blood chemistry and absence of changes in the skull(5).

Although various measures like prednisolone, oral buffered phosphates and enzyme replacement therapy have been tried in this disease, there is no well established therapy(4,6). Death in the perinatal form occurs either *in utero* or few hours or at the most a few days after birth due to marked inability to ventilate the lungs properly in such patients(7). The unusual survival in the present case may have been due to the absence of severe involvement of the thoracic cage in the rachitic process.

REFERENCES

1. Rathbun JC. Hypophosphatasia, A new developmental anomaly. Am J Dis Child 1948, 71: 822-831.
2. Whyte MP. Hypophosphatasia. *In:* The Metabolic Basis of Inherited Disease, 6th edn, Scriver CR, Beaudet AL, Sly WS, Walle W. USA, McGraw Hill Information Services Co, 1989, pp 2843-2856.
3. Fraser D. Hypophosphatasia. Am J Med 1957, 22: 730-746.
4. Teree TM, Klein L. Hypophosphatasia: clinical and metabolic studies. J Pediatr 1968, 72: 41-50.
5. Kumar A, Kapoor R, Sethi GR, Saha MM, Chopra K, Mittal M. Metaphyseal dysostosis Schmidt type. Indian Pediatr 1988, 25: 1211-1215.
6. Whte MP, Valdis R Jr. Ryam LM, Mc Alister WH. Infantile hypophosphatasia: Enzyme replacement therapy by intravenous infusion of alkaline phosphatase rich plasma from patients with Paget bone disease. J Pediatr 1982, 101: 379-386.
7. Kellsey DC, Danville PA. Hypophosphatasia and congenital bowing of the long bones. JAMA, 1962, 179: 187-190.

Neonatal Adrenal Hemorrhage

R.H. Merchant
V.S. Sakhalkar
A.K. Karnik
M.S. Joshi
M.P. Desai

The relatively large size and vascularity of the neonatal adrenal gland makes them vulnerable to traumatic and asphyxial injuries, with incidence of adrenal hemorrhage at necropsies being 1.7 per 1000(1). Though the exact cause of neonatal adrenal hemorrhage (NAH) is unknown, postulated etiologies include maternal diabetes, obstetric trauma, asphyxia, thrombocytopenia and coagulation defects(2,3). Our recent encounter of four patients with NAH who were evaluated by ultrasound(USG) prompted this report. USG which is a safe, portable, noninvasive, accurate, useful tool for the diagnosis of NAH, was the only imaging modality employed for diagnosis and follow-up of our cases.

Case Reports

From June to November 1990, four male neonates with mean birth weight of 3450 g presented with a flank mass in the

From the Bai Jerbai Wadia Hospital for Children and Dr. Balabhai Nanavati Hospital, Bombay.

Reprint requests: Dr. R.H. Merchant, Honorary Professor of Pediatrics, Division of Neonatology, B.J. Wadia Hospital for Children, Parel, Bombay 400 012.

Received for publication: September 3, 1991; Accepted: December 4, 1991

first four days of life. All were term vaginal deliveries, except case 4 who was a preterm infant of a diabetic mother (IDM). Their salient features are summarized in the *Table*. Both neonates with birth asphyxia (Cases 2 and 4) had leucocytosis, of which the IDM also had hypoglycemia. Two had icterus within physiological limits and one had a ventriculoseptal defect with congestive cardiac failure (Case 3). Urine, hematocrit, clotting time, platelet count, prothrombin time, partial thromboplastin time were normal in all. USG abdomen showed a well defined suprarenal mass suggestive of NAH with a normal ipsilateral kidney in all. Follow-up revealed complete clinical and USG resolution within four months.

In all cases a tender flank mass, three on the right and one on the left, was detected on routine clinical examination within the first four days of life. Abdominal USG showed a well defined mass situated above the superior pole of a normal ipsilateral kidney, extending lateral to the kidney in two. In Cases 1 and 3, a predominatly hyperechoic solid lesion (*Fig. 1*) was suggestive of neuroblastoma. Serial USG in these two cases showed regression of mass with total resolution which confirmed this lesion to be NAH. The other two masses had heterogenous solid cystic echotexture. Follow-up within 30 days showed regression of lesion in all patients, with change to a cystic echotexture (*Fig. 2*). Complete regression was seen by 120 days in all, with normal adrenal configuration but with calcification in two (Cases 3 and 4). No other radiological investigation was done.

