CASE REPORT

Unusual presentation of a pancreatic mass in an infant: pancreatic haemangioendotheliomatosis

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ABSTRACT. Primary pancreatic tumours are extremely rare in children. We report a case of a 5-month-old male with a diffuse invasive tumour of the head of the pancreas. The tumour demonstrated peripancreatic extension into the porta hepatis, which occluded the portal vein and invaded the superior mesenteric artery. It was found to be haemangioendotheliomatosis of the pancreas. Imaging, pathological findings and a brief relevant classification of haemangioma are discussed.

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Haemangioendotheliomas are vascular neoplasms that demonstrate histological features and clinical behaviour in-between the benign, well-differentiated haemangioma and the frankly malignant angiosarcomas [1]. Although infantile haemangioma is the most common tumour in an infant and may involve any organ, involvement of the pancreas is extremely rare. Very few cases of infantile haemangioendotheliomas of the pancreas have been reported [2–6]. Often, terms describing vascular lesions have been used interchangeably, which has resulted in controversy regarding the classifications of these lesions. Pathological descriptions and the most current classification of the various types of these vascular lesions are reviewed.

Case report

A 5-month-old male was referred to our hospital for evaluation and management of jaundice, which began 9 weeks prior to presentation. The patient had a liver biopsy at another hospital with results suggestive of biliary atresia. The patient presented with the following laboratory values: total bilirubin, 7.5 mg dl⁻¹; direct bilirubin, 6.1 mg dl⁻¹; serum glutamic-oxalocetic transaminase, 253 u l⁻¹; serum glutamic-pyruvic transaminase, 127 u l⁻¹; alkaline phosphatase, 457 u l⁻¹; gamma-glutamyl transpeptidase, 692 u l⁻¹; amylase, <30 u dl⁻¹ and lipase, 225 u l⁻¹. An ultrasound was performed that showed diffuse homogeneous enlargement of the pancreatic head (Figure 1). The mass was not hypervascular on colour flow Doppler assessment. The remainder of the pancreas was normal in appearance. The pancreatic duct distal to the head was

dilated. The liver parenchyma appeared heterogeneous and mild dilatation of the common bile duct and the intrahepatic biliary system was noted. Findings suggested an underlying mass in the pancreatic head, so CT and MR studies were performed.

The CT scan of the abdomen demonstrated a large and diffuse homogeneously and intensely enhancing soft tissue mass involving the head, body and uncinate process of the pancreas (Figure 2). Enhancing soft tissue, which extended beyond the confines of the pancreas into the region of the porta hepatis and appeared contiguous with this mass, was also noted. The superior mesenteric vein (SMV) narrowed just prior to its junction with the splenic vein and was surrounded by this mass; therefore, involvement of the vein could not be excluded. The superior mesenteric artery (SMA) was partly encased by the mass. As noted on the ultrasound, there was dilatation of both the distal pancreatic duct and the biliary system, which suggested obstruction.

MRI demonstrated similar findings to those seen on the CT and ultrasound studies. A homogeneously enhancing mass was again noted involving the pancreatic head, proximal body and uncinate process (Figure 3). The mass was isointense on the T₁ weighted sequence and appeared slightly hyperintense on the T₂ weighted sequence. The mass was found to extend into the porta hepatis following the course of the portal veins. The mass displaced the duodenum laterally and anteriorly.

The main diagnoses considered at this point were pancreaticoblastoma or a pancreatic haemangioma. Pancreaticoblastoma was favoured, owing to the presence of the enhancing soft tissue seen outside the pancreatic parenchyma in the region of the porta hepatis. This was thought to be either due to extension of the tumour or to associated lymphadenopathy.

The patient then underwent surgery. The mass was centred in the head of the pancreas. It was very vascular and bled upon touching. The SMA and distal SMV and

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Figure 1. Transverse ultrasound image of the abdomen, demonstrating diffuse homogenous enlargement of the pancreatic head (arrow). The mass is not hypervascular on colour Doppler.

splenic veins were invaded by the tumour. As a result of the extensive nature of the tumour and vascular involvement, it was decided that the only way to remove this tumour was via autotransplantation. A Whipple procedure with intestinal autotransplantation was performed. The distal splenic vein was resected and a splenorenal shunt placement was performed. The mass was sent for pathological evaluation.

