Traumatic presentation of a solid pancreatic pseudopapillary neoplasm in a 7 year old girl

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Abstract

Solid pseudopapillary neoplasms of the pancreas are rare tumors that present in adolescence after having grown to a large size. We present the case of a young girl who had emesis and abdominal pain after hitting her abdomen on the side of her bathtub. She underwent workup and successful surgical resection in the same admission. In reviewing the literature, these tumors often present with a palpable mass or abdominal pain. They are diagnosed with various imaging modalities and have an excellent prognosis with complete surgical resection. Chemotherapy is reserved for unresectable or metastatic disease. The young age and traumatic presentation make this a notable case.

1. Case report

A 7 year old girl presented to our emergency department after falling and reportedly hitting the side of the bathtub with her abdomen. She had developed nonbloody, nonbilious vomiting and due to her persistent symptoms, her mother brought her to the hospital. Her past medical history included a diagnosis of Attention Deficit Hyperactivity Disorder and there was no family history of malignancy. On exam, she was afebrile with normal vital signs; a heart rate of 103 beats per minute and a blood pressure of 116/59 mm Hg. She was mildly tender to her epigastrium but without signs of peritonitis. There were no abrasions or bruising on her abdominal wall. She was a Tanner stage 3. Her white blood cell count was 15,300 cells/μL and her hemoglobin was 13.3 g/dL. Her basic metabolic panel was within normal limits but her amylase and lipase were elevated at 331 U/L and 352 U/L respectively. An abdominal CT scan demonstrated a 4.9 cm by 4.4 cm mass, without calcifications or evidence of hemorrhage, in the body of the pancreas (Fig. 1). She was admitted to the hospital for resuscitation and operative planning.

On hospital day 3 she was taken to the operating room for surgical resection of the pancreatic mass. Via a bilateral subcostal incision we approached the large tumor abutting the neck of the pancreas (Fig. 2). Splenic preservation was attempted but not successful as the splenic artery passed through the tumor capsule. This was not apparent from the CT scan. The pancreas was divided with a TA 60 linear stapler (Covidien, Massachusetts) and the staple line was over-sewed with a running 3-0 polydioxanone suture. A surgical drain was placed by it to bulb suction and clips were placed at the tumor site in case of adjuvant radiation.

The patient tolerated the procedure well. Post-operatively, pain was controlled with an epidural catheter and intravenous opioids as needed. Overnight, her temperature went as high as 103.2 °F and she was started on piperacillin with tazobactam. Her fever resolved over the next few days and her antibiotics were narrowed empirically to cefazolin. Her laboratory values showed normal bilirubin...
and stable hemoglobin values. A chest radiograph showed a small left-sided pleural effusion but she was not tachypneic or hypoxic. By postoperative day 4 she was tolerating a liquid diet, her epidural catheter was removed along with her Foley catheter, and she was switched to oral penicillin with potassium (Pen VK). The hematology—oncology service was consulted for management of her post-splenectomy vaccinations and follow-up imaging. She was discharged home the next day in good condition on Pen VK and hydrocodone. Pathology confirmed a solid pseudopapillary neoplasm of 5 cm confined to the pancreas with uninvolved margins and no metastasis to the spleen. It was surrounded by a tan-white fibrous capsule and did have extensive hemorrhagic necrosis. Microscopic examination showed islands of viable tumor with pseudo-papillary, rosette-like architecture (Fig. 3).

At follow-up two weeks later she was doing well with no complaints, her abdomen was unremarkable with well-healed surgical incisions.

2. Discussion

Solid pseudopapillary neoplasm (SPPN) of the pancreas is an uncommon tumor. The young age of the patient and the discovery after a traumatic injury are unusual features associated with this case. First described by Virginia Frantz in the 1950’s, SPPN has been described with terms including adenocarcinoma, cystic, solid, papillary, and epithelial as well as simply Frantz's tumor. This is one reason for a disparity in incidence from <1% in the past to as high as 6% of exocrine pancreatic tumors in more modern series [1]. A recent 20-year case series by Austin at the Texas Children’s Hospital found 22 patients with only 3 under 10 years of age and abdominal pain being the most common presentation [2]. Embryonal pancreatic pluripotent cells are suggested as the cell of origin as they are negative for mucin, enzymes, or hormones [3]. They do stain for CD99 however in a dot like pattern different from that seen in other solid tumors [4]. They are also characterized by the presence of abnormal presence of beta-catenin within the nucleus of the tumor cells. This pattern of beta-catenin expression is often associated with mutations within the Wnt-signaling pathway. Females are predominantly affected leading to theories about hormonal or genital tract association. These tumors characteristically show expression of progesterone receptor via immunohistochemistry. One possibility is that during embryogenesis the pancreas is adjacent to the female genital ridge and SPPN may represent a tumor of embryonal cells from this region [4].

SPPN typically shows an indolent growth pattern causing abdominal pain or a palpable mass. The most common presentation is due to abdominal pain about half the time followed by a palpable mass in a third then by asymptomatic incidental finding in 15% [1]. Ultrasound is commonly the first imaging modality and shows a heterogenously hypoechoic mass with a pseudo-capsule; Doppler shows reduced vascularity [5,6]. This is followed by CT in which it presents as a hypodense, well circumscribed, heterogenous mass.
often greater than 5 cm [5]. Magnetic resonance imaging (MRI) is particularly sensitive for evidence of bleeding which can be a diagnostic clue [6]. Although imaging is often sufficient for diagnosis, a percutaneous ultra-sounded guided needle biopsy can be helpful for cases in which pancreatoblastoma or lymphoma is on the differential diagnoses [7].

Treatment is surgical with complete resection being curative in over 90% of patients in which disease is limited to the pancreas. Recurrence is found in less than 15% and is associated with large tumor size and invasion into muscular vessels [8]. Some series show no metastases or mortality at over a year out from surgery but some recurrence associated with positive surgical margins [9]. Chemotherapy and radiation are reserved for unresectable or metastatic disease. Surveillance is controversial as metastatic potential and recurrence is very low but can be done with either ultrasound or CT scan annually for up to 10 years to look for hepatic or splenic lesions. This is especially important for those with positive margins, aggressive characteristics, or metastatic spread [10]. Liver metastasis after initial resection has recently been treated with adjuvant chemotherapy but there is not a well defined treatment algorithm [11]. Radiotherapy has also been used for unresectable disease [12,13]. Fig. 4 gives an algorithm for the diagnosis and treatment of SPPN.

3. Conclusion

A seemingly unfortunate traumatic event leads to an early discovery of SPPN in this young girl. These rare tumors are often very large at presentation but respond well to surgical resection. There is a role for surveillance for liver lesions with ultrasound for those with high-risk characteristics. Chemotherapy and radiation are reserved for unresectable or metastatic disease.

References