

2. Levy HL. Inborn errors of carbohydrate metabolism. *In: Schaffer's Diseases of the Newborn*, 5th edn. Eds Avery MA, Taeusch Jr MW. Philadelphia, WB Saunders Co, 1984, pp 520-534.
3. Purohit KR, Dwaraki Bai Y. A case of galactosemia. *Indian Pediatr* 1967, 4: 454-455.
4. Anand VR, Sindhi KC. Kinase defect in galactosemia. *Indian Pediatr* 1971, 14: 379-380.
5. Kalra SK, Kalra AS, Sarin NP, Sarkar P. Galactosemia. *Indian Pediatr* 1979, 14: 1143-1144.

Hydrops of the Gall Bladder in a Child with Wilson's Disease: A Rare Association

Z. Karrar
P.J. Patel
M.B. Abdurrahman

Hydrops of the gall bladder is an uncommon condition in childhood. It has been described in preterm infants and newborns secondary to parenteral hyperalimentation, sepsis as well as in normal

From the Department of Pediatrics and Radiology, College of Medicine, King Saud University, Riyadh, Saudi Arabia.

Reprint requests: Dr. P.J. Patel, Associate Professor, Radiology Department (40), P.O. Box 2925, Riyadh 11461, Kingdom of Saudi Arabia.

*Received for publication October 16, 1990;
Accepted December 10, 1990*

newborns(1-3), older infants and children. It has been reported in association with upper respiratory tract infections, gastroenteritis and enteric infections, leptospirosis, scarlet fever, lymphadenitis, and mucocutaneous syndrome, polyarteritis nodosa and familial mediterranean fever(4-8).

We report a case of asymptomatic hydrops of the gall bladder in an 11-year-old Saudi Girl with hepatic cirrhosis secondary to Wilson's disease.

Case Report

A 12-year-old Saudi girl was admitted to hospital for re-evaluation because of recurrent hematuria, abdominal pain and occasional joint pains and ankle swellings.

She had been evaluated extensively in another hospital in Riyadh an year ago and referred for further evaluation and management in the United Kingdom, with a provisional diagnosis of glomerulonephritis. In the United Kingdom in addition to her suspected renal disease she was found to have a disturbed clotting profile, raised hepatic enzymes and a positive HbsAg screening test. Because of her clotting disturbances, liver and renal biopsies were not done, and a provisional diagnosis of chronic active hepatitis with possible immune complex renal disease was made. An abdominal ultrasound at that time was reported as showing a normal gall bladder.

There was no history of jaundice or neurological symptoms. Her parents are cousins and she has 5 siblings who are living. There was no family history of liver disease or neurological disease.

On physical examination she was pale, without jaundice or obvious Kayser Fleischer (KF) ring. Her weight was 31 kg and height was 128 cm. She had pitting edema

over both ankles as well as the sacrum. Her BP was 110/60 mm of Hg. Abdominal examination revealed splenomegaly of 6 cm below the costal margin. The liver was not palpable, the upper margin was in the 6th intercostal space. There was no ascites.

Laboratory results showed a total and direct bilirubin of 258 and 8 mmol/L respectively, albumin 27 g/L, alkaline phosphates 806 units/L, alanine and aspartate aminotransferases 35 and 127 units/L, respectively and gammagluteryl transpeptidase of 61 units/L. The prothrombin and partial thromboplastin times were prolonged. Screening for hepatitis-B, TORCH, syphilis and autoimmune markers was negative. Alpha-1-antitrypsin level was normal. Her serum ceruloplasmin was markedly low (less than 7 mg/dl); 24 hours urinary copper excretion was high (93 mg) and was markedly enhanced to 1040 mg, after a challenge with penicillamine for 48 hours.

A liver biopsy showed marked cirrhosis with some copper deposition. A slit lamp examination confirmed the presence of KF rings bilaterally. Real time ultrasonography of the upper abdomen using 3.5 MHz linear array system revealed a dilated gall bladder (size 7 × 3.2 cm) without evidence of calculi or dilatation of the biliary tree. No other significant abnormality was demonstrated. (Fig. 1). Intravenous cholangiography showed distended gall bladder, confirming the ultrasonographic findings and excluding obstruction of the cystic duct (Fig. 2).

On the basis of her clinical, laboratory and pathological findings a diagnosis of Wilson's disease was made and she was started on penicillamine 2 g/day and Vitamin B₆. After discussion with the surgeons it was decided that no surgical intervention

was indicated because her symptoms were not related to the gall bladder findings.

She showed good response to treatment: her liver function tests improved and she lost her edema by 4 weeks. She did not show any symptom or sign of gall bladder disease and a repeat ultrasound 6 weeks later showed normal sized gall bladder (Fig. 3).

Discussion

Hydrops of gallbladder is rare in childhood. Recently, more cases have been described in preterm and sick newborns and it was suggested that the condition might be relatively common in that groups(2).

Among infants and other children the condition was described secondary to acalculous cholecystitis and in association with childhood infections, systemic diseases including salmonellosis, shigellosis, scarlet fever, mucocutaneous syndrome, familial mediterranean fever and polyarteritis nodosa(4-8). None of these conditions was suggested in our patient.

To the best of our knowledge this is the first case reported to have hydrops of the gall bladder in association with confirmed Wilson's disease. The benign course in our patient suggests that the condition is transient and self-limiting and supports reports suggesting that there is no need for cholecystectomy or drainage as advocated previously(1-6,8). It is not clear whether therapy with penicillamine is linked to this resolution.

