Intrarenal Neuroblastoma Presenting as Orbital and Multiple Skeletal Metastases

Lalit Verma
S. Sandramouli
S.P. Garg
S. Vashisht

Primary renal neuroblastoma, a rare clinical entity, is known to mimic Wilms' tumor both clinically and radiologically(1). We herein report a case of intrarenal neuroblastoma that presented with rapidly enlarging unilateral proptosis.

Case Report

A 12-year-old boy presented to us with a rapidly progressive protrusion of the right eye of 20 days' duration, associated with marked, progressive deterioration of vision and two bony swellings, one each on the forehead and the right temple.

On examination, the boy was cachectic with marked pallor but with no significant lymphadenopathy. Both on the forehead and the right temple region, a non-tender, hard swelling with irregular surface, measuring 3 × 3 cm was present. Both the swellings were fixed to the underlying bones and had areas of softening. Palpation of the abdomen revealed a large, non-tender, irregular lump in the region of the right renal angle, which was of firm consistency and was bimanually ballotable. The rest of the systemic examination was remarkable.

Ocular examination of the left eye was unremarkable with unaided Snellen's visual acuity of 20/20. In the right eye, there was no light perception. There was marked and irreducible proptosis of the right eye with downward and medial displacement of the globe (*Fig. 1*) and a generalized severe restriction of the ocular movements. On valsalva manoeuvre and neck vein compression, there was no increase in the prominence of the globe. The conjunctiva was xerotic with marked chemosis. The dry, desiccated and opaque cornea prevented visualization of the structures posterior to it. Investigations of hemoglobin, total and differential leucocyte counts, platelet

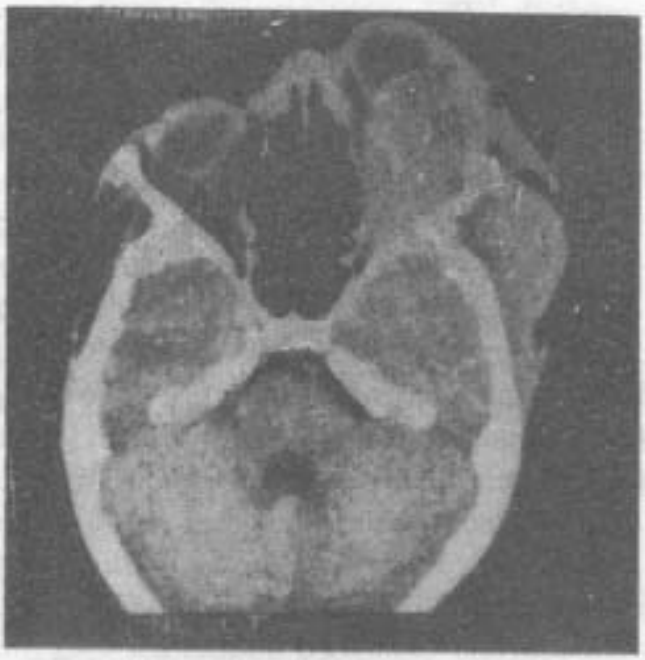

Fig. 1. CT Scan of the orbit and the skull showing proptosis of the left eye with medial displacement. Large soft tissue masses are seen retrocularly in the left orbit and in the left frontoparietal region.

From the Dr. Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, Ansari Nagar, New Delhi 110 029.

Reprint requests: Dr. Lalit Verma,4654, Shora Kothi, Pahar Ganj,, New Delhi 110 055.

Received for publication: April 6, 1992;
Accepted : December 1, 1992

count, peripheral smear, erythrocyte sedimentation rate, urine examination, blood sugar and liver function tests yielded normal results. Urinary catecholamine levels of vanil mandelic acid (VMA) and homovanillic acid (HVA) were within normal limits.

X-rays of the orbit revealed enlargement of the right orbit with marked soft tissue haziness. The roof of the orbit was normal but the greater wing of the sphenoid could not be defined. *X*-ray of the skull in both anteroposterior and the lateral views showed an osteolytic lesion in the posterior frontal region with soft tissue swelling over the scalp adjacent to it. *X*-ray of the right humerus showed an osteolytic area in the upper end of the humerus. *X*-ray chest and rest of the skeletal survey were normal. CT scan of the head and orbit demonstrated a large soft tissue mass occupying the retrobulbar space (*Fig. 1*) with displacement of the globe anteriorly, inferiorly and medially. The mