

CASE REPORT

Pediatric Pancreatic Hemangioma: A Case Report and Literature Review

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ABSTRACT

Context The pancreas is an unusual site for a hemangioma in an infant. A child with obstructive jaundice caused by a pancreatic hemangioma is presented and management strategies for this benign tumor are discussed.

Case report A 5-month-old girl presented with a 2-week history of jaundice, pale stools and dark urine. Liver function tests confirmed obstructive jaundice. An abdominal ultrasound scan and magnetic resonance imaging showed an enhancing mass in the head of the pancreas. At laparotomy, a wedge biopsy of the pancreatic tumor was taken and a tube cholecystostomy inserted. Histological examination of the specimen revealed a pancreatic hemangioma with sclerotic features. The high volume of bile loss from the cholecystostomy proved problematic and biliary diversion with a Roux-en-y hepaticojejunostomy was therefore performed. The tumor subsequently regressed spontaneously and was no longer visible on follow-up imaging two years later. The child has since thrived.

Conclusions Pancreatic hemangiomas are rare and may cause diagnostic confusion. Pancreatic resection should be avoided since the natural history of these benign tumors is

that of spontaneous involution. Various strategies can be used to manage any associated obstructive jaundice.

INTRODUCTION

Vascular anomalies can be divided into two distinct categories, hemangiomas and vascular malformations, based on clinical and histological characteristics. Hemangiomas are the most common tumor of infancy and typically present as cutaneous lesions. Their natural history consists of a proliferative phase during the first year of life followed by an involuting phase of variable duration lasting up to 12 years of age [1]. Hemangiomas may cause cosmetic problems or be complicated by ulceration and bleeding, high output cardiac failure, Kasabach-Merritt syndrome [2], or compression of adjacent structures.

Visceral hemangiomas have been described in various organs including the brain, parotid, thorax, liver, spleen and gastrointestinal tract. Pancreatic hemangiomas are rare. Less than 10 have been described in children. A pancreatic hemangioma may present with obstructive jaundice when it can lead to diagnostic confusion and difficulties in management. This report documents a further case of an infantile pancreatic hemangioma

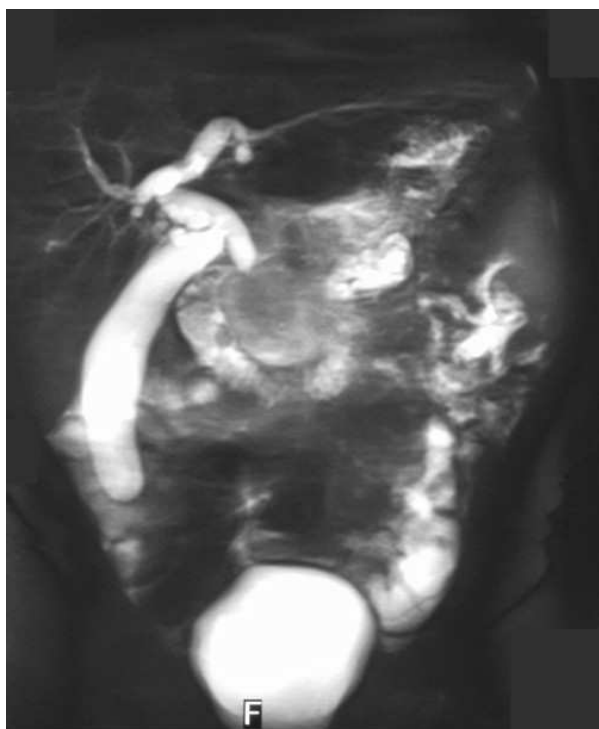


Figure 1. Magnetic resonance cholangiopancreatography demonstrates a distended gallbladder and dilated intra- and extrahepatic ducts down to the level of the head of pancreas.

and highlights some of the difficulties encountered in clinical management.

