

CASE REPORT

Solid Pseudopapillary Tumor of the Pancreas in a Child: Imaging Findings with Diffusion-Weighted MR Imaging

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ABSTRACT

Context Solid pseudopapillary tumor of the pancreas is a rare tumor more common in young girls and rare in males. **Case report** We present a case of a solid pseudopapillary tumor of the pancreas in a 13-year-old boy, with typical imaging features. **Conclusions** Our case report specifically illustrates the potential of diffusion-weighted imaging findings on solid pseudopapillary tumor in pediatric patients.

INTRODUCTION

Solid pseudopapillary tumor of the pancreas is a rare exocrine tumor, described for the first time by Franz in 1959 [1]. Solid pseudopapillary tumor of the pancreas predominates in young Asian women in the second or third decade of life, while it is uncommon in male subjects [2]. We report a case of solid pseudopapillary tumor in which the preoperative clinical and imaging findings strongly suggested the diagnosis. A few reports of imaging features of solid pseudopapillary tumor in children are reported in the literature but this is probably the first case that includes diffusion-weighted MR imaging (DW-MRI).

CASE REPORT

A 13-year-old boy referring postprandial fullness and nausea for the past three months. Weight loss and pain in the upper quadrants of the abdomen recently occurred. Blood workup, including tumor markers, was unremarkable. Abdominal ultrasound (Figure 1) showed a well-circumscribed heterogeneous hypo-echoic mass in the region of the pancreatic body/tail. Unenhanced CT (Figure 2) confirmed the pancreatic lesion with a slightly heterogeneous density. No

calcifications were identified. T2-weighted MR images (Figure 3a) showed a 4.8 cm heterogeneously hyperintense lesion in the pancreas, showing a hypointense rim. This rim was also hypointense on T1-weighted images (Figure 3c), consistent with the presence of a tumoral fibrous pseudocapsule. T1-weighted images also showed some internal areas of increased signal intensity compatible with hemorrhage.

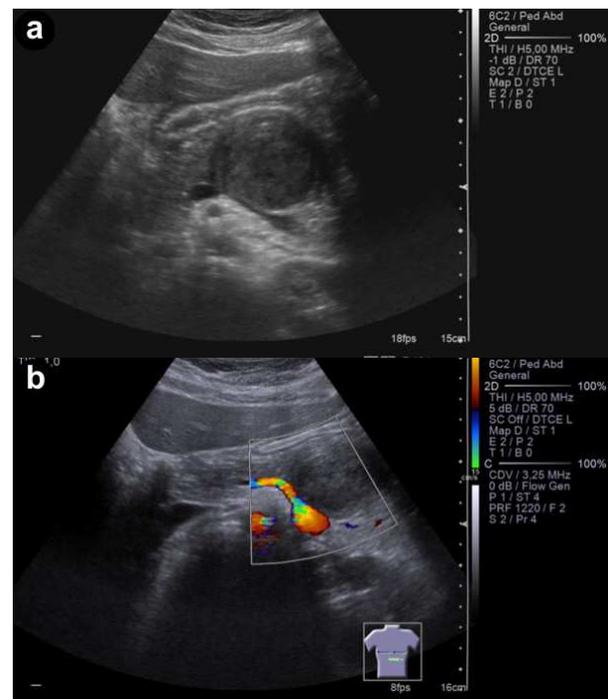


Figure 1. a. Transverse upper abdominal ultrasonography: hypo-echoic heterogeneous mass of the pancreas. b. Doppler ultrasonography showing no apparent tumoral vascularization.

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Key words Diffusion Magnetic Resonance Imaging; Pancreatic Diseases /diagnosis; Pancreatic Neoplasms

Abbreviations ADC: apparent diffusion coefficient; DW: diffusion-weighted

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Figure 2. Unenhanced axial abdominal CT scan: hypodense, slightly heterogeneous mass of the body/tail of the pancreas, without calcifications.

DW-MRI (Figure 4a) was performed (using *b* values of 50, 400 and 800) and the mean apparent diffusion coefficient (ADC) map was calculated (Figure 4b). The lesion presented a relatively high ADC value ($1.8 \times 10^{-3} \text{ mm}^2/\text{sec}$) compared to the ADC of the normal pancreatic gland ($1.4 \times 10^{-3} \text{ mm}^2/\text{sec}$), although there were some intralesional small areas of relatively low signal (minimum values of $1.3 \times 10^{-3} \text{ mm}^2/\text{sec}$). T2/DW fusion imaging was performed (Figure 4c). Gadolinium-enhanced MR imaging (Figure 5) showed early peripheral enhancement of the fibrous pseudocapsule. No central contrast enhancement was observed.

Based on the previous imaging findings the diagnosis of solid pseudopapillary tumor was suggested and distal pancreatectomy was performed. Pathology showed a neoplasm with extensive necrosis, with preserved tissue found in the tumor periphery under the fibrous capsule. Central aggregates of neoplastic cells with a pseudopapillary pattern were also observed and the diagnosis of solid pseudopapillary tumor was confirmed. There was no lymph node involvement.

DISCUSSION

Clinical presentations of solid pseudopapillary tumor of the pancreas range from asymptomatic “incidental” to unclear clinical features like abdominal pain or discomfort, poor appetite and nausea, which are related to tumor compression on adjacent organs [2].

Solid pseudopapillary tumor of the pancreas is a low malignant potential exocrine epithelial tumor composed of monomorphous cells forming solid and pseudopapillary structures, frequently with hemorrhagic-cystic changes [3]. The tumor varies from solid to nearly completely cystic, with the majority composed of a mixture of the two components [4]. Occasionally, the tumors are nearly completely cystic with a small amount of residual solid tumor at the periphery [4].

It is a well-encapsulated solitary tumor, round to ovoid and occur throughout the pancreas [2, 4]. The tumor is usually large at presentation [4] but it generally

displaces surrounding structures rather than invading them [5]. Because of its softness, solid pseudopapillary tumor rarely causes bile duct or pancreatic duct obstruction, even when it is located in the head of the pancreas [5].

The imaging features of solid pseudopapillary tumor reflect the pathologic findings of cystic and solid

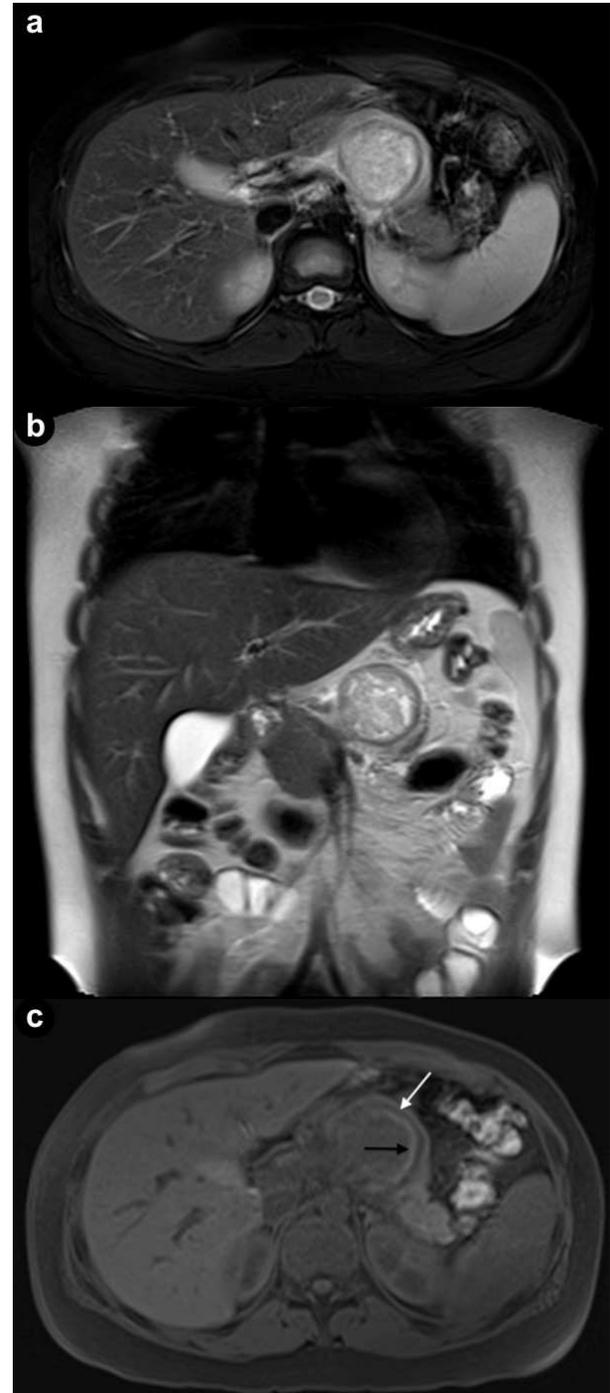


Figure 3. Axial fat-suppressed (a.) and coronal (b.) T2-weighted MR images show a well-margined lesion, heterogeneously hyperintense, in the body/tail of the pancreas, with a hypointense fibrous pseudocapsule. c. On an axial unenhanced fat-suppressed T1-weighted MR image the fibrous pseudocapsule is also hypointense (white arrow) and there is an internal peripheral high signal intensity rim (black arrow), a finding consistent with hemorrhage.

components, intratumoral hemorrhage, a fibrous capsule, and, less commonly, calcifications [4]. On ultrasound, solid pseudopapillary tumor presents as a well-circumscribed heterogeneous mass surrounded by a pseudocapsule of compressed pancreatic tissues and reactive fibrosis, sometimes with central cystic areas of necrosis [6]. The capsule may be visualized as an echogenic or, less commonly, hypoechoic rim at ultrasound and typically hypoattenuating at CT [4]. Unenhanced CT may identify hemorrhage and tiny calcifications [6, 7]. MRI should be considered the best imaging technique for children due to the absence of radiation and its

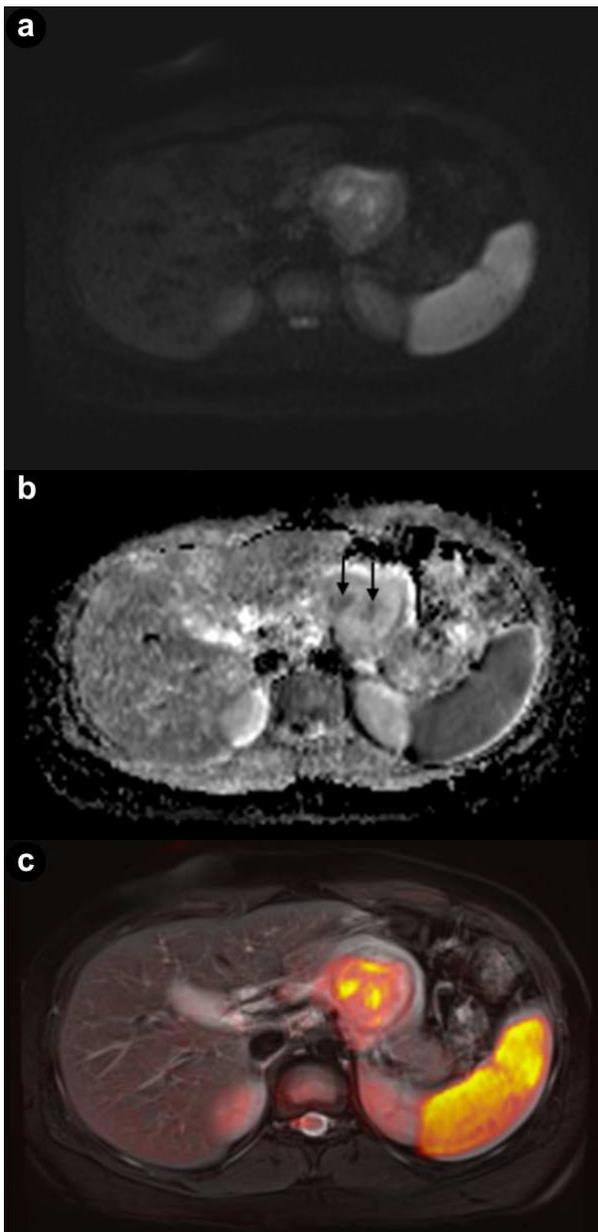


Figure 4. a. Axial diffusion-weighted image (b value of 800 sec/mm^2) shows a lesion with low signal intensity with some small central areas of higher signal intensity from restricted diffusion. b. Apparent diffusion coefficient mean value of the lesion is $1.8 \times 10^{-3} \text{ mm}^2/\text{sec}$ with the central hypointense areas having a minimum value of $1.3 \times 10^{-3} \text{ mm}^2/\text{sec}$ (arrows). c. T2-weighted and diffusion-weighted imaging were fused displaying a colored map of abnormal diffusion parameters.

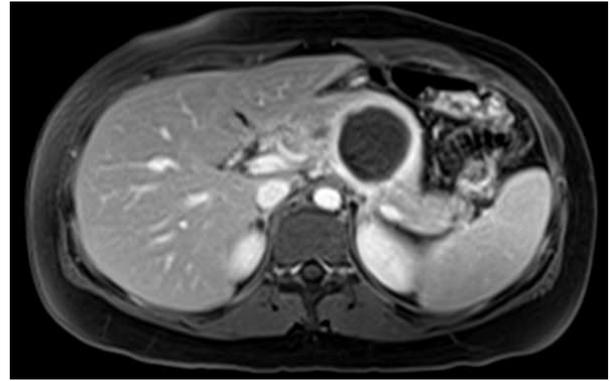


Figure 5. Axial contrast-enhanced fat-suppressed T1-weighted MR image shows peripheral enhancement of the neoplasm.

improved capacity for visualizing tumor components [6]. At MR imaging, T1-weighted images show a surrounding hypointense fibrous pseudocapsule and high signal intensity areas corresponding to internal hemorrhage, distinguishing features of solid pseudopapillary tumor [4, 6, 7]. Similar dark rim is also seen on T2-weighted images corresponding to the pseudocapsule [4]. The solid portions of the tumor are usually iso- to hypo-intense to pancreas on T1-weighted images and slightly heterogeneously hyperintense on T2-weighted images [4, 7].

The most common enhancement pattern of solid pseudopapillary tumor consists of early, peripheral enhancement of the tumoral pseudocapsule during the arterial phase, compared with the normal pancreas and the lesion itself [4, 5]. In some cases there is a progressive but heterogeneous fill-in of the lesion during the portal venous and equilibrium phases [5], showing less enhancement than the adjacent normal pancreas [4] consistent with a hypovascular tumor.

CONCLUSIONS

Our case report specifically illustrates the potential of DW-MRI findings in solid pseudopapillary neoplasms in pediatric patients. Wang *et al.* [8] describe DW-MRI in pancreatic lesions in adult patients and suggest that the components of the neoplasm (solid, cystic or hemorrhagic) determines the degree of diffusion and the ADC values, with the solid components accounting for the relatively low ADC values. In our case multiple b value DW-MRI allowed to discriminate better than T2-weighted or contrast-enhanced series the different components of the lesion according to signal behavior along with the increase in b value. Areas with low ADC values (compared to both normal pancreas and necrotic areas of the lesion) correspond probably to areas of increased cellularity. There are few reports about DW-MRI in cystic pancreatic neoplasms [8], so it remains to be established the role of DW-MRI to discriminate between benign and malignant lesions. Nevertheless, our findings emphasize the potential of DW-MRI to substitute contrast-enhanced imaging, particularly in the pediatric population.

Conflict of interest There is no conflict of interest with any financial organization regarding the material discussed in the manuscript

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