed when the radiological presentation is atypical. Treatment is exclusively surgical and must consist of a partial or even total pancreatectomy, depending on the tumor's location. Resection should be as extensive as possible to minimize the risk of tumor recurrence. Simple tumorectomy, even if technically feasible, can lead to recurrence and metastasis. The overall prognosis is good with complete resection, with an estimated 5-year survival rate of 95%. Oncological medical monitoring is essential to prevent any adverse event. **Conclusion:** Pseudopapillary solid pancreatic tumor is a rare tumor in pediatrics, with varied and nonspecific clinical presentations. Imaging allows for

diagnosis, and histopathological examination confirms the tumorous nature. The only treatment is complete surgical resection. Long-term follow-up is required due to the risk of recurrence and diabetes.

**Keywords:** Pituitary Stalk Interruption Syndrome, Autoimmune Polyendocrinopathy, Posterior Pituitary Ectopia, MRI, Hypogonadotropic Hypogonadism.

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## INTRODUCTION

- Pancreatic tumors are quite rare in children, causing less than 0.2% of malignant pediatric children deaths [1].
- In general, pancreatic tumors in children have a different histologic spectrum and better clinical outcome compared to those in adults [2].
- It has a low potential for malignancy and has a good prognosis when completely resected.
- All pancreatic neoplasms in children are capable of producing metastases, usually to the liver and lymph nodes [3].
- On the whole, these tumors have a better clinical outcome than most pancreatic tumors in adults. Knowledge of the differential diagnosis of pancreatic masses in children and their relatively good prognosis may promote correct preoperative diagnosis and appropriate treatment [1].

## OBERVATIONS

We report two cases of solid pseudopapillary tumors operated on in 2019 and 2024 in the Pediatric Visceral Surgery Department of the Abderrahim HAROUCHI Mother-Child Hospital, IBN ROCHD University Hospital, Casablanca. We analyzed the mode of presentation, the initial clinical examination, paraclinical tests, as well as the type of surgical procedure and postoperative course.

### Case 1:

The case of a 12 years old girl, with no particular pathological history, initially admitted to our hospital structure, for diffuse abdominal pain evolving 2 months with preservation of the general state.

The patient underwent a couple of morphological exams: abdominal ultrasound and abdominal computed tomography, which concluded that is a tissue mass, tail and body of the pancreas 10.4x 8.3 x 6.8 cm with bilateral pulmonary micronodules (less than 4 mm). Echo-guided biopsy made it possible to make the diagnosis.