CASE REPORT

A 5-month-old girl presented with a 2-week history of jaundice, pale stools and dark urine. She was born at term and had been otherwise well. On examination she was markedly jaundiced and had mild hepatomegaly but no cutaneous hemangiomas. Biochemical liver function tests were abnormal (total bilirubin 200 $\mu\text{mol/L}$, reference range: 5-21 $\mu\text{mol/L}$; conjugated fraction 173 $\mu\text{mol/L}$, reference range: 3-18 $\mu\text{mol/L}$; alanine aminotransferase 221 IU/L, reference range: 0-40 IU/L; alkaline phosphatase 2,136 IU/L, reference range: 100-400 IU/L; gamma glutamyl-transpeptidase 957 IU/L, reference range: 0-50 IU/L). Plasma lipase was mildly elevated at 132 IU/L (reference range: 22-51 IU/L) but plasma amylase was normal. Her hemoglobin was 9.5 g/dL (reference range: 9-14.1 g/dL) with a normal platelet count. Other hematological investigations included a

normal immunoreactive trypsinogen, no detectable common mutations for cystic fibrosis, and normal alpha-fetoprotein and alpha-1-antitrypsin levels. Urinary catecholamines were normal. An abdominal ultrasound scan revealed a 3x2 cm mass in the head of the pancreas but the echotexture of the pancreatic head was similar to the remainder of the gland. The common bile duct was dilated (9 mm) and the gallbladder distended with marked intrahepatic duct dilatation. Magnetic resonance imaging (MRI) including magnetic cholangiopancreatography (MRCP) confirmed a 3 cm mass in the head of the pancreas with associated biliary obstruction (Figures 1 and 2). The pancreatic duct in the body and tail of the gland was not dilated but there was some inflammatory change around the pancreas and evidence of mild lymphadenopathy at the porta hepatis and in the small bowel mesentery. The mass showed delayed enhancement after gadolinium injection with centripetal filling (Figure 3).

A percutaneous needle biopsy of the tumor was considered but it was decided to take an open biopsy in order to obtain sufficient tissue for histology. At laparotomy a hard, irregular mass involving the head and neck of the pancreas was identified. Some mesenteric vascular congestion was noted. Several



Figure 2. Coronal oblique MIP (maximum intensity projection) post gadolinium image demonstrates a low signal mass in the head of the pancreas causing nipping of the portal vein.

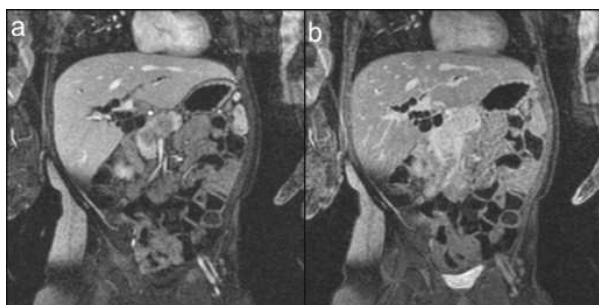


Figure 3. Coronal oblique post gadolinium image in the portal venous phase (a.) demonstrates a low signal mass in the head of pancreas with delayed enhancement demonstrated on the delayed coronal oblique post gadolinium acquisition (b.).

superficial wedge biopsies of the pancreatic mass were taken along with a lymph node from the porta hepatis. A tube cholecystostomy was inserted.

Histopathology showed replacement of pancreatic tissue by lobules of spindle cells lining slit-like spaces and capillary sized vascular channels (Figure 4). The lobules were separated by dense fibrous tissue. Immunohistochemistry showed the spindle cells to be strongly positive for CD31 and CD34 markers, consistent with an endothelial origin (Figure 5). The stroma was strongly positive for actin but negative for vimentin and desmin. There was no evidence of malignancy and the lymph node biopsy was benign. It was concluded that the specimen

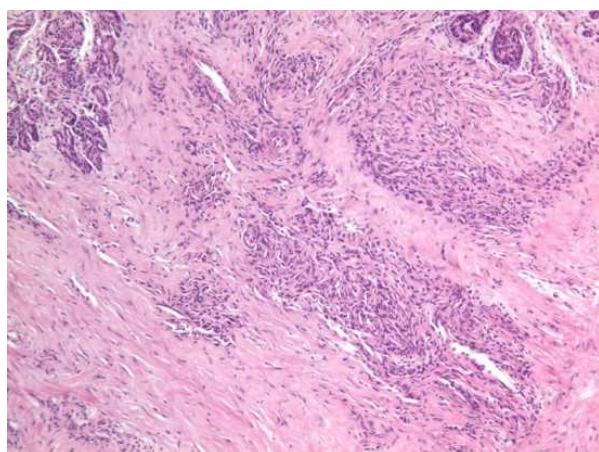


Figure 4. The biopsy shows lobules of small capillary vascular channels separated by densely sclerotic stroma replacing the pancreatic tissue. Some residual acinar pancreatic tissue is seen in the upper right of the specimen. (Hematoxylin and eosin, x100).

represented a pancreatic infantile capillary hemangioma with marked sclerosis. Cytogenetic analysis of the tumor showed a normal female karyotype.

