

bone cyst, and both hearing loss and aneurysmal bone cyst were results of the initial bacterial meningitis. The association between bacterial meningitis and hearing loss is well known. Bacterial infection as an aetiological factor in the development of an aneurysmal bone cyst may be due to an accompanying vasculitis resulting in local ischaemia and growth disturbance of the bone as mentioned above. Proof of whether either of our theories are true awaits further observations.

Even if the diagnosis of an aneurysmal bone cyst can be made by radiography, this diagnostic method is not reliable. 'Typical' signs on radiography have been described by several authors,^{2,3} for example soap bubble or blow out appearance (see above), eggshell type calcification, or lytic skull lesions, but the morphological changes vary considerably from patient to patient.^{2,3} Moreover, in cases of skull base cysts, plain radiographs of the skull and even axial computed tomograms are of little diagnostic value. Only coronal thin section computed tomograms will provide diagnostic clues, as shown in our patient. We therefore recommend performing these scans in every case of suspected malformation of the

skull base and in the diagnosis of aetiologically obscure recurrent bacterial meningitis.⁷

Because of the possible coincidence with malignant tumours,² destructive growth of the lesion *per se*, and the high recurrence rate of nearly 21%,³ total surgical dissection of the cyst should be the primary therapeutic goal.

We thank surgeons Professor Dr W Draf, head of the department of ear, nose, and throat, head, neck, and facial reconstructive surgery, Fulda, and Professor Dr H P Richter, then head of the department of neurosurgery, Fulda.

- 1 Jaffe HL, Lichtenstein L. Solitary unicameral bone cyst: with emphasis on the roentgen picture, the pathologic appearance, and the pathogenesis. *Arch Surg* 1942; **44**: 1004-25.
- 2 Wachter W, Adler CP. Aneurysmale Knochenzyste des Os temporale. *Laryngorhinootologie* 1984; **63**: 622-5.
- 3 Keuskamp PA, Horoupian DS, Fein JM. Aneurysmal bone cyst of the temporal bone presenting as a spontaneous intracerebral hemorrhage: case report. *Neurosurgery* 1980; **7**: 166-70.
- 4 Delorit GJ, Summers GW. Aneurysmal bone cyst of the sphenoid sinus. *Transactions of American Academy of Ophthalmology and Otolaryngology* 1975; **80**: 438-43.
- 5 Revuelta M, Jos VL, Trujillo F, Albert P. Dos casos raros de tumor de la base del cráneo: osteoclastoma y quiste oseo aneurismático. *Revista Española Oto-Neuro-Oftalmología Y Neurocirugía* 1975; **33**: 19-23.
- 6 Yee RD, Cogan DG, Thorp TR, Schut L. Optic nerve compression due to aneurysmal bone cyst. *Arch Ophthalmol* 1977; **95**: 2176-9.
- 7 Draf W. Malformations of the skull base. In: Pfeifer G, ed. *Craniofacial abnormalities and clefts of the lip, alveolus and palate*. Stuttgart: Thieme, 1991: 40-3.

Hepatic abscess in sickle cell anaemia: a rare manifestation

Meera Lama

Abstract

A child with sickle cell anaemia developed a hepatic abscess, which was managed successfully by percutaneous drainage under ultrasound control. A history of attacks of pain dissimilar to usual vaso-occlusive crises should be treated with suspicion and investigated appropriately.

(*Arch Dis Child* 1993; **69**: 242-243)

Hepatic abscess as a complication of sickle cell anaemia has been described only once before in childhood.¹ The first case managed successfully by percutaneous drainage guided by ultrasound is reported here.

Case report

A 10 year old Saudi girl, known to have sickle cell anaemia, presented to the casualty department with a one month history of recurrent right abdominal pain and intermittent fever. She had been seen repeatedly during this time and was thought to be suffering from mild abdominal vaso-occlusive crises and sent home on analgesics, with little response.

The casualty officer had considered the child was probably using the pain as an excuse for

missing school. Interestingly though, this was the child's first presentation with abdominal pain and she was emphatic that it was totally dissimilar to any previous vaso-occlusive episodes.

On examination she looked unwell and was feverish (39°C), in severe abdominal pain, and pale with a tinge of jaundice. Abdominal examination revealed tender hepatomegaly (5 cm below the costal margin in the mid-clavicular line).

Initial investigations included a haemoglobin concentration of 62 g/l, a raised white cell count ($27.3 \times 10^9/l$), mainly neutrophils with a left shift, and a platelet count of $927 \times 10^9/l$. Blood urea and electrolyte concentrations were within normal limits. Chest and upper abdominal x ray films showed abnormal elevation of the right cupola of the diaphragm with some right basal reaction (fig 1). Abdominal ultrasound confirmed the presence of an abscess in the right lobe of the liver - showing as two hypoechoic areas containing some necrotic elements, lying approximately 8.5 cm from the surface (fig 2).

