Pediatr Radiol (1991) 21: 152–153

Pediatric Radiology



# Sarcoma botryoides of the common bile duct: preoperative diagnosis by coronal CT and PTC

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Received: 27 February 1990; accepted: 11 June 1990

**Abstract.** Sarcoma botryoides of the extrahepatic bile ducts is a rare cause of obstructive jaundice in the pediatric population. It is rarely diagnosed preoperatively. We present a case of this tumor which was diagnosed by ultrasound, computerised tomography and PTC. Coronal CT sections were particularly useful in demonstrating the relationship of the tumor to the porta hepatis, pancreas and duodenum.

Rhabdomyosarcoma is the commonest primary malignancy of the bile ducts in childhood [1]. In the extrahepatic biliary system it usually forms a botryoid mass resembling a cluster of grapes [1, 2]. This characteristic morphology allowed us to make a preoperative diagnosis of this extremely rare tumor utilising coronal computerised tomography (CT) and percutaneous transhepatic cholangiography (PTC).

## Case report

A 4-year-old boy, one of monozygotic twins, was admitted for the investigation of fever, progressive jaundice, abdominal distension, loss of appetite and fatigue which had developed over a three week period. His past history was uneventful and his psychomotor development normal.

On examination, the child was alert and in discomfort. Jaundice and pallor were noted. There was fever of 39.8°C. The abdomen was markedly distended and a firm tender liver could be palpated 8 cm below the costal margin. There was moderate splenomegaly and no evidence of ascites.

Laboratory tests demonstrated markedly abnormal liver function tests including a total bilirubin of 110 mol/l and a direct bilirubin of 88 mol/l. The hemoglobin was 11.8 gm% and the white cell count 10,800/mm$^3$ with 35% lymphocytes, 57% granulocytes and 8% monocytes. The erythrocyte sedimentation rate was 110 mm in the first hour. Blood cultures grew Hemophilus-influenzae.

Ultrasound demonstrated a markedly dilated extrahepatic biliary system containing echogenic material interspersed with small echofree areas (Fig. 1). The liver parenchyma was homogenous with dilated intrahepatic bile ducts which did not contain any echogenic material. Axial CT scans after the administration of intravenous contrast material demonstrated an $8 \times 5.5 \times 4.5$ cm mass arising at the porta hepatis and extending caudally to the level of the head of the pancreas. The mass had a lower attenuation value than normal liver. Small areas of even lower attenuation were seen within this mass (Fig. 2) 5 mm thick coronal slices through the area demonstrated the mass to be within a markedly dilated common bile duct (Fig. 3). PTC demonstrated massive dilatation of the distal common hepatic duct and the common bile duct. These contained large filling defects with contrast filling the interstices between the tumor masses (Fig. 4).

Following two weeks of intravenous antibiotic therapy for Hemophillus-influenzae bacteremia, the child underwent surgery. At laparatomy, markedely dilated common hepatic and common bile ducts were found. These were filled with the typical grape-like lesions of sarcoma botryoides extending from the confluence of the two main hepatic ducts down to within half a centimeter of the papilla of Vater. No liver metastases were seen or palpated. The common hepatic duct, common bile duct and the gallbladder were resected and a Roux-en-Y jejunal segment interposed in the form of a hepaticojejunostomy. On pathological examination, a diagnosis of embryonal rhabdomyosarcoma was made. There was microscopic invasion of the wall of the common bile duct and two cystic lymph nodes and several scattered foci of tumor within the gallbladder wall.

The post operative course was uneventful and the patient was referred for adjuvant therapy.

## Discussion

Obstructive jaundice in childhood is rare. Choledochal cyst, rhabdomyosarcoma and choledocholithiasis are the commonest causes after the newborn period when biliary atresia accounts for most cases [3]. Rhabdomyosarcoma arising from the intrahepatic biliary tree usually presents late with a large hepatic mass which has no distinguishing features on ultrasound or CT. These tumors do not usually cause obstructive jaundice [1]. Rhabdomyosarcoma arising from the extrahepatic biliary tree is more likely to result in obstructive jaundice usually preceeded by malaise, abdominal pain and fever. These tumors tend to form a botryoid mass consisting of tumor masses interspersed with lakes of bile [1, 2].

With surgical resection alone, the prognosis of this tumor has been uniformly poor [4]. Aggressive use of chemotherapy and radiotherapy in addition to resection has resulted in several cases of disease free survival [5].

Since most of the reported cases of this entity occured before the availability of ultrasound, CT or PTC, there have been few reports which included details of preoperative imaging. Some early reports included operative cholangiography demonstrating filling defects within the extrahepatic bile ducts [2, 6] and in a more recent report the diagnosis was established preoperatively using ultrasound, axial CT and PTC [7].