Kumari in 1979 reported the first case to be diagnosed preoperatively by ultrasound(8). Since then, ultrasound has proven its value in assessing preterm infants and sick newborns with gall bladder distension(2,3). In asymptomatic children it is a valuable tool for safe and non-



Fig. 1. Ultrasound study showing a distended gall bladder (7 × 3.2 cm).



Fig. 3. Ultrasound study six weeks later showing resolution of the gall bladder distension.

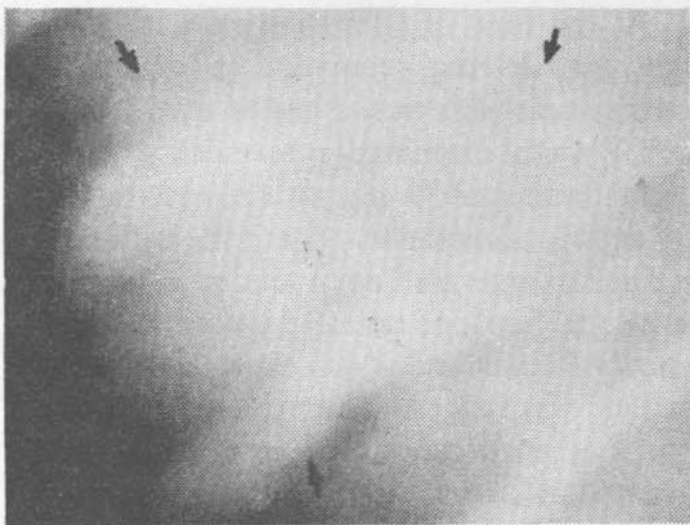


Fig. 2. Intravenous cholangiogram showing a distended gall bladder.

invasive follow-up method that will save the child unnecessary surgical intervention.

REFERENCES

1. Neu J, Arvin A. Hydrops of the gall bladder. *Am J Dis Child* 1980, 134: 891-893.
2. Saldanha RL, Stein Ca, Kopelman AE. Gall bladder distension in ill preterm infants. *Am J Dis Child* 1983, 137: 1179-1180.
3. Liechty EA, Cohen MD, Lemons JA, *et al.* Normal gall bladder appearing as abdominal mass in neonates. *Am J Dis Child* 1982, 136: 468-469.
4. Bloom RA, Swain VA. Non-calculous distension of the gall bladder in childhood. *Arch Dis Child* 1966, 41: 503-508.
5. Chamberlain JW, Hight DW. Acute hydrops of the gallbladder in childhood. *Surgery* 1970, 68: 899-905.

6. Ternberg LJ, Keathing JP. Acute acalculous cholecystitis complication of other illnesses in childhood. *Arch Surg* 1975, 110: 543-547.
7. Barth RA, Brasch RC, Fill RA. Abdominal pseudotumor in childhood. *AJR* 1981, 136: 341-343.
8. Kumari S, Lee WJ, Baron MG. Hydrops of the gall bladder in a child—diagnosis by ultrasonography. *Pediatrics* 1979, 63: 295-297.

Acanthameba Meningoencephalitis Complicating Pyogenic Meningitis

S.C. Karande
K.R. Lahiri
S.S. Sheth
U.S. Nadkarni
M.K. Jain
M.D. Shah

Two genera of free-living amebae, *Naegleria* and *Acanthameba* are known to cause primary amebic meningoencephalitis (PAM) in man. Fowler and Carter in 1965(1) were the first to report 4 fatal cases of PAM. Despite their widespread distribution in soil, water, sewage and even

From the Department of Pediatrics, Seth G.S. Medical College and K.E.M. Hospital, Parel, Bombay 400 012.

Reprint requests: Dr. Keya R. Lahiri, B/10, Vijay Kunj, Jawaharlal Nehru Road, Santacruz (East), Bombay 400 055.

Received for publication October 2, 1990;

Accepted December 13, 1990

air, PAM is a very rare disease. Less than 120 cases have been reported in world literature, of which less than 30 are due to *Acanthameba*(2). From India, Pan *et al.*(3) and Gogate *et al.*(4) have reported 2 cases each, all 4 due to *Acanthameba*. The extreme rarity and unusual clinical presentation in our case, has prompted us to report it.

Case Report

On November 11, 1989, a 7 week-old-male infant was referred, from Baroda, for treatment of resistant ventriculitis. The child had developed pyogenic meningitis on the sixth day of life, following a normal vaginal hospital delivery. He responded well to 3 weeks treatment with intravenous antibiotics and other supportive measures. Four days later, the child was readmitted with complaints of poor sucking and convulsions. Although full details of treatment given were not available, the mother stated that 4 lumbar punctures and 2 ventricular taps were done during the two hospital admissions at Baroda.

At the time of admission with us, the baby was having recurrent generalized tonic spasms with partial loss of consciousness. Physical examination revealed a head circumference of 39 cm, with a tense bulging anterior fontanelle, absent menace reflex and exaggerated deep tendon reflexes. Investigations done revealed a normal total leucocyte count and X-ray chest. Ventricular fluid was suggestive of pyogenic meningitis, but no organisms were detected on Gram stain and routine culture (Table I). CT showed evidence of moderate hydrocephalus, with a hyperdense area in the right frontal region (Fig.). This mass lesion was thought to be a hematoma resulting from the earlier ventricular taps done at Baroda.