Postoperatively, bile drainage via the cholecystostomy tube increased to 200-400 mL/day (maximum 3 mL/kg/h). During the next two weeks, this fluid was replaced with intravenous 0.9% sodium chloride and potassium since the patient was unable to tolerate full enteral feeding together with oral fluid and electrolyte replacement. A cholangiogram demonstrated persistent and almost complete obstruction of the distal common bile duct and a narrow, tortuous cystic duct.

The patient's bile losses precluded her discharge home. Since it was anticipated that it would take several months for her hemangioma to show signs of spontaneous involution an internal biliary bypass was planned. She therefore underwent a hepaticojejunostomy using a 35 cm Roux-en-y loop of jejunum. Thereafter, she recovered rapidly and uneventfully with complete resolution of her jaundice. Two years later she remains well with normal biochemical liver function and no clinical or biochemical evidence of pancreatic insufficiency. The pancreatic mass is no longer detectable on ultrasound imaging.

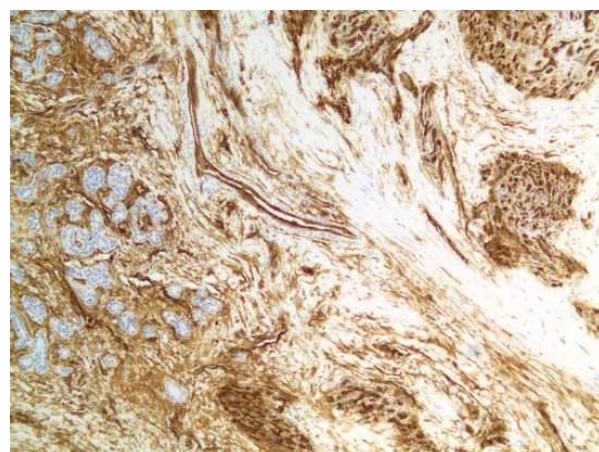


Figure 5. The lobules of capillaries are highlighted by immunohistochemistry using the endothelial cell marker CD34. Note the negative staining of pancreatic glands to the left. (CD34 immunoreactivity, x100).

DISCUSSION

Infantile hemangiomas, are vascular tumors composed of blood vessels lined by mitotically active endothelial cells. Unlike vascular malformations which grow with the individual, hemangiomas tend to undergo proliferation during infancy followed by a period of slow involution lasting several years and eventual regression leaving a fibro-fatty residuum [1]. They may occur in any region of the body but have a predilection for the head, neck and trunk. Treatment is variable and a conservative approach is often justified

because of their natural history and benign tendency. However, complications due to their size or site may warrant medical or surgical intervention. Visceral hemangiomas may occur in isolation or coexist with cutaneous lesions [3, 4].

Pancreatic hemangiomas in children are very rare. Of more than 5,000 children in the vascular anomalies database at the Children's Hospital Boston, only two had an infantile hemangioma of the pancreas [5]. Only 9 children with a pancreatic 'infantile' hemangioma/hemangioendothelioma have been reported in the English literature (Table

Table 1. Pediatric pancreatic hemangiomas in the English literature.

Case	Year	Authors	Age at presentation	Sex	Presenting features
#1	1973	Chappell [6]	11 months	M	Jaundice, hepatomegaly
#2	1976	Tunell [7]	3 months	M	Jaundice, hepatosplenomegaly, abdominal mass, Kasabach-Merritt syndrome
#3	1985	Horie <i>et al.</i> [8]	3 years	M	Jaundice, hepatomegaly
#4	1987	Sauer <i>et al.</i> [9]	5 months	F	Jaundice, hepatomegaly
#5	2003	Tebboune <i>et al.</i> [10]	2 months	F	Jaundice, hepatomegaly
#6	2003	Tebboune <i>et al.</i> [10]	18 months	M	Gastrointestinal bleeding, splenomegaly
#7	2005	Hibi <i>et al.</i> [11]	2 years	M	Jaundice
#8	2006	Vogel <i>et al.</i> [5]	2 months	F	Gastrointestinal bleeding
#9	2006	Vogel <i>et al.</i> [5]	2 years	M	Jaundice and abdominal pain

Table 1. (Continues)