Gross examination of the mass revealed an irregular firm tan-brown tumour measuring $3.2 \times 3.2 \times 2.9$ cm involving the pancreatic head, which was surrounded by several arterial and venous blood vessels (Figure 4a). The tumour was locally invasive and extended to the common bile duct and mesenteric duodenal serosa and

infiltrated the liver, local nerves, lymph nodes and blood vessels. On histological examination, the tumour was characterised by thin-walled vessels interspersed with coalescent nodules of spindle neoplastic cells forming small slit-like spaces filled with red blood cells, and mixed with nests of round epithelioid cells and fibrin thrombi-rich capillaries. The cells had pale cytoplasm and oval nuclei with delicate nuclear chromatin. The nuclear atypia was minimal and the rate of mitosis was low (Figure 4b). Neoplastic cells were positive for endothelial cell marker CD31, focally for glucose transporter 1, and negative for keratin and chromogranin. Immunostain for Ki-67 showed a high proliferative index. The final pathological diagnosis was haemangioendotheliomatosis.

The patient had a complicated post-surgery course, with thrombosis of the SMA, intestinal necrosis and thrombosis of the right superficial femoral artery. The thrombosis of the right superficial femoral artery was probably due to catheterisation. Since discharge, the patient has been doing well.

Discussion

Cellular haemangiomas of infancy, also known as "infantile haemangioendotheliomas," are benign tumours whose dense cellularity can be confused with other aggressive masses [7]. They are vascular tumours that may involve numerous organs including the skin, liver, spleen and salivary glands [4]. The pancreas is a rare site of occurrence for this tumour, with only a few previously reported cases [2–6]. Presenting signs and symptoms for these tumours include obstructive jaundice, hepatomegaly, palpable mass, duodenal obstruction and intestinal bleeding [6]. Most cases of pancreatic haemangiomas, including the one presented here, have been diagnosed following surgical biopsy.

Imaging findings of haemangioendotheliomatosis is similar to haemangioma, except for the more extensive and locally invasive feature of the lesions. On ultrasound,

(a)

(b)

Figure 2. Axial post-contrast CT scan at the level of the pancreas (a,b) a well defined intensely and homogeneously enhancing mass in the head, body and uncinate process of the pancreas (short thick black arrows). Peripancreatic enhancing soft tissue is also noted (white arrow). Dilatation of the distal pancreatic duct is shown (long thin black arrow).

Figure 3. (a) Axial and (b) coronal T_1 weighted fat saturated post-contrast and axial fat saturated T_2 weighted. (c) MRI of the abdomen demonstrating the large enhancing mass involving the head and uncinate process of the pancreas (long thin arrows). Peripancreatic extension of the mass into the porta hepatis surrounding the right and left portal veins is well seen (short black arrows). The mass is isointense to the kidneys on the T_2 weighted images and there is minimal peripancreatic fluid (white arrow).

these lesions are homogenous and can be isoechoic or hyperechoic to the pancreatic parenchyma. All previously reported cases, including the one presented in this report, have demonstrated a mass in the head of the pancreas. CT and MRI not only help in localising the mass, but can also give information on any associated findings. Signs of obstruction, such as dilated pancreatic and biliary ducts, may be seen. The cause of the heterogeneous appearance of the liver in this case is not entirely clear, but may have been due to the obstructed biliary system and resultant cholestasis. Intense and homogenous enhancement, similar to haemangiomas in other organs, can be seen. In some reported cases, ultrasound studies have demonstrated portal vein stenosis and portomesenteric vascular thrombosis [4].

MRI findings frequently show a lobulated mass with moderately increased signal intensity on T_2 weighted images and marked enhancement following intravenous gadolinium [6].

The other differential diagnosis considered was a pancreaticoblastoma, given the large size and the peripancreatic extension. However, pancreaticoblastomas are heterogeneous in appearance on imaging and have both cystic and solid components [8]. They can also be multiloculated with enhancing septations, and demonstrate calcification: findings not seen in this case.

The treatment of pancreatic haemangiomas is variable and dependent upon the associated biliary obstruction and extension of mass and vascular invasion. Resection has been reported in previous cases [9]. Systemic steroids and

(a)

(b)

Figure 4. (a) Gross cut specimen an irregular, firm, tan-brown tumour encased by the C-loop of the duodenum. The superior mesenteric artery and the superior mesenteric vein (arrows) are seen encased by the tumour. (b) Histological slide shows thin wall vessels interspersed with coalescent nodules of spindle neoplastic cells (arrows) forming small slit-like spaces filled with red blood cells, and mixed with nests of round epithelioid cells and fibrin thrombi rich capillaries.

interferons have shown variable results. There have been successful attempts at biliary stenting and external drainage pending spontaneous tumour regression [5].

Conclusion

We describe a new case of pancreatic haemangioendotheliomatosis. Although imaging characteristics were fairly characteristic for a haemangioma, the peripancreatic extension made the diagnosis difficult and the mass was confused with a pancreaticoblastoma. A difficult Whipple procedure, with intestinal autotransplantation and the formation of a splenorenal shunt, was required to treat this patient, because of tumour infiltration of the superior mesenteric vessels and the splenic vein. As a result of their variable nature and presentation, it remains to be seen what the appropriate treatment is for these rare tumours.

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