

INTRODUCTION

Pancreatic tumors have an annual incidence of 0.19 cases per million in the pediatric population in North America. Solid pseudopapillary neoplasms (SPN), an indolent tumor with low malignant potential, comprise 70% of pediatric pancreatic tumors. It has been described mostly in adolescents, mean age of 13-14 years. SPN commonly occurs in the pancreatic tail with surgery being the mainstay of treatment. We present a case of a 13-year-old female who presented with acute onset abdominal pain found to have SPN based on endoscopic ultrasound assisted biopsy with spontaneous regression of the mass

CASE HISTORY

A 13-year-old female presented with 4 days of acute epigastric pain with nausea and vomiting in the setting of positive sick contacts. Initial blood work was normal and abdominal ultrasound showed only right lower quadrant mesenteric lymph node enlargement. She was discharged home with supportive care. However, due to continued severe epigastric pain, she presented to an ED 2 days later.

WORK UP

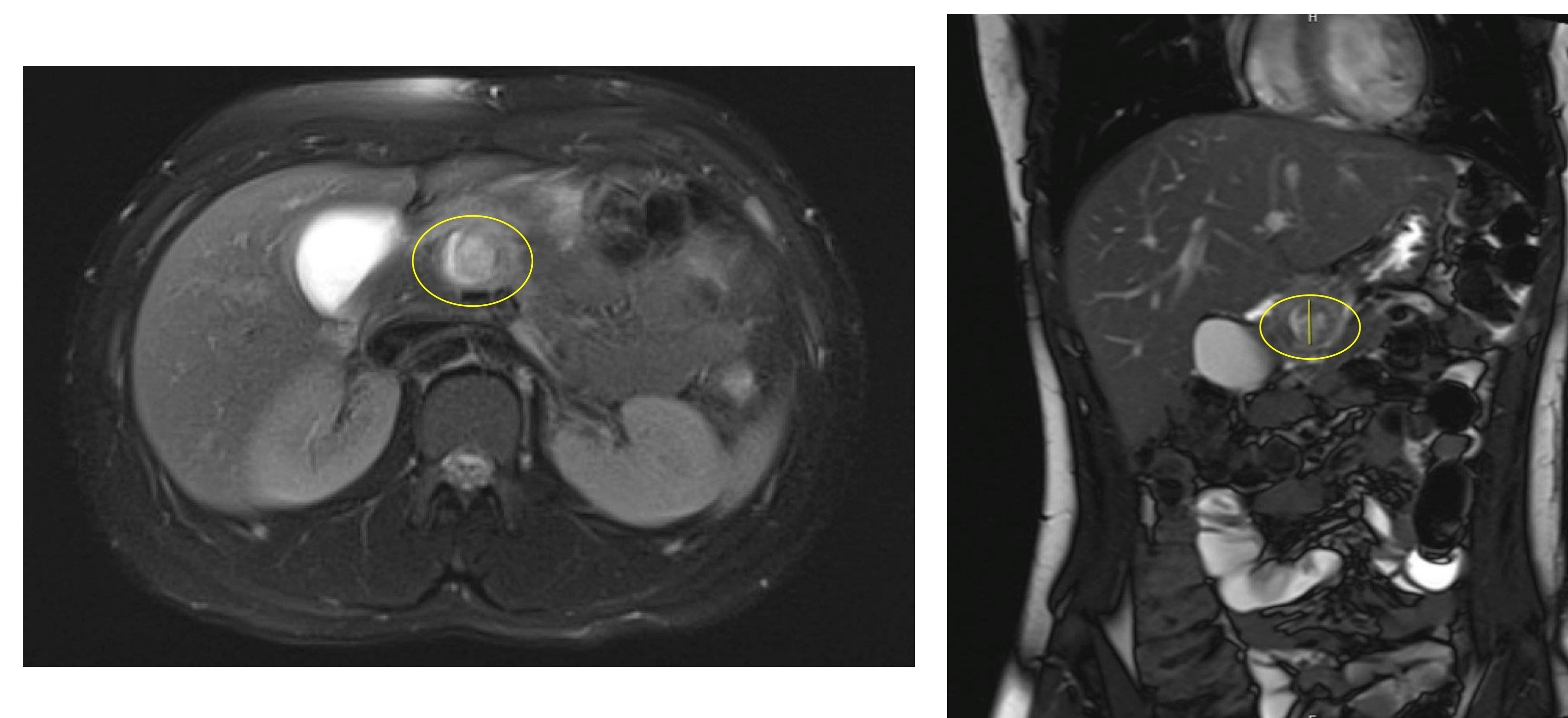
CT abdomen and pelvis (completed at outside hospital): A small 1.9 x 1.9 cm homogeneous hypoattenuating ring enhancing lesion in the neck of the pancreas. Pancreatic duct does not appear dilated. Confirmed on MRI at our hospital.
Tumor Markers: CA19-9, CEA, and IgG subclasses normal.

Endoscopic guided ultrasound biopsy: Small hypoechoic lesion suggestive of a thin-walled cyst in genu of the pancreas. Fine needle aspirate smear showed largely necrotic tumor, few viable tumor cells with mild nuclear atypia and abundant eosinophilic cytoplasm. Solid pseudopapillary neoplasm favored.
Immunohistochemistry stain (IHC): Beta-catenin: Positive. Insulinoma associated protein-1 (INSM1): Negative

INTERVENTION

Surgical excision was planned a few months later due to parental preference. Follow up imaging obtained 8 months later for surgical planning showed: Ill-defined, hypointense focus in the pancreatic neck measuring ~ 8 mm, located at site of 1.9 cm low-density lesion seen on prior CT. No main duct dilatation. Due to spontaneous reduction in the lesion, surgical resection was postponed with follow up imaging in 6-8 months.

MRI ABDOMEN WITH CONTRAST



ENDOSCOPIC ULTRASOUND

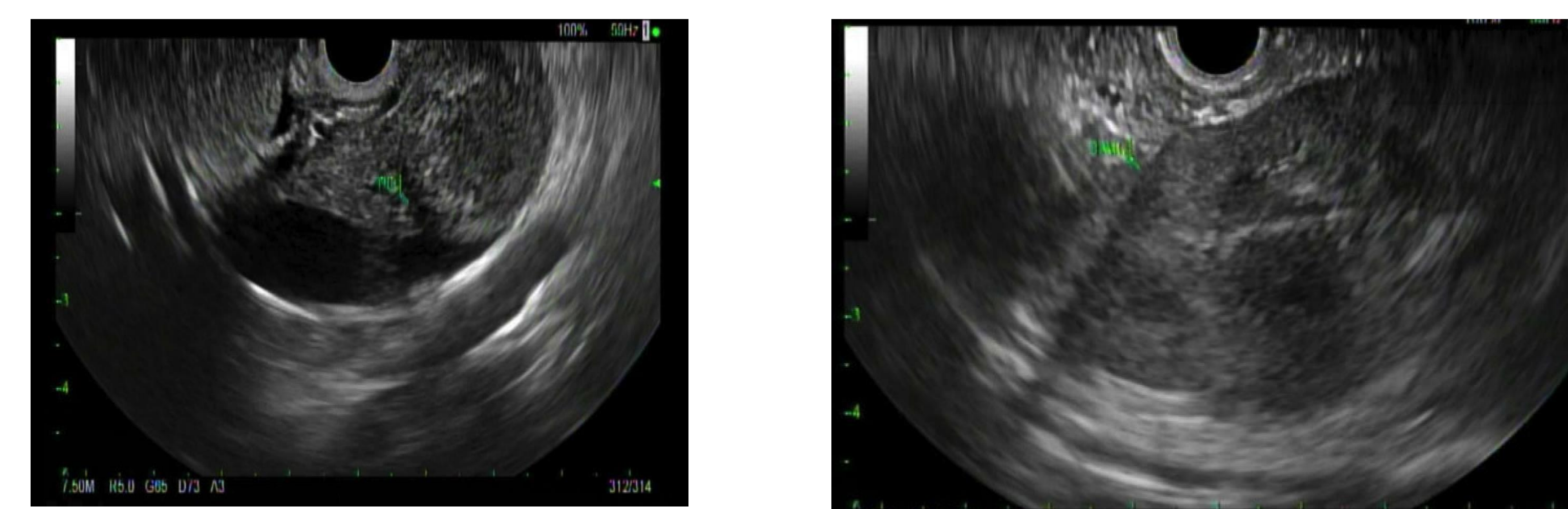


Figure: EUS showing 20x 21 cm hypoechoic cystic lesion in genu of pancreas

PANCREATIC PATHOLOGY

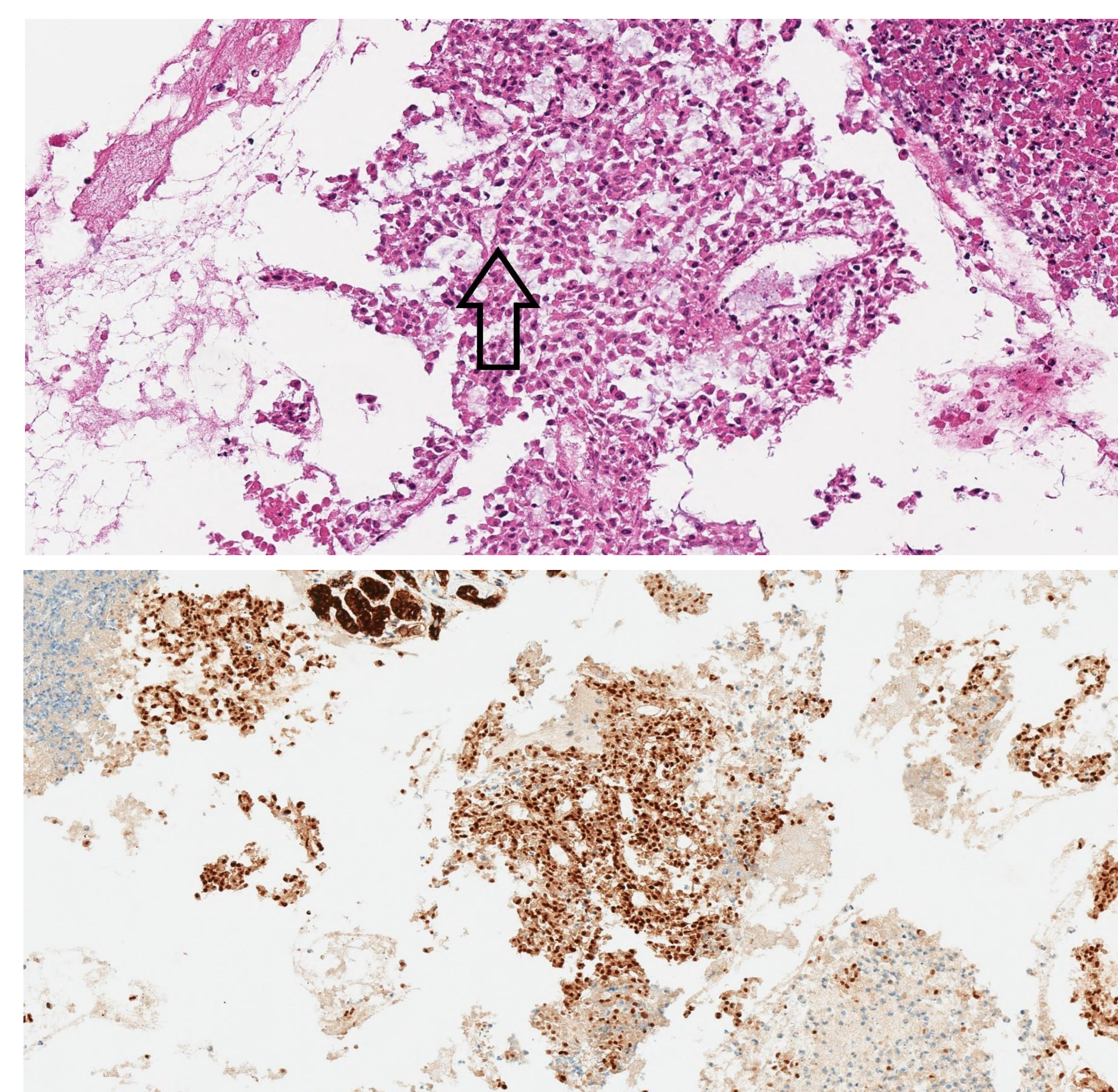


Figure: Small cells with bland, monotonous nuclei, and rare vascular elements (indicated by arrow).

Figure: Immunostaining showing nuclear positive (center of the screen); background normal pancreas with nuclear negativity (upper left)

*Pathology images courtesy Dr. William Twaddell

DISCUSSION

Pancreatic tumors are rare in children and require a high index of suspicion. Symptoms are often non-specific. MRI is helpful to demonstrate fibrous capsule and internal hemorrhage which are characteristic of SPN. IHC is a reliable diagnostic adjunct as almost all SPN tumors demonstrate nuclear expression for beta-catenin. SPN is mostly benign, but there are rare instances of metastasis. Hence, complete surgical resection is typically the treatment of choice. Our case demonstrates a unique outcome where no surgical intervention was necessary. From our literature review there has been only one other reported case of spontaneous regression of SPN in a 48-year-old woman 1 year later correlating with menopause.⁽¹⁾

CONCLUSION

Our case represents the first spontaneous regression of SPN in a pediatric patient. Overall SPN in pediatrics have excellent oncologic outcomes with survival rates > 95% after adequate resection even if there is no regression.

CONTACT

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