Case	First line treatment	Subsequent treatment	Pathological description
#1	T-tube choledochotomy	Pancreaticoduodenectomy	Hemangioendothelioma in head of pancreas
#2	Prednisone	Gastrojejunostomy and cholecystojejunostomy (later reversed); radiotherapy	Hemangioendothelioma in head of pancreas
#3	Choledochojejunostomy	None	Hemangioendothelioma in head of pancreas
#4	Prednisone and biopsy	Prolonged percutaneous transhepatic biliary stenting and external drainage	Hemangioendothelioma in head of pancreas
#5	Percutaneous transhepatic external biliary drainage	Hepaticojejunostomy	Hemangioendothelioma in head of pancreas
#6	Percutaneous needle biopsy and observation	None	Capillary hemangioma in head of pancreas
#7	Percutaneous transhepatic gallbladder drainage	Roux-en-Y choledochojejunostomy	Capillary hemangioma in head of pancreas
#8	Corticosteroids	None	Infantile hemangioma
#9	Corticosteroids	Temporary percutaneous transhepatic external biliary drainage	Infantile hemangioma

1) [5, 6, 7, 8, 9, 10, 11] but the true incidence of the condition may well be higher since similar lesions in the body and tail of the gland are likely to be asymptomatic. In these reports, the terms hemangioma and hemangioendothelioma appear to have been used interchangeably to describe the same clinical entity. Presenting features have included obstructive jaundice, hepatomegaly, a palpable mass, duodenal obstruction, and intestinal bleeding. In one case, the lesion was first detected by prenatal ultrasonography at 26 weeks' gestation [10]. Cutaneous hemangiomas were present in only one patient [5]. All tumors were located in the head of the pancreas. Table 1 does not include children with more extensive retroperitoneal hemangioendotheliomas involving multiple organs including the pancreas, all of whom also had the Kasabach-Merritt syndrome [4, 5, 12, 13, 14]. The latter is now recognized to be a feature of the more aggressive Kaposiform hemangioendothelioma rather than the typical common infantile hemangioma [2]. One other infant who died from cardiac failure and was found at autopsy to have multiple cutaneous and visceral hemangiomas, including three on the surface of the pancreas, was also excluded [3]. One of the 9 children with an infantile hemangioma arising from the pancreas (Table 1) had the Kasabach-Merritt syndrome but it is difficult to be certain about the exact histology of this case reported in 1976 [7].

Ultrasound imaging of a pancreatic hemangioma demonstrates a mass in the pancreas frequently associated with biliary obstruction. Portal vein stenosis and portomesenteric vascular thrombosis have been reported [8, 10]; our patient had extrinsic compression of the portal vein (Figure 2). MRI usually demonstrates more definitive features of a hemangioma i.e. a lobulated mass with a moderately high intensity signal on T2 weighted images and marked enhancement after intravenous gadolinium [5, 11]. MRCP sequences help to define associated biliary obstruction. In our patient, tumor enhancement after gadolinium administration was delayed. This atypical

feature may have been due to the unusually dense sclerosis within the tumor. Conventional arteriography has been described as an adjunct to diagnosis but does not provide any additional useful information unless selective embolization of feeding vessels is being considered [5, 10].

The treatment of pancreatic hemangiomas in childhood is variable and depends in part on the presence of associated biliary obstruction. Some tumors have been resected [4, 6] but this is unnecessary. An adequate biopsy of tumors at this site is important to eliminate any doubt about the diagnosis. Treatment with corticosteroids or alpha interferon may be successful but tumor response to therapy is variable [4, 5, 6, 9, 14]. Vincristine has been used to treat the more aggressive Kaposiform hemangioendothelioma involving the pancreas [5]. One report described successful percutaneous transhepatic biliary stenting and external drainage in a 5-month-old infant pending spontaneous tumor regression; the infant required three catheter changes during a 22-month period [9]. A similar attempt at percutaneous transhepatic external drainage in another infant had to be abandoned following the development of septic complications and the patient required an emergency hepaticojejunostomy [10]. Other reports have described internal surgical bypass procedures using cholecystojejunostomy or choledochojejunostomy [6, 7, 8, 11].