Discussion

NAH occurs most frequently between the second and seventh postnatal day. In the present series all four cases were detected within the first four days. Almost

TABLE—*Summary of Relevant Information of 4 Cases*

Case No.	Birth weight (g)	Salient clinical features & investigations	Ultrasound findings
1	3500	Tender Rt. flank mass PCV—0.60 Blood sugar—72 mg/dl Indirect serum bilirubin—123 μmol/L	Day 5 — Solid hyperechogenic mass (4.8 × 4.2 cm) Rt. suprarenal area, Rt. kidney normal Day 14 — Lesion smaller, predominantly cystic Day 120 — Lesion regressed completely
2	3400	Tender Rt. flank mass, seizures WBC—28 × 10^9/L USG skull—normal Blood sugar—75 mg/dl Indirect serum bilirubin—160 μmol/L	Day 8 — Mixed echogenic predominantly cystic mass (3.9×3.8 cm) Rt. suprarenal area Rt. kidney normal Day 100 — Lesion regressed completely
3	3400	Tender Lt. flank mass, VSD PCV—0.52 Blood sugar—60 mg/dl Indirect serum bilirubin—208 μmol/L	Day 4 — Mixed echogenic predominantly solid mass (5.2×4.6 cm) Lt. suprarenal area Day 30 — Mass smaller, predominantly cystic Lt. kidney normal Day 120 — Suprarenal dense echoes, posterior acoustic shadow indicating calcification
4	3500	Tender Rt. flank mass, bruises lower abdomen WBC—21×10^9/L PCV—0.46 Blood sugar—25 mg/dl Indirect serum bilirubin—144 μmol/L	Day 7 — Heterogenous predominantly cystic mass (3.3×3 cm) Rt. suprarenal area Rt. Kidney normal Day 14 — Mass smaller, cystic echoes Day 100 — Suprarenal dense echoes, posterior acoustic shadow indicating calcification.

PCV-Packed cell volume; VSD-Ventricular septal defect; WBC-White blood cells.

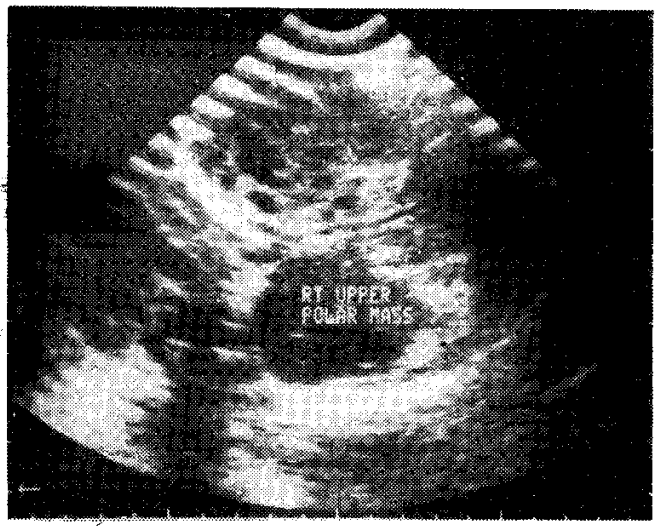

Fig. 1. Longitudinal upper abdominal sonogram showing a hyperechoic mass above right kidney representing an acute adrenal hemorrhage.

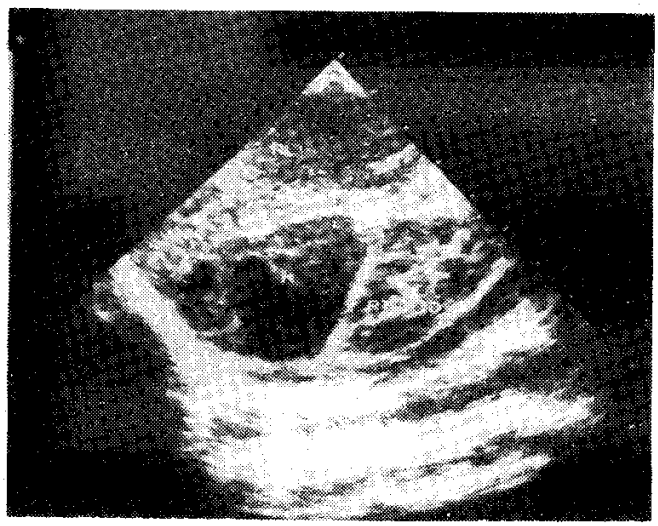

Fig.2. Sonogram of same patient 30 days later now demonstrates the suprarenal mass to be predominantly sonolucent.

70% NAH occurs on the right side with bilateral involvement in 5-10% cases(4). The difference in susceptibility to hemorrhage between the two sides appears to lie in their venous drainage. The right adrenal vein drains directly into the inferior vena cava (IVC), thus the right adrenal would suffer severe venous engorgement as a result of rise in IVC pressure. The left adrenal vein joins the left renal vein thus supplying the left adrenal with alternative drainage channels in event of fluctuations in IVC pressure occurring during delivery(4). In our series three had right sided NAH. Large neonates who undergo abdominal compression and consequently increased IVC pressure during vaginal or stressed delivery(3) are at greater risk of developing NAH.