MANAGEMENT

The child was commenced on intravenous fluids, analgesics, and broad spectrum anti-

Department of Paediatrics, National Guard King Khalid Hospital, Jeddah, Saudi Arabia

Correspondence to: Dr Meera Lama, Department of Paediatrics, East Birmingham Hospital, Bordesley Green East, Birmingham B9 5SS.

Accepted 29 April 1993

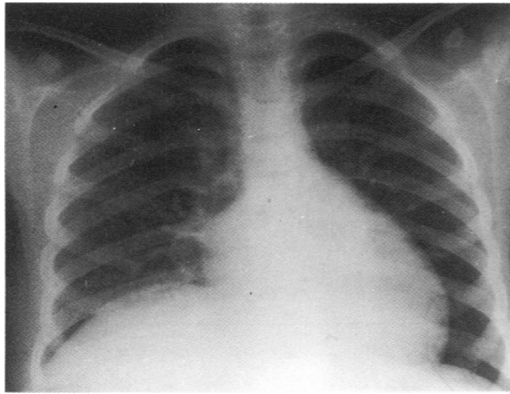


Figure 1 Radiograph of chest and upper abdomen.

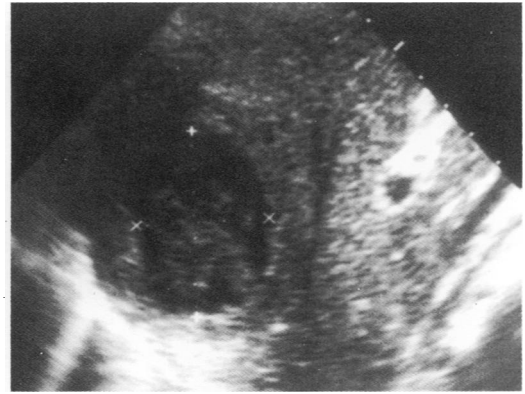


Figure 2 Ultrasound scan of the abdomen.

biotics (intravenous cefotaxime and metronidazole).

As a result of her poor general condition she was transfused to raise her haemoglobin to 100 g/l in an attempt to reduce circulating concentrations of haemoglobin S. After her condition stabilised, 100 ml of yellow fluid was drained from the liver by percutaneous drainage under ultrasound guidance, using a short acting anaesthetic. Culture of the aspirated fluid subsequently grew a scanty growth of bacteroides sensitive to penicillin and metronidazole.

PROGRESS

The child's general condition improved dramatically over the next two days. The fever subsided, the liver regressed to 2 cm, and the tenderness disappeared. Antibiotics were discontinued after 10 days and the child discharged home in good general condition. Follow up by serial ultrasound examination revealed almost complete obliteration of the abscess cavity after six weeks.

Discussion

Children with sickle cell anaemia may suffer a variety of hepatobiliary abnormalities.² These range from biochemical abnormalities, including raised conjugated bilirubin concentrations and transaminases, to clinically evident hepatomegaly, hepatic crisis, cholelithiasis and cholecystitis, progressing often in adulthood to hepatic fibrosis and hepatic cirrhosis. Rare presentations include a hepatic biloma³ – an intrahepatic bile filled cyst – and hepatic abscess.

Patients with sickle cell anaemia are prone to frequent episodes of vaso-occlusive crisis and infarction secondary to sludging associated with increased viscosity of hypoxic haemoglobin S rich blood. Such episodes are usually precipitated by infection, acidosis, dehydra-

tion, and hypoxaemia and can occur in any tissue.

Infarctive crises in the liver have been reported in about 10% of patients with sickle cell disease. Hepatic crises are relatively rare, perhaps because the liver is protected against sickling by its dual blood supply.

Infection may reach the liver via the hepatic artery, portal vein, lymphatics, or biliary tree. In the majority of cases, however, abscesses are cryptogenic.

I postulate that the liver abscess in this patient represents a secondary infection in a hepatic infarct because:

- (1) Patients with sickle cell anaemia are prone to infarction episodes.
- (2) Primary liver abscesses are very rare.
- (3) Hepatic infarcts are likely to become secondarily infected and produce an abscess.

Infarcted liver may become infected for a number of reasons:

- (1) Enteric organisms including commensals like bacteroides may invade the portal system because of compromise of gastrointestinal integrity after mucosal capillary thrombosis.
- (2) Functional asplenia, low IgM concentrations, and defective antibody response.
- (3) Impaired opsonin phagocytic function and defective complement pathways may impair effective removal of bacteria from the blood stream.

In sickle cell disease, abdominal crises are fairly frequent but persistence of symptoms and, more importantly, history of pain dissimilar to previous infarctive/vaso-occlusive episodes should alert the paediatrician to look more closely for a specific underlying cause.

- 1 Shulman ST, Beem MO. A unique presentation of sickle cell disease: pyogenic hepatic abscess. *Pediatrics* 1971; **47**: 1019–21.
- 2 Schubert TT. Hepatobiliary system in sickle cell disease. *Gastroenterology* 1986; **90**: 2013–21.
- 3 Middleton JP, Wolper JC. Hepatic biloma complicating sickle cell disease. *Gastroenterology* 1984; **86**: 743–4.