Our patient underwent an open biopsy of the pancreatic mass to provide sufficient tissue for diagnosis. This was considered especially important because of the slightly atypical features of the tumor on imaging. The obstructive jaundice was initially managed by a tube cholecystostomy which we hoped would provide satisfactory external biliary drainage. However, fluid and electrolyte losses prevented this being a sustainable long term option and a straightforward hepaticojejunostomy was therefore performed. This avoided treatment with corticosteroids, vincristine or interferon, all of which have a significant risk of side-effects. If the expertise is available, an alternative option worth considering is temporary endoscopic biliary

stenting but this would require regular stent changes with the risk of cholangitis and post-procedural pancreatitis. Cholecystojejunostomy was considered in our patient but was rejected for two reasons. Firstly, pancreatic hemangiomas in the head of the pancreas can cause cystic duct obstruction in infants [6, 9]. Secondly, we were concerned that our patient's narrow tortuous cystic duct might cause problems in sustaining satisfactory biliary diversion during the period of involution of the hemangioma which may take several years [6, 11].

In conclusion, pancreatic hemangiomas are rare in children and often present with biliary obstruction. The tumor does not require resection because the natural history is that of spontaneous involution. Associated biliary obstruction can be managed in various ways and, to some extent, the approach depends on institutional expertise. In our patient, surgical internal biliary diversion led to a good outcome.

Received July 13th, 2006 - Accepted July 27th, 2006

Keywords Hemangioma; Jaundice, Obstructive; Pancreas

Abbreviations MIP: maximum intensity projection

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References

1. Takahashi K, Mulliken JB, Kozakewich HP, Rogers RA, Folkman J, Ezekowitz RA. Cellular markers that distinguish the phases of hemangioma during infancy and childhood. *J Clin Invest* 1994; 93:2357-64. [PMID 7911127]

2. Sarkar M, Mulliken JB, Kozakewich HP, Robertson RL, Burrows PE. Thrombocytopenic coagulopathy (Kasabach-Merritt phenomenon) is associated with Kaposiform hemangioendothelioma and not with common infantile hemangioma. *Plast Reconstr Surg* 1997; 100:1377-86. [PMID 9385948]

3. Burman D, Mansell PWA, Warin RP. Miliary haemangiomas in the newborn. *Arch Dis Child* 1967; 42:193-7. [PMID 6024468]

4. Schulz AS, Urban J, Gessler P, Behnisch W, Kohne E, Heymer B. Anaemia, thrombocytopenia and coagulopathy due to occult diffuse infantile haemangiomatosis of spleen and pancreas. *Eur J Pediatr* 1999; 158:379-83. [PMID 10333119]

5. Vogel AM, Alesbury JM, Fox VL, Fishman SJ. Complex pancreatic vascular anomalies in children. *J Pediatr Surg* 2006; 41:473-8. [PMID 16516618]

6. Chappell JS. Benign hemangioendothelioma of the head of the pancreas treated by pancreaticoduodenectomy. *J Pediatr Surg* 1973; 8:431-2. [PMID 4715749]

7. Tunell WP. Hemangioendothelioma of the pancreas obstructing the common bile duct and duodenum. *J Pediatr Surg* 1976; 11:827-30. [PMID 993954]

8. Horie H, Iwasaki I, Iida H, Takizawa J, Itoh F, Kohda S. Benign hemangioendothelioma of the pancreas with obstructive jaundice. *Acta Pathol Jpn* 1985; 35:975-9. [PMID 4072679]

9. Sauer L, Harrison MR, Bond SJ, Flake AW, Heyman MB, Ring EJ. Long-term percutaneous biliary drainage in an infant with hemangioendothelioma. *J Pediatr Surg* 1987; 22:606-8. [PMID 3612453]

10. Tebboune N, Lazure T, Fabre M, Pariente D. Pancreatic haemangioma in infancy: the place of radiology. *Pediatr Radiol* 2003; 33:621-3. [PMID 12879316]

11. Hibi M, Tokiwa K, Fukata R, Nakajima F. Obstructive jaundice in a child with pancreatic hemangioma. *Pediatr Surg Int* 2005; 21:752-4. [PMID 16047179]

12. Niedt GW, Greco MA, Wiczorek R, Blanc WA, Knowles DM 2nd. Hemangioma with Kaposi's sarcoma-like features: report of two cases. *Pediatr Pathol* 1989; 9:567-75. [PMID 2813203]

13. Tsang WY, Chan JK. Kaposi-like infantile hemangioendothelioma. *Am J Surg Pathol* 1991; 15:982-9. [PMID 1928553]

14. Shih SL, Chen BF, Chen SH, Chi T, Sheu CY. Spindle cell hemangioendothelioma of the pancreas treated with interferon alpha 2a. *Pancreas* 1998; 16:215-6. [PMID 9510148]