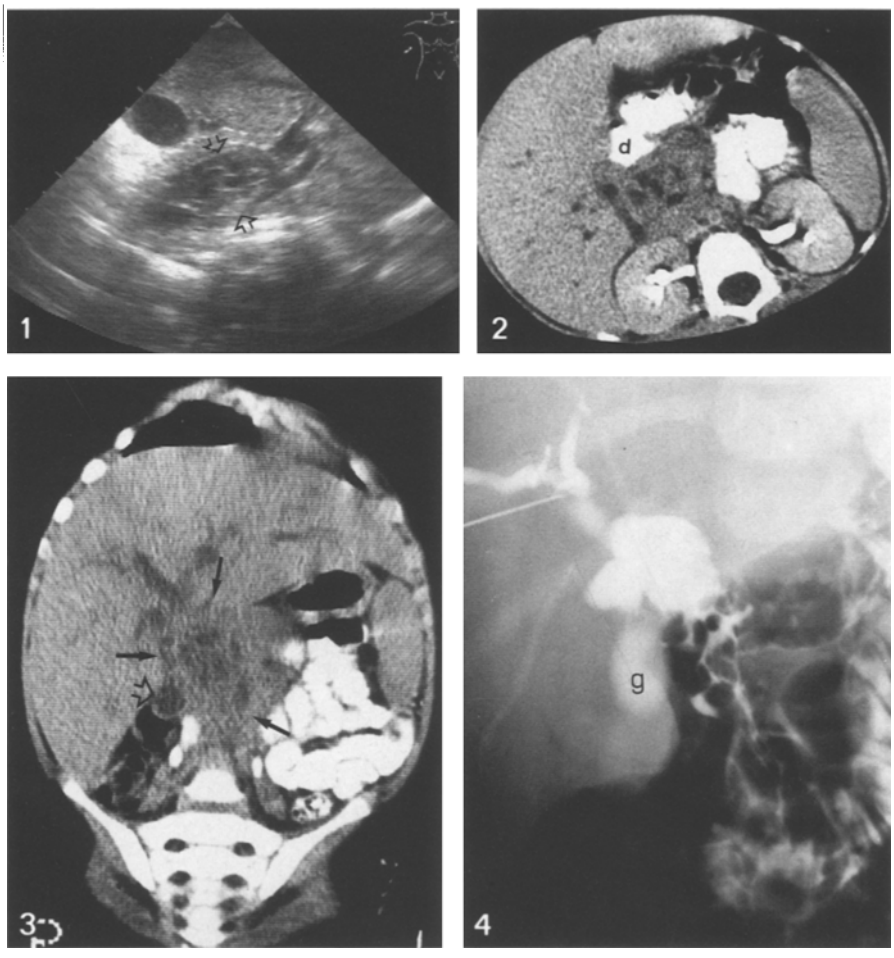


**Fig. 1.** Right parasagittal ultrasound scan demonstrating massively dilated extrahepatic bile ducts filled with tumor *(arrows)* and echo-free bile lakes

**Fig. 2.** Axial CT scan demonstrating tumor filled extrahepatic bile ducts behind the duodenal cap (d). Low attenuation bile lakes are seen within the tumor mass

**Fig. 3.** Coronal CT scan through the porta hapatis and head of pancreas demonstrating the tumor mass *(arrows)* and its precise relationship to the surrounding structures. The gallbladder *(hollow arrow)* is seen lateral to the tumor

**Fig. 4.** Antegrade cholangiogram via a right hepatic duct demonstrating markedly dilated common hepatic and common bile ducts with free passage to the duodenum and filling of the gallbladder (g). Multiple grape-like filling defects are present in the common hepatic and common bile ducts. Notice the similarity between this appearance and the coronal CT (Fig. 3)

In our case, the ultrasound, CT and cholangiographic appearances corresponded closely to the gross pathology of the tumor which showed the affected bile ducts to be markedly dilated by masses of solid tumor interspersed with lakes of bile. On ultrasound, small echo-free bile lakes were seen between the echogenic tumor masses (Fig. 1). Contrast enhanced axial and coronal CT demonstrated the tumor to have a lower attenuation value than normal liver. Within the tumor there were small areas of lower attenuation corresponding to bile lakes (Figs. 2, 3). To our knowledge this is the first case of this tumor that has been studied by coronal CT. The coronal sections provided much clearer delineation of the relationship of the tumor to the porta hepatis, pancreas and duodenum and bore a striking resemblence to the images obtained by PTC. The sections were obtained by laying the patient supine in a semi-sitting position, his back elevated 50° and with the gantry angled 30° cranially [8]. Coronal CT can also be performed with the patient lying prone with his chest elevated and the gantry angulated cranially [9].

Detailed preoperative assessment is of great value in establishing the diagnosis, determining resectability and planning surgery.

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