Case Report

Incidently detected solid pseudopapillary neoplasm of pancreas in a child with an ovarian cyst: a case report

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ABSTRACT

Solid pseudopapillary neoplasms (SPN) are the most common as well as very rare pediatric tumors of the pancreas. Most of the literature is derived from adult SPNs. As per world health organization, these tumors considered as low grade malignant with excellent survival outcomes after complete surgical resection. We report a case of incidentally detected SPN in a 16-year-old female child with an ovarian cyst. She underwent pylorus preserving pancreaticoduodenectomy and ovarian cystectomy. Histopathology revealed well differentiated SPN of the pancreas head and ovarian cystadeno. She is symptom-free at the 6th month follow up.

Keywords: Pancreas, Pediatric, Solid pseudopapillary neoplasm

INTRODUCTION

Solid pseudopapillary neoplasm (SPN) of the pancreas constitutes less than 1% of all pancreatic tumors.¹ Even though these are the most common tumors of the pancreas in children, they occur very rarely among them.² As per the world health organization (WHO) classification of pancreatic tumors, SPNs are considered to have the low-grade malignant potential.³ The majority of the literature on SPN is based on adult data and there are only several case series and reports available from childhood SPN. These tumors have excellent survival with complete surgical excision and limited data available on pediatric SPN behavior. Authors report a case of SPN in a female child aged 16 years with a brief review of the literature.

CASE REPORT

A 16 year old girl presented with abdominal pain and distension in the lower abdominal region for 3 months without any significant past history. On clinical examination, her abdomen was soft with 10X10 cm mass palpable in the hypogastric region probably arising from the pelvis. On ultrasound examination, a 15X14X8 cms cystic lesion noted in the pelvis extending up to umbilicus with bilateral normal-appearing ovaries. Another 7X5 cm hypoechoic mass noted in the gall bladder and head of the pancreas with calcification. Computed tomography showed a 7X5X7 cms hypodense mass lesion with areas of calcification attached to the pancreatic head with large enhancing para-ovarian cyst in the pelvis measuring about 15X8X13 cms (Figure 1). Her cancer antigen 19-9 level is 32 (reference range, <39 U/mL). She underwent pylorus preserving pancreaticoduodenectomy with para-ovarian cystectomy. On visual inspection both ovaries are normal. Her postoperative recovery was uneventful and discharged on her 11th postoperative day. She is symptom-free at the 6th month of follow up.

On gross examination, the resected specimen from the pancreas showed 7X6X6 cms grey-white to brown solid lesion attached to head of the pancreas with areas of
hemorrhage and necrosis (Figure 1). On microscopic examination, the lesion consists of sheets of small papillae with thin vascular cores lined by cells with abundant eosinophilic cytoplasm with a round to oval nuclei with well differentiation denoting grade 1 tumor (Figure 1). Some of the papillae showed myxoid changes (Figure 1). The lesion is separated from the normal pancreatic acini by a fibrous capsule. There is no perineural or vascular invasion. On immunohistochemistry, the cells are strongly positive for CD 56, vimentin; strong cytoplasmic and nuclear positive for β-catenin and negative for cytokeratin-19 consistent with the diagnosis of solid pseudopapillary neoplasm. The resected ovarian specimen showed a thin capsule with clear fluid. The final pathological diagnosis was serous cystadenoma of the ovary.

Figure 1: (A) CT scan of abdomen showing pancreatic mass (arrow) and large ovarian cyst (arrow head), (B) Gross features of resected specimen showing grey white areas and necrosis, (C) Microscopic image showing cells with well eosinophilic cytoplasm with well differentiation, (D) Numerous papillae along with areas of myxoid changes.

DISCUSSION

SPNs are rare tumors arising from the pancreas and they are even rarer in children. These tumors are common in females and tend to occur more commonly from the tail of the pancreas. The pathogenesis of SPN is poorly understood. The presence of progesterone receptors and female preponderance favors an origin from neuroendocrine cells. Even though these tumors are considered as low grade malignant, approximately 8-14% will have distant metastasis. The common sites of metastasis are liver, inferior vena cava, and spleen. Complete resection is the definitive modality of therapy with 95% survival rates. Progression-free survival is longer even in incompletely resected tumors because of their anatomical locations. Other predicted factors for recurrence or malignancy are angio invasion, perineural infiltration, infiltration into the surrounding tissues and solid component. The data on pediatric SPN is limited and most of the available literature is derived from adult SPNs. Lee et al, compared 15 pediatric SPN to 47 adult cases and reported that children typically have a larger mass than adults and fewer chances of incidental detection. Bender et al, performed a systematic review of literature on pediatric SPN. They found SPN is more common among female children.

Approximately 54% arises from the tail and 46% from the head of the pancreas. The majority of them underwent surgical resection. Chemotherapy and radiation were given to 3.8% of patients as an adjuvant modality. The role of chemotherapy or radiotherapy is doubtful and mostly based on anecdotal reports. The recent increase in the incidence of incidental SPNs are probably due to the widespread use of CT scan.

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