Pancreatic Tumors in Children

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Learning objectives

· To describe the spectrum of the imaging findings of epithelial and nonepithelial pediatric pancreatic lesions for making the correct diagnosis.

· To illustrate cases of a children with tumoral pancreatic lesions in our hospital.

Background

Paediatric pancreatic tumours are rare and diverse entities usually benign. Histologically, these tumors can resemble pancreatic embryonic elements or more well-differentiated pancreatic structures, in a subset the cellular origin can’t be delineated (1).

These tumors may be the first manifestation of a genetic syndrome (such as familial adenomatous polyposis or Wiedemann-Beckwith syndrome) or could occasionally present with life-threatening symptomatology. Additionally, they need to be properly characterized and distinguished from the more common, metastatic tumors to the pancreas (1-7).

Pediatric pancreatic neoplasms can be divided into epithelial and nonepithelial types. Most pancreatic neoplasms are epithelial lesions, which can be further subdivided into exocrine and endocrine subtypes; include solid pseudopapillary tumor, pancreatoblastoma, islet cell neoplasms and cystic lesions. Nonepithelial lesions include benign entities such as lymphatic malformations, malignant neoplasms such as lymphoma, and intermediate lesions, including inflammatory myofibroblastic tumors (3,4).

Clinical symptoms are often non-specific and presentation may be late.

Pediatric pancreatic neoplasms are capable of producing metastases, usually to the liver and lymph nodes; however, children have distinct clinical courses and have a better prognosis than adults (3,8).

Surgical resection is dictated by tumor location and remains the treatment of choice (1,2,9).
Findings and procedure details

Eight patients were identified with, serous cystadenomas, solid-pseudopapillary tumor, acinar cell carcinoma, lymphomas, intrapancreatic cystic spleen and other cystic lesions which can mimic tumors.

They are well demarcated with expansile growth patterns, may be quite large at diagnosis, central cystic necrosis is common, infrequently cause biliary duct obstruction (3).

Pancreatoblastoma

Pancreatoblastoma is the most common pancreatic tumor in children. Most occur in the first decade of life, there is a male predominance. Congenital cases of pancreatoblastoma have been described in association with Beckwith-Wiedemann syndrome (1,2,6,7,10).

In the majority it presents as an incidental abdominal mass, occurs with equal frequency in the head and tail of the pancreas, often the mass is as large at presentation as to make determination of the organ of origin quite difficult. The tumors appear as well-circumscribed heterogeneous masses with solid and cystic components, with prominent lobulation and multiloculated with enhancing septa, calcifications are common on CT. (1-3,11,12).

Metastatic involvement at presentation is found in one third of patients, and the liver is the most common site of spread, management of pancreatoblastoma consists of complete surgical resection, surgery is typically curative in the absence of metastatic disease, but recurrence is frequent (1,3,4).

Solid pancreatic pseudopapillary tumor

Solid pancreatic pseudopapillary tumors make up 1%-3% of all pancreatic tumors, occur predominantly in adolescent girls, and in the third and fourth decades of life. Less frequently, these tumors may present in children. Solid-pseudopapillary tumor, has low potential for malignancy, it generally behaves as a low grade neoplasm with a good prognosis, though aggressive behavior has been reported (1,2,12,13).

The pancreatic head was the most common site, solid pancreatic pseudopapillary tumor is commonly large, circumscribed and encapsulated, with cystic and solid components,
marked degenerative and hemorrhagic change, and, less commonly, calcification, smaller tumors tend to be predominantly solid and larger tumors tend to be more cystic (4,13).

Complete surgical excision is the treatment of choice with excellent outcomes. Tumors located in the body or tail of the pancreas can be managed with a laparoscopic distal pancreatectomy (14,15).

Fig. 1: 9-year-old boy with solid pseudopapillary tumor. Large, well-circumscribed, and heterogeneous mass with fibrous capsule, arising from body of pancreas. Cystic component is due to underlying hemorrhagic and necrotic change.

**References:** Radiology, Hospital Universitario y Politecnico LA Fe, B - Valencia/ES

Fig. 1 on page 12

**Acinar cell carcinoma**
Acinar cell carcinoma is quite rare in children, is pathologically divergent from ductal adenocarcinoma but similar to pancreatoblastoma, which is considered by some authors to be the embryonic counterpart to acinar cell carcinoma (3).

The presenting symptoms are most often nonspecific and related to local tumor growth or metastases. The most common manifestations are palpable abdominal mass or abdominal pain (2,3,16).

Acinar cell carcinoma and pancreatoblastoma are very similar in appearance owing to their common acinar differentiation; the histologic differentiation may be particularly difficult. This tumor can cause hyperlipasemia due to a unusual paraneoplastic syndrome called lipase hypersecretion syndrome, which may lead to diffuse subcutaneous nodules and polyarthropathy (3,16,17).

The tumor is most often a large, well-demarcated, soft, round to lobular mass, the masses are frequently exophytic, heterogeneous, with a central hypodense area, which represents tumoral necrosis, internal septations and peripherally located calcifications may be also seen. The pancreatic head was the most common site. Masses enhanced homogeneously but less than the surrounding normal pancreas (1-3,16).
**Fig. 2**: 11-year-old boy with acinar cell carcinoma who presented with abdominal pain and emesis. Ultrasound image depicts nonspecific heterogeneously predominantly hypoechoic lesion. Axial contrast-enhanced CT image shows large, well-marginated, round, hypodense and homogeneous mass arising from pancreatic head. MRI shows isointense mass to surrounding glandular parenchyma with partial capsule, non-enhancing central portion and peripheral mild enhancement.

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**Serous cystadenoma**

Cystic pancreatic neoplasm, which account for less than 1% of pancreatic neoplasm in adults, and are rare in children. There are two different types of cystic pancreatic neoplasms: microcystic serous adenomas and mucinous cystic neoplasm. While microcystic serous adenomas are benign, mucinous cystic neoplasm represent a
continuum of benign to malignant disease. Calcifications are less frequent with mucinous tumors than serous tumors (1,3,4).

Serous cystadenoma of the pancreas is a benign cystic tumor, which radiological diagnosis is easy in its typical microcystic variant, consists of numerous cysts, each measuring less than 2 cm in diameter, though larger cysts have been described (18-20).

The presence of multiple small cysts may mimic a solid lesional appearance at ultrasound and CT.

Although the tumor is not potentially malignant, patients require partial pancreatic resection (18,21).
**Fig. 3:** 13-year-old girl with Von Hippel Lindau syndrome. Cystic lesions with water-attenuation, serous cistoadenomas.

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**Fig. 3 on page 14**

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**Fig. 4:** Serous Cystadenoma in an 9-year-old girl who presented with diarrhea. US images of upper abdomen show homogeneously hypoechoic mass arising from body of pancreas. Axial and coronal T2-weighted MR images show high signal intensity.

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**Fig. 4 on page 15**

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**Cystic pancreatic lesions**
Cysts can be subdivided into false and true cysts. Microscopically the distinction between true cysts and pseudocysts is more apparent true cyst has an epithelial lining, but pseudocyst does not (1,3,4).

Pseudocysts are the most common cystic pancreatic lesion in the pediatric population, these lesions most commonly occur after blunt abdominal trauma and following pancreatitis (1-4).

Solitary true cysts may develop in the absence of a defined syndrome, whereas multiple true cysts can be seen in the setting of systemic disorders such as autosomal dominant polycystic kidney disease, von Hippel-Lindau syndrome, Beckwith- Wiedemann syndrome or Meckel-Gruber syndrome (1,3,4,21).

**Fig. 5**: 10-year-old boy with abdominal pain and emesis. Axial US image shows a round, anechoic cyst. Axial T2-weighted MR image shows uniform fluid signal intensity internally surrounded by low-signal-intensity wall, T2-weighted hypointensity within cyst wall is likely due to fibrous content.

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**Fig. 5** on page 16

**Lymphoma**

Pancreatic involvement in childhood lymphoma is unusual. Lymphoma can involve the pancreas, either primarily or secondarily, and is the most common nonepithelial pancreatic tumor. Variants of non-Hodgkin lymphoma, large-cell lymphoma and Burkitt lymphoma most commonly involve the pancreas in children (1,3,4,22).
Two different morphologic patterns of pancreatic involvement are seen: a localized, well-circumscribed tumoral form and diffuse enlargement infiltrating or replacing most of the pancreatic gland. The diffuse infiltrating pattern may mimic the imaging findings of acute pancreatitis with gland enlargement and irregular infiltration of the peripancreatic fat and the well-circumscribed tumoral form can be easily misinterpreted as a carcinoma, lymphoma can be distinguished from other tumors by the multiplicity of large nodal masses (1-4,22,23).

Chemotherapy is the treatment of choice for most patients with pancreatic lymphoma (23).

**Fig. 6**: 13-year-old boy with B-cell non-Hodking lymphoma. Axial ultrasound image of left upper abdomen shows heterogeneously hypoechoic mass. CT image shows ill-defined hypodense mass occupying body and pancreatic tail.

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**Fig. 6** on page 17
Fig. 7: Burkit lymphoma in a 12-year-old boy with epigastric pain for 3 weeks. Infiltrative hipodense lesion, leading to glandular enlargement and poor definition with minimal enhancement, in pancreatic tail a well-defined hipodensity mass. Enlargement peripancreatic nodes.

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Fig. 7 on page 17

Intrapancreatic accessory spleen

Although the tail of the pancreas is the second most common site of an accessory spleen, cyst formation in an intrapancreatic accessory spleen is extremely rare. Most patients with this condition have no clinical symptoms and it is often detected incidentally by imaging studies and generally does not require therapy. It is difficult to differentiate a splenic cyst in the pancreas from a primary pancreatic neoplasm because it lacks particular characteristics on radiological examination (24, 25).

Intrapancreatic accessory spleen is usually seen as a round or oval mass with a mildly echogenic and homogeneous. Typical location, similar attenuation of the lesion to the spleen on noncontrast, and postcontrast CT at different phases are helpful to make diagnosis. In particular, characteristic heterogeneous contrast enhancement of intrapancreatic accessory spleen on the arterial phase may be helpful for correct diagnosis (24-26).
Fig. 8: 17-month-old baby who underwent abdominal ultrasonography because of abdominal discomfort. US findings reveal a big cystic mass in the left upper quadrant. CT scan enhanced with intravenous contrast material shows a predominantly cystic mass in the pancreatic body and tail. No solid components or surrounding inflammatory changes are. The intraoperative finding was cyst in an intrapancreatic accessory spleen.

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Fig. 8 on page 18

Images for this section:
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Conclusion

· Pancreatic neoplasms are rare in children, knowledge of features imaging of the variety of lesions allows more appropriate diagnosis.

· Epithelial tumors are most common and include solid pseudopapillary tumor, pancreatoblastoma, islet cell neoplasms, and cystic lesions.

· Pancreatic neoplasms are rare in childhood and have a different histologic spectrum and better clinical outcome than in adulthood.

Personal information

References


