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Solid Psudopapillary Neoplasm of the Pancreas in 11 Years Old Saudi Girl: Case Report and Review of Literature

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Abstract: ***Solid psudopapillary neoplasm (SPN) of the pancreas is a rare neoplastic entity representing around 0.13-2.7% of pancreatic neoplasms [1]. In recent years, SPN has been reported more frequently and this might be attributed to increased awareness of this disease [2]. This entity was described in details in 1959 by Frantz but was first reported by Lichtenstein in 1933 [3,4]. This neoplasm has been designated with many names through out years, until it was official recognized as a distinctive entity by the World Health Organization (WHO) in 1996 and was given the name SPN [1]. Information about SPN is mostly case reports and small case series. The first published case of SPN was by Frantz in 1959 when he described " papillary cystic tumor of the pancreas ", in a 2 years old male patient, who died while they attempted to perform pancreaticodoudenectomy on him [3]. In 1970, Hamoudiet al added an additional patient to the literature where they ultrastructure the features of the tumor with electron microscopy which in turn led to its acceptance of SPN as separate clinicopathological entity [5]. Since 1953, more than 700 cases of SPN were reported, the majority of them were reported in the last 10 years [7]. This can be attributed to increase of awareness of the tumor behavior and its clinicopathological features. Herein, we aim to summarize our experience with diagnosis and the surgical treatment of SPN in a female child, as we were suspecting the tumor radiologically and confirmed intra-operatively by cytology.***

Keywords: Solid, pseudopapillary, pancreatic, neoplasm

1. Case Report

11 years old girl, medically and surgically free, presented to the emergency unit complaining of a sudden severe left upper quadrantpain. The pain was associated with one episode of vomiting. There was no diarrhea, constipation, jaundice or weight loss.

On examination there was mild tenderness in the left upper quadrant, with no distention, or a palpable mass. Initial enhanced CT scan was performed, which showed a well-circumscribed lesion in the pancreatic tail that measured approximately 8 cm in diameter. There were peripheral enhancement and a central area of cystic degeneration present(Figure1). Further evaluation by ultrasound demonstrated a large hypo/isoechoic rounded mass (10.5 X 6.8 X 9.6 cm) with central necrosis occupying the epigastrium and left hypochondrium, arising from the tail of the pancreas. (Figure 2). CT of the chest was unremarkable for any metastatic lesions.

The lab investigations, except for the ESR (48) and CRP (142), were within the normal values, including the amylase, lipase and LFT parameters. Serum assays of tumor markers (CEA, CA-19.9, AFP) were also within normal values, except for CA-125 (70.99), however, that is neither a specific nor a sensitive sign.Given the radiological findings, along with the age, gender and clinical presentation raised suspicion of SPN.

Distal pancreatectomy with splenectomy was planned and the tumor was resected by means of open surgery. Moreover, a segment of the transvers colon was resected, as the tumor was attached to it. A drain was placed at the pancreatic bed. The patient had an uneventful week after the surgery with no complications. The drain was removed on the fourth postoperative day, and the patient was discharged two days later. (Figure 3)

On gross examination an encapsulated tumor (9 X 7 X 5.5 cm) was distorting the distal pancreas (Figure 4) It had a solid cystic cut surface with extensive hemorrhage. It was 2cm away from the resection margin. The attachment to the transvers colon showed fibrous adhesions and hemorrhage, but no gross tumor extension. Histological appearance of Solid-pseudopapillary neoplasm was reported (Figure 5, The neoplasm shows numerous irregular pseudopapillae of varying sizes)
(Figure 6, heterogeneous growth pattern, with a combination of solid areas, typically located at the periphery, and pseudopapillary-microcystic structures with nercotic-hemorrhage degeneration at the center of the tumor and deposition of calcific material)

On the follow up appointment 3 months after the surgery, the patient had no new complaints and he remains asymptomatic with no evidence of local recurrence.

2. Discussion

SPN is a rare entity with low malignant potential, which is common entity in females more then males, with a female to male ratio of 9:1, and the presentation of the disease usually appears from the second to forth decades of life [6,7]. Moreover, this entity is rare in children and might represent only forth of pediatric pancreatic tumors, as reported by Lack and his colleagues [8]. Utilizing PubMed to search for reports of SPN cases in children, there has been a an increase in reporting due to increase awareness of the disease, but it is important to note that this entity is still underreported in literature in comparison with cases occurring in older age [7,9]. Reports of SPN in children in Saudi Arabia are lacking, only two cases by Meshikhes*et al*, and Mohammed *et al* reported such entity [9,10]. (Table 1) represents the reported cases in the last 10 years.

SPN occurs everywhere in pancreas put specially in the head and tail [7,11]. The cellular origin of this tumor is unknown, but it has been proposed that it might be originated from primordial cells, and lack differentiation of normal pancreatic tissue, ductal epithelial cells, or neuroendocrine origin [6,7]. It is important to note that since it is more common in females, the hormonal factors must be put in account [7]. The tumor appears grossly as an encapsulated, well-circumscribed, and clearly demarcated mass from pancreatic tissues [6,7]. The size of the tumor is usually larger in children in comparison with adults [12]. In cut section, it contains spongy areas with alternation between solid, cystic, and hemorrhagic components [6].

Clinical presentations of SPN are non-specific, with abdominal pain being the most common presenting symptoms, followed by enlarging mass in the right upper quadrant that is not associated with tenderness [6,7]. It can be detected incidentally, since it can be asymptomatic in some patients [7]. Imaging studies such as abdominal ultrasound (U/S) and computed tomography (CT), reveal a well-encapsulated mass, with areas of hemorrhagic, solid, and cystic components, with displacement of surrounding structures [11]. Magnetic resonance imaging (MRI) is considered as a superior imaging modality to U/S and CT, since it can aid in narrowing the differential diagnoses of a cystic lesions in the pancreas [11]. It is important to note that preoperative diagnosis of such entity is still difficult, even with technological advancement, since SPN share many similarities with other pancreatic cystic lesions such as: retention cysts, psudocysts, hydatid cysts, and cystic tumors including cystadenoma, cystadenocarcinoma, hemangioma [7]. In children involvement of pancreas is usually due to secondary malignancy rather then a primary one such as neuroblastoma, and lymphoma [6].

Biopsy remain the gold standard in diagnosing SPN, and it can be obtained by fine needle aspiration [FNA], or as preferred by many authors through endoscopic ultrasound guided biopsy [EUS] [6,7,9]. SPN is characterized by high activity with vimentin, α1-antitrypsin, α1-antichymotrypsin, and neuron specific enolase [6,11,12].

Once the diagnosis of SPN is confirmed, surgery would be indicated as a curative choice [6,7,11]. Location of the tumor determines the operation to be done; for lesions in the body or tail of pancreas distal pancreatectomy with or without splenic preservation can be performed [6,7,11]. Moreover, lesions occurring in the head necessitate performing pancreaticoduodenectomy either Wipple or Longmire procedures [6,7,11]. Chemotherapy can benefit certain patients as reported by many authors, and radiotherapy proved to benefit many patients but still lack evidence [6].

Prognosis of SPN is good, even with metastasis or local invasion, and surgery proved curative [6,7].

3. Conclusion

Our case demonstrates that SPN can occur in in younger age, and that physicians dealing with such cases, should have high degree of suspicion of SPN and include it in the differential diagnosis, when encountering cystic lesions by any imaging modality.

4. Authors' Contributions:

AH, AN, MH, MS, MS, and YM participated in the whole diagnostic and treatment process of the described patients. MH, MS, MS, and YM conceived the concept of the study. MH and MS collected the data. YM, MS wrote the article. YM, MS, MH, and AN participated in the sequence alignment, and helped to draft the manuscript. NS dealt with pathological aspects of the lesion. All authors read and approved the final manuscript.

5. Financial and Material Support

None in conjunction with this work

6. Conflicts of Interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

7. Ethical Approval

All procedures were in accordance with the ethical standards at our institution and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

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Figures

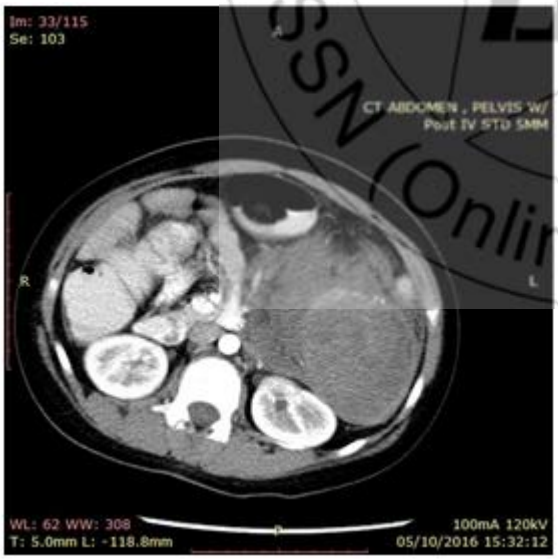

Figure 1

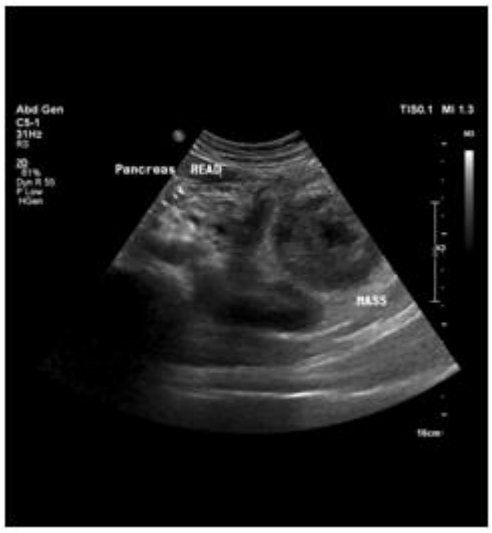

Figure 2

Figure 3

Figure 4

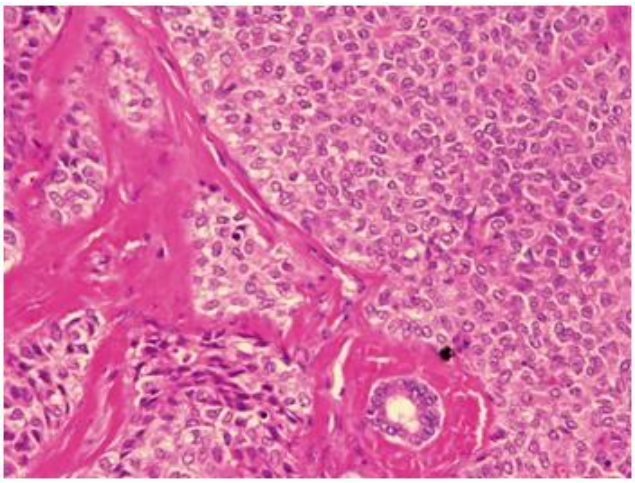

Figure 5

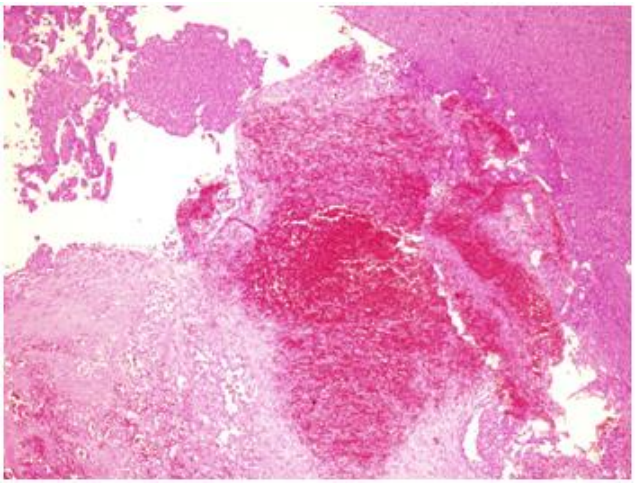

Figure 6

Tables

Table 1

No	Reference	Country	Year	No. Cases	Case description	Sex	Age (in years)	Presentation	Procedure type	Procedure	Location in pancreas	LOE (in days)
1	Meshikhes et al	Saudi Arabia	2004	1	Case 1	Male	12	Abdominal pain	Open	Distal panceartectomy and splenectomy	Body, tail	7
2	Al-Qahtani et al	Switzerland	2010	3	Case 1	Female	14	Abdominal swelling	Open	Pancreatecoduodenectomy and partial gastrectomy	Head	NR
					Case 2	Female	13.5	Abdominal swelling	Open	Pancreatecoduodenectomy	Head	NR
					Case 3	Female	11	N/V, abdominal pain	Open	Distal panceartectomy and splenectomy	Body, tail	NR
3	Arafah et al	Saudi Arabia	2010	1	Case 1	Female	16	Hypoglycemia	Laparoscopic	Distal panceartectomy and splenectomy	Tail	7
4	Crucitti et al	Italy	2010	1	Case 1	Female	15	Abdominal swelling	Open	Distal panceartectomy and splenectomy	Body, tail	13
5	Uchida et al	Japan	2010	1	Case 1	Female	12	Abdominal swelling	Laparoscopic	Spleen-preserving distal pancreatecomty	Body	15
6	Tsai et al	Taiwan	2011	1	Case 1	Female	10	Abdominal fullness	Laparoscopic	Pancreatecoduodenectomy	Head	4
7	Akhavan et al	Iran	2012	1	Case 1	Female	11	Abdominal swelling	Open	Distal pancreatecomty	Tail	NR
8	Nasit et al	India	2012	1	Case 1	Male	10	Abdominal swelling	Open	NR	Head	NR
9	Laje et al	USA	2013	6	Case 1	Female	15	Abdominal pain	Open	Distal panceartectomy and splenectomy	Tail	8
					Case 2	Female	15	Abdominal pain	Open	Pancreatecoduodenectomy	Head	16
					Case 3	Female	11	Abdominal pain	Open	Spleen-preserving distal pancreatecomty	Body	5
					Case 4	Male	14	Abdominal pain	Open	Distal panceartectomy and splenectomy	Tail	12
					Case 5	Female	17	Jaundice	Open	Pancreatecoduodenectomy	Head	15
					Case 6	Female	18	Abdominal pain	Open	Distal panceartectomy and splenectomy	Body	5
10	Rodrigues-Duarte et al	Portugal	2013	1	Case 1	Male	13	Abdominal fullness	Open	Distal pancreatecomty	Head	NR
11	Tlili et al	France	2013	3	Case 1	Female	14	Abdominal pain	Open	Median pancreatecomy with pancreatecogastricanastomsis	Head	15

					Case 2	Female	13	Abdominal pain	Open	Median pancreatecomy with pancreatecogastricana stomsis	Body	21
					Case 3	Female	12	Abdominal pain	Open	Pancreatecoduodenectomy	Head	20
12	Escobar et al	USA	2014	1	Case 1	Female	11	Abdominal pain	Open	Pancreatecoduodenectomy	Head	17
13	Mohammed et al	Saudi Arabia	2014	1	Case 1	Female	15	N/V, abdominal pain	Open	Distal panceartectomy and splenectomy	Body, tail	10
14	Petrosyan et al	USA	2014	3	Case 1	Female	14	Abdominal pain	Laparoscopic	Distal panceartectomy and splenectomy	Tail	NR
					Case 2	Female	13	Abdominal pain	Laparoscopic	Distal panceartectomy and splenectomy	Body, tail	NR
					Case 3	Female	13	Abdominal pain	Laparoscopic	Spleen-preserving distal pancreatecomty	Body, tail	NR
15	Nachulewicz et al	Poland	2015	1	Case 1	Female	13	Asymptomatic	Open	Roux-en-Y pancreaticojejunosmty	Head	30
16	Słowik-Moczydłowska et al	Poland	2015	2	Case 1	Female	15	Abdominal pain	Open	Pancreatecoduodenectomy	Head, body	NR
					Case 2	Female	12	Asymptomatic	Open	Left pancretecomy and spleenectomy	Body	NR
17	Bidassek et al	Germany	2016	1	Case 1	Female	8	N/V, abdominal pain	Open	NR	Head	7
Total				29	25 Females, 4 Males (Female-Male ratio 6.25:1)		Average= 13.12 years					Average= 7.8

LOE: Length of stay, NR: not reported