Neuroblastoma presenting with paraplegia

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Introduction

Neuroblastoma presenting with paraplegia in a 7 month old Fijian is discussed. This is a rare presentation of this tumour. Neuroblastoma in infancy is one tumour that tends to undergo spontaneous regression. This malignancy is less common in developing countries but clinicians in these areas need to remain sensitised to its diagnostic features, prognostic factors and methods of evaluation.

Case report

A 7 month old Fijian boy was admitted with inability to move the limbs from a presumed fall 2 months prior to admission. He had been constipated and was dripping urine for about the same duration.

He was pale, afebrile and non-toxic. Tone and deep tendon reflexes were decreased in both lower limbs. Upper limb movements were normal. Sensation to pain was preserved up to the umbilicus. The child-health card showed that he had been fully immunised. The initial diagnosis was epidural abscess; other causes of spinal cord compression were to be excluded.

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The boy's haemoglobin on admission was 9.0 g/dl, white blood cell count 4000/mm³ and platelet 400 000/mm³. Electrolytes and urea were normal. Initial lumbar puncture yielded a clear cerebrospinal fluid with elevated protein of 11 000 mg/dl and sugar of 0.6 mmol/L. The random blood sugar was 4.8 mmol/L. Abdominal ultrasound showed dilatation of the ureters, a distended bladder and a mass in the left lumbar region which displaced the left kidney. Plain X-rays showed marked kyphosis at the thoracolumbar junction with poorly defined pedicles in the four upper lumbar vertebrae and scalloping of posterior aspects of T12 and L1-3. There was also a widening of the spinal canal (see Figure 1a and 1b).

Intravenous urogram and anterior posterior tomograms of the kidneys (see Figure 2a and 2b) showed displacement of the left kidney by a paraspinal mass; the upper tracts were however normal. A computerised tomography (CT) scan of the lower thoracic and lumbar region showed fairly well the paraspinal mass which was of soft tissue density with speckled calcification. In addition, there was a loss of retrocrural space from T12 to L4 vertebrae, evidence of intraspinal extension.

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A presumptive diagnosis of neuroblastoma was made. This was further strengthened by elevated urinary dopamine and noradrenaline. The child had decompressive lumbar laminectomy. The tumour mass was not excised because of its extent but scrapings taken showed mainly necrotic tissue containing nonspecific round and elongated cells.

Discussion

Paraplegia is one of the rare presentations of neuroblastoma[1]. The history of fall in this baby was a 'red herring'. Our diagnosis was made on the evidence of calcified abdominal mass with retroperitoneal and intraspinal extensions. In this age group neuroblastoma should be top of the list of diagnoses. It is the commonest childhood malignancy involving the spinal cord in infancy.

Figure 1(a) and 1(b)
AP and LATERAL views of the thoracolumbar spines showing small and poorly defined pedicles of L1-4. There is posterior scalloping of T12, L1-4 with widening of the spinal canal.

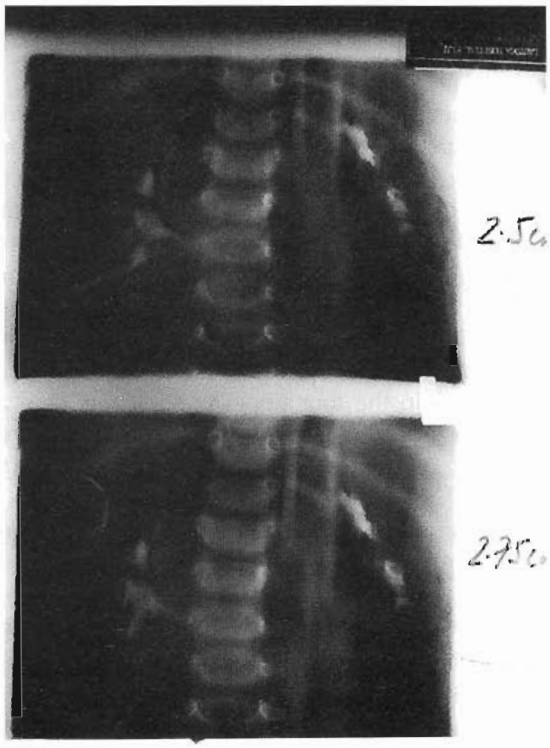
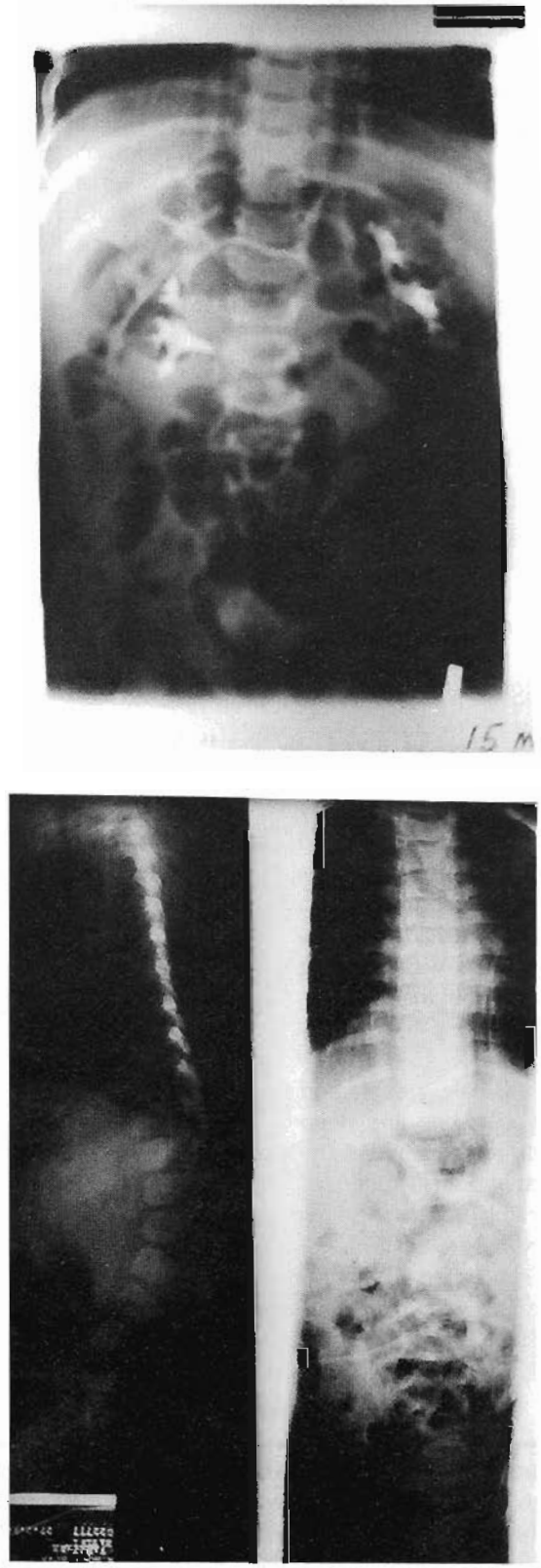


Figure 2(a) and 2(b)
IVU-AP Tomography of the kidneys (56 minutes) and abdominal film (15 minutes) illustrating the displacement of the left kidney by a paraspinal mass. There is good secretion of dye by both kidneys.



Neuroblastoma arises from cells of neural crest origin, mainly in the spinal sympathetic ganglia or suprarenal glands but also from other organs including the liver[2,3]. The wide extent of the tumour in this patient suggests multicentricity rather than metastasis. Ultrasonography is a useful investigation in delineating tumour masses including those arising from the suprarenal gland both in the prenatal and postnatal periods[4,5]. This has made intravenous urogram (IVP) virtually an unnecessary investigation. But IVP is useful in demonstrating displacement of the kidneys and collecting systems, distinguishing this condition from nephroblastoma, another childhood malignancy common in the first year of life. Ideally, lumbar puncture (LP) should not be done as part of investigation of compressive cord disorders unless a myelogram is contemplated. In our patient, the elevated CSF protein is in keeping with spinal compression. But myelography has been superseded by ultrasonography and CT scan. The latter represents the gold standard investigation although some workers believe that it is inferior to MRI in this condition[1].

Neuroblastoma is the commonest solid tumour in infancy particularly in the more industrialised countries. In developing countries it has been found mainly in areas of improved standard of living[6,7]. There are associations of this tumour

with congenital abnormalities, fetal hydantoin/alcohol syndromes and various drug intoxications[2, 6, 8, 9]. These conditions are more common in more affluent societies.

This tumour tends to regress spontaneously especially when diagnosed in infancy but if there is involvement of the spinal cord the prognosis is usually worse[1]. Some authors suggest that aggressive and immature tumours produce dopamine whereas more differentiated tumours produce noradrenaline[6]. But generally over 90% of neuroblastoma secrete cathecolamines and their metabolites, except tumours involving the spine.[10] Our patient's elevated catecholamines may therefore be a result of the extra-spinal involvement.

" Unfortunately, neuroblastoma of the spinal cord tends to have a poor prognosis in spite of invasive chemotherapy and other treatment modalities. "

In summary, we have described a 7-month old Fijian boy with neuroblastoma who presented with paraplegia. Neuroblastoma is the commonest solid tumour of infancy and should therefore be considered in any infant presenting with paraplegia. Unfortunately, neuroblastoma of the spinal cord tends to have a poor prognosis in spite of invasive chemotherapy and other treatment modalities[1].

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