The diagnosis of NAH is often difficult. It is important to distinguish it from renal vein thrombosis in which there is a variable degree of hematuria and proteinuria which were not noted in our cases. Small to moderate unilateral adrenal bleeds are usually asymptomatic as in our series. Large unilateral or bilateral extensive bleeds may occasionally present with jaundice, anemia, hypoglycemia, shock and adrenal insufficiency(2,3) none of which were observed in this series. Preponderance of subcapsular hemorrhage explains the rarity of Addisonian crisis in NAH(3).

Before USG was introduced various invasive and expensive procedures were utilized for the diagnosis and follow-up of NAH(4). In our cases the initial diagnosis as well as the follow-up was by USG. A fresh clot following active bleeding is visualized on USG as a hyperechoic solid mass which may closely resemble neuroblastoma(5) as in Cases 1 and 3. The cystic appearance of liquified NAH with or without low level echoes, should be distinguished from adrenal abscess, neonatal urinoma, cortical renal cyst and cystic neuroblastoma(5). Later, calcification in the resolved hematoma leads to a bright echogenic gland with posterior shadowing as in Cases 3 and 4. Treatment depends upon the severity of illness; blood, intravenous fluids and hydrocortisone may be required in large bleeds to combat shock and Addisonian crisis.

Judicious re-examinations documenting the sequential changes in echogenicity within the hematoma with its ultimate resolution is virtually diagnostic of NAH and prevents unnecessary, expensive and invasive investigations and surgical intervention. USG has hence become the modality of choice both for the initial diagnosis and clinical follow-up of these lesions(6).

REFERENCES

1. DeSA DJ, Nicholls S. Hemorrhagic necrosis of the adrenal gland in perinatal infants: A clinicopathological study. J Pathol 1972, 106: 133-149.
2. Lawson EE, Teele RL. Diagnosis of adrenal hemorrhage by ultrasound. J Pediatr 1978, 92: 423-426.
3. Black J, Williams DI. Natural history of adrenal hemorrhage in the newborn. Arch Dis Child 1973, 48: 183-190.
4. Gwinn JL, Barnes Gr. Radiological case of the month: Massive adrenal hemorrhage in the newborn. Am J Child 1965, 109: 259-260.
5. Wu C. Sonographic spectrum of neonatal adrenal hemorrhage: Report of a case simulating solid tumor. J Clin Ultrasound 1989, 17: 45-49.
6. Perry M, Kaftori JK, Bar-Maor JA. Sonography for diagnosis and follow-up of neonatal adrenal hemorrhage. J Clin Ultrasound 1981, 9: 397-401.

Kala Azar – Diagnostic Dilemma

T.S. Raghu Raman
S.A.M. Ahuja
K.V.S. Mani
Y.P. Jalpota
S.K. Adaval

There has been sudden resurgence of kala azar in recent times. Most of the children present no difficulties in diagnosis. However it is important to be aware of atypical clinical manifestations, leading to diagnostic problems. Three cases being reported illustrate the diagnostic difficulties, atypical hematological features, and rare complications in kala azar.

Case Reports

Case 1: A 12-year-old girl presented with fever, headache, altered sensorium, and cough with expectoration of five days duration. Examination revealed a malnourished and toxic child. Other signs were moderate pallor, generalized lymphadenopathy, bilateral fine crepitations on chest examination, no hepatosplenomegaly, and meningeal irriation with no neurological deficit. Investigations showed the level of hemoglobin 10 g/dl, total leucocyte count 11700/cu mm, with polymorphs 84%. The blood and sputum cultures were sterile, and the *X*-ray chest showed bilateral non homogeneous opacities suggestive of bronchopneumonia.

Management included ampicillin (100 mg/kg/day) and gentamicin (5 mg/kg/day). At the end of three weeks, the child continued to be febrile; clinical and radiologic signs of lower respiratory tract infection persisted. On empirical grounds a therapeutic trial with anti tubercular drugs was started. At the end of fifth week of

From the Departments of Pediatrics, Obstetrics, Medicine, and Pathology, Command Hospital, Bangalore and No. 5 A.F. Hospital C/o 99 APO.

Reprint requests: Dr. T.S. Raghu Raman, Department of Pediatrics, Command Hospital (A.F.), Agaram, Bangalore 560 007.

Received for publication: September 27, 1991; Accepted: January 16, 1992