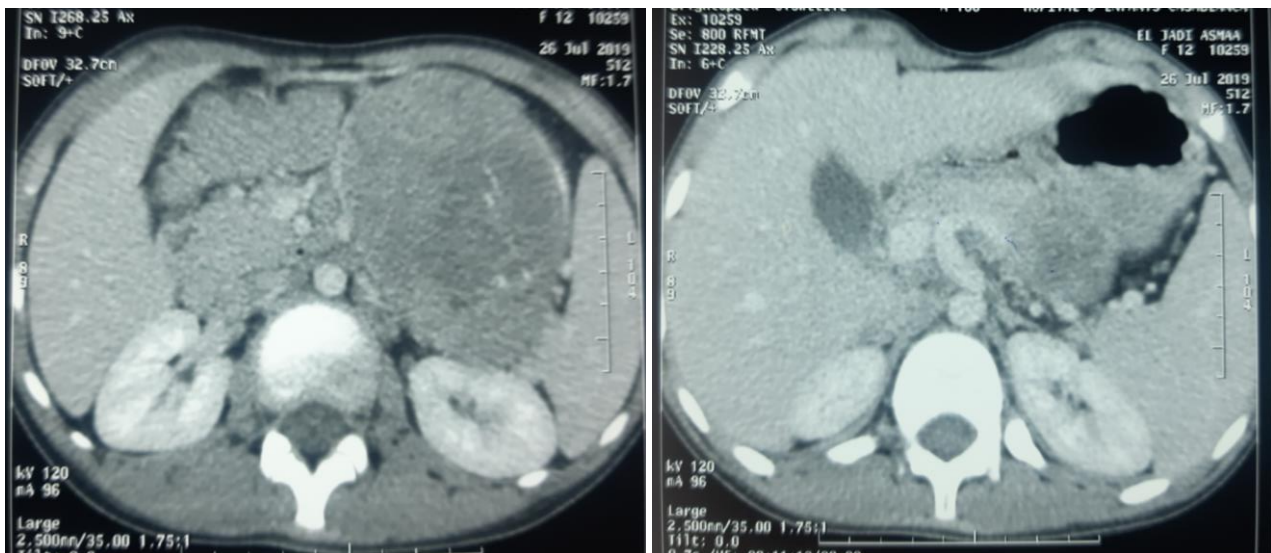


Figure 1: Initial Abdominal CT scan: a well-defined mass in the body and head of the pancreas.

After discussion with the multidisciplinary concertation staff, there was no indication for chemotherapy. We propose the surgical cure; we carried out a corporo-caudal pancreatectomy by laparotomy. The post-operative suites were simple.

The evolution was marked by a clear clinical and radiological improvement. The patient underwent

regular monitoring at the oncology clinic during the first month, followed by quarterly, six-monthly and annual check-ups, comprising an abdominal ultrasound and a clinical examination, with no abnormalities noted. An abdominal scan was carried out a year later and found no abnormalities.

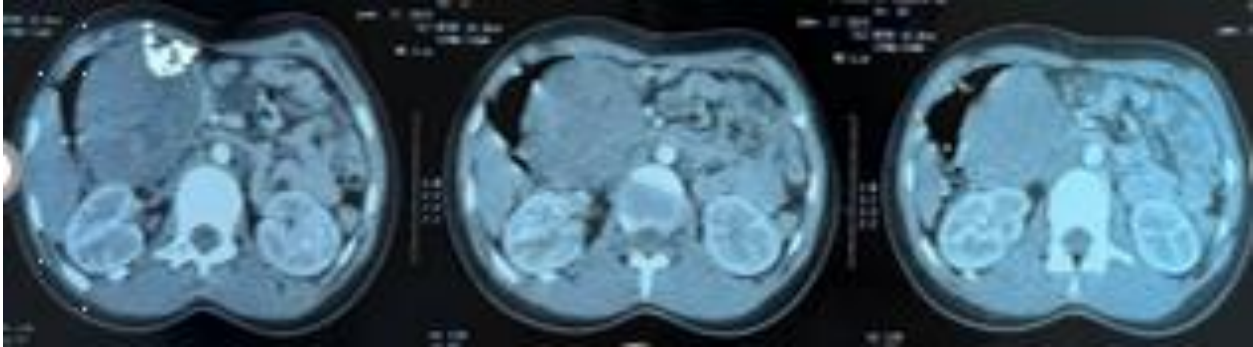
The patient received endocrinology follow-up, without any complications, respectively at 1 month, 3 months, 6 months.

#### Case 2:

A 14-year-old girl was admitted with acute abdominal pain associated with abdominal distension, occurring in the context of afebrile status and preserved general condition. Physical examination revealed a hard,

fixed, painful abdominal mass straddling the epigastrium and right hypochondrium.

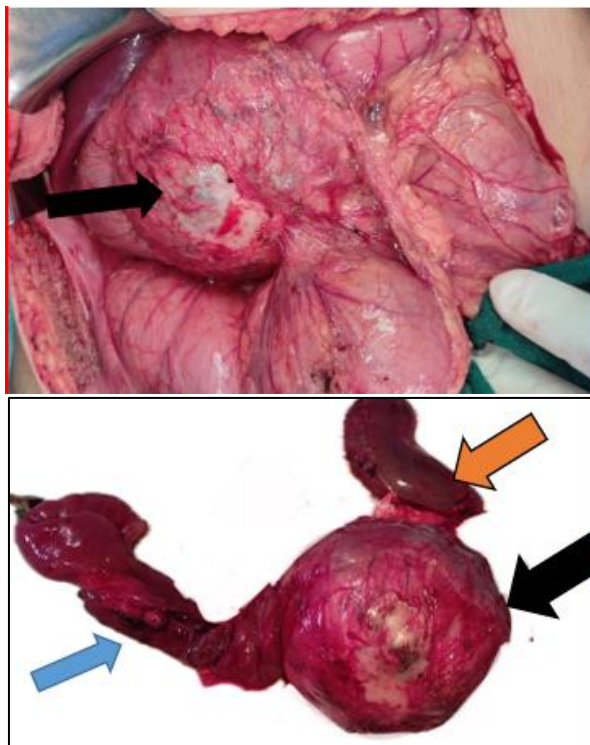
An abdominal CT scan revealed a 99x76x95 mm tissue mass at the head of the pancreas with two cystic formations in the body and tail, associated with dilation of the Wirsung duct. Serum amylase and lipase levels were elevated.



**Figure 2: Initial Abdominal CT scan: a well-defined mass in the head of the pancreas**

An ultrasound-guided biopsy was performed. Pathology concluded that the lesion was a solid pseudopapillary neoplasm of the pancreas.

A cephalic duodenopancreatectomy was therefore indicated and performed with a healthy resection margin, as confirmed by the pathological examination of the surgical specimen.



The patient underwent regular monitoring at the oncology clinic during the first month, followed by quarterly, six-monthly and annual check-ups, comprising an abdominal ultrasound and a clinical examination, with no abnormalities noted. An abdominal scan was carried out a year later and found no abnormalities.

## DISCUSSION

- Solid pseudopapillary tumors and insulinomas are the most common type of pancreatic tumors in children with the most literature available reviewing surgical methodology. Surgery

remains the treatment for pediatric pancreatic tumors [4].

- Solid Pseudopapillary Tumors (SPT) were first described in 1959 by Frantz and subsequently classified in 1996 by the WHO. These tumors were previously known as papillary cystic epithelial neoplasms, solid/ papillary neoplasms, papillary cystic tumors, and solid/ cystic tumors. Typically, SPT are found in young females in the second or third decade of life.
- Female to male ratio has been reported ranging from 2:1 to 10:1[5].
- SPT in children are typically found as incidental masses given the benign, indolent nature of these tumors. [5]
- Typical presenting symptom is abdominal pain [6]. Though rare, there have been reported cases of hemoperitoneum secondary to traumatic or spontaneous rupture of SPT in children [7].
- Multiple imaging modalities have been used to identify these lesions, including abdominal ultrasound (US), computed tomography (CT), and magnetic resonance imaging [7].
- There is general consensus that surgical resection offers the only cure. The type of pancreatic resection is dependent on location of the tumor. Because these patients are children there is the dual purpose of complete excision of the tumor with functional preservation of the pancreas if at all possible [8].
- Laparoscopic surgery is acceptable too [9], especially for benign lesions if negative margins can be achieved. Given the rarity of many pediatric pancreatic tumors, only a few case reports were available for review in many instances.

## CONCLUSION

- Solid-pseudopapillary tumor of the pancreas (SPT) is an exceptionally rare neoplasm in children. Its origin remains enigmatic.
- It is of low malignant potential and occurs most frequently in young females.
- It is mandatory to establish this diagnosis since complete surgical removal of the tumor even in case of metastases or local invasion offers an excellent prognosis.
- Individual countries have formed databases for these rare tumors with the purpose of elucidating presentation, diagnosis, and treatment of these rare tumors. An international collaborative would be ideal to capture potential missed cases.
- Finally, we would like to emphasise the importance of holding a multidisciplinary

consultation meeting for every paediatric tumour.

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