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Case Series

Solid Pseudo-Papillary Tumor of the Pancreas

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Introduction

Solid pseudo-papillary tumor of the pancreas is rare and usually non-metastatic mass of the pancreas. Mostly affecting young women, it has diverse presentations at the beginning of the symptoms. The overall prognosis is excellent and only few patients become complicated. Here we discuss three cases.

Case Series

Case 1

First case was 13-year-old girl who was admitted in surgery ward with a mass in head of pancreas. Her problem had started 1 year prior to admission, with post-feeding infirmity. Sometime after, she became a bit pale and icteric; these progressed into general weakness and biliary vomiting 6 months after the first presentation. Defecation habit was changed after that. 2 months later, following a minor abdominal trauma, she became very ill and vomited past days undigested food. Sonography and CT-scan revealed a cystic well-defined mass, with mixed density in sub hepatic area in head of pancreas; suggestive for pseudocyst of the pancreas, duplication cyst, seromucinous cyst of the pancreas, and hydatid cyst. Peritoneal fluid was also detected. A large cystic mass was detected at the head of the pancreas at the time of operation therefore wipple procedure was done for patient. Diagnosis was confirmed by two pathologists

Case 2

Second case was a 10-year-old girl with an acute progressive pain in left flank, radiating to back, being accompanied by nausea and bloodless, non-biliary vomiting consisted of undigested food. She had no previous constitutional symptom or weight loss and no change in defecation. No complaint of fever she had, but axillary temperature of 38.5°c was recorded in admission with stable vital signs. Abdominal tenderness without any rebound tenderness in epigastric zone was detected. Laboratory evaluations revealed mild leukocytosis (13700/mm^3 with 68% neutrophil and 29% lymphocyte). Ultrasound imaging depicted a solid echogenic soft tissue, 89*60 mm in dimensions, near the spleen hilum, above left kidney, at the end of pancreas tail. CT scan with contrast media showed the same mass without enhancement, other lesions and lymphadenopathy. Imaging was suggestive for lymphoma, neuroblastoma and Wilm's tumor.

She went under operation under general anesthesia and supine position with midline laparatomy. A solid-cystic tumor was located at tail of the pancreas that was totally attached to the hilum of the spleen and we could not separate them by surgical maneuvers, therefore distal pancreatectomy and total splenectomy was done to excise the tumor. Diagnosis was confirmed by two pathologists.

Case 3

Third case was a 10-year-old boy with recurrent non-significant abdominal pain from one year ahead of admission, was admitted with additive progressive jaundice, and dark urine. No history of warning signs, weight loss and fever was present. Good general conditions, stable vital signs, soft abdomen with no tenderness or guarding and normal span liver were found in physical findings.

Ultra sonographic evaluation and CT-Scan showed a solid-cystic mass at the head of the pancreas. Upper midline incision in general anesthesia and supine position was made for the child in surgery. The mass was attached to duodenum, portal and inferior vena cava veins and head of the pancreas. Duodenum and distal end of CBD was excised with tumor because of tumor adhesions to these organs. The mass excision was completed by shaving the adhesions from the IVC. Whipple surgery was done for the patient after complete excision of the tumor.

Pathologic findings

Sections showed neoplasm composed of massive areas of necrosis and presence of pseudopapillae, covered with single to multiple layers of epithelial cells, consisting of ovoid and folded nuclei, indistinct nucleoli and rare mitotic figures. Fibrovascular cores of papillae were

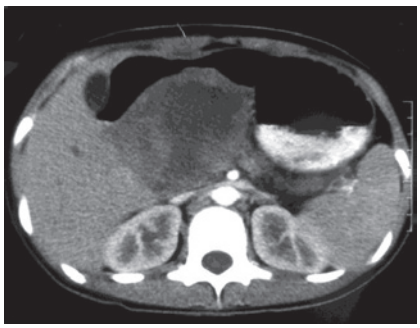

Figure 1: 1st Case: CT-Scan.

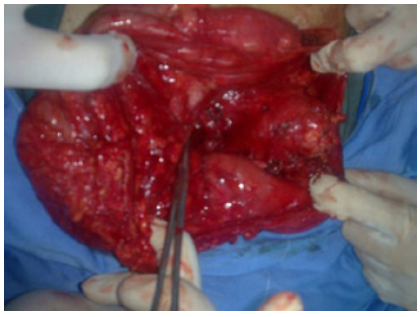

Figure 2: 1st Case: Intraoperative photo.

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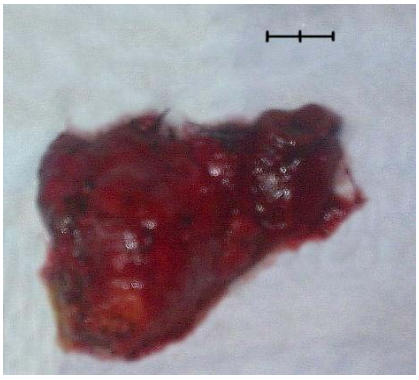

Figure 3: 1st Case: Dissected head and neck of pancreas.

thick with mucinous changes focally (Figures 1,2 and 3).

Immune histochemistry (IHC) evaluation revealed positive reaction for α1-antitrypsin, vimentin, calretinin and CD10. It was also negative for carcinoembryogenic antigen (CEA), CA19-9 and chromogranin. Despite being moderately positive in 3rd case for chromogranin in some foci, but generally it was negative.

Follow up

All the patients were free of sign and symptoms in the next months' follow up. Radiologic studies also showed clear abdomen and thorax of any mass.

Discussion

Solid pseudo-papillary tumor of the pancreas is a rare and mostly benign entity accounting of pancreatic non-endocrine tumors. Papillary cystic epithelial neoplasm, solid-cystic papillary epithelial neoplasm, solid and pseudo-papillary epithelial neoplasm, have been used to nominate this tumor interchangeably [1,2], but "solid pseudo-papillary tumor of the pancreas" (SPTP) is in favorite of WHO [2]. Despite its rarity, it is good for physicians to keep this in mind because of the curability and 5-year survival of 95%. In this report, we explained three cases which were diagnosed and treated as SPTP.

No presentation can be attributed specifically to SPTP, but the routine signs and symptoms resulting from an occupying mass. Non-significant pain, palpable mass, obstructive jaundice, bloating, dyspepsia [3], and less commonly pancreatitis and hemoperitoneum are other presentations [4]. Having this distinct tumor in mind is important in young females, coming with an abdominal mass, to prevent misdiagnosis. In our 1st case, as the patient had a history of an abdominal trauma, it was crucial not to presume the mass for pancreas pseudocyst; which could lead to mismanagement of the patient.

Since the first introduction half a century ago by Frantz [5], there have been numerous case reports of the entity; yet not enough to say it's not rare. Mostly affecting young adolescent girls and young females in all races [1,2], It's overall occurrence ranges between 0.17% of nonendocrine tumors [6,7] to 2.7% of all pancreatic tumors [8,9], yet it is a rare tumor in children.

Surgery is still the best modality for the patient with resecting the whole capsulated tumor [1]. Distal pancreatectomy and Whipple surgery, depending on the situation, are common [10]. Kanter et al, used neo-adjuvant chemotherapy so as to downsize the tumor in a 14-year old girl, to respectable dimensions [11].

All the patients were free of sign and symptoms in the next months' follow up in our study. Radiologic studies also showed clear abdomen and thorax of any mass. Therefore, we should have a suspicious of this tumor in differential diagnosis of pancreatic masses in children. Papavramidis et al, reviewed 718 case reports of SPTP from 1933 through 2003, suggest an increasing rate owing to the better diagnosis, accounting for 6% of all exocrine pancreas tumors [10], compared with 2% in literature [2], most of them are in adult not in children.

Underlying pathogenesis leading to tumor formation is not clarified till now [10]. Some reports suggest β-catenin mutations anyway [12,13]. This mass can be detected in ultrasonography, CT-Scan and MRI.

Excessive necrosis, varying amounts of cystic changes, solid areas rather than small cells with pseudorosette formation mimicking endocrine tumors, cystic areas with papillary structure [1,14] are the pathologic presentation.

Immune histochemistry (IHC) reveals granules containing periodic Acid-Schiff positive granules with positive progesterone receptors, α-antitrypsin, α-antichymotrypsin, phospholipase A2, neuroendocrine enolase, synaptophysin. They are free of carcinoembryogenic antigen (CEA), CA19-9, and tissue peptide antigen; which are usually seen in pancreatic carcinomas [1,15].

No clinical definite diagnosis can be made, unless it is proved by the pathologic studies, therefore the correct diagnosis was performed after pathologic evaluation in our patients.

Surgery is still the best modality for the patient with resecting the whole capsulated tumor [1]. Distal pancreatectomy and Whipple surgery, depending on the situation, are common [10]. Kanter et al, used neo-adjuvant chemotherapy so as to downsize the tumor in a 14-year old girl, to resectable dimensions [11].

Few cases have been reported with metastasis, leaving more than 95% of them with complete cure by single resection [10].

All the patients were free of sign and symptoms in the next months' follow up. Radiologic studies also showed clear abdomen and thorax of any mass.

Conclusion

We should keep in mind pseudo-papillary tumor of pancreas as a differential diagnosis of pancreatic cysts or masses in pediatric age group, to avoid mismanagement of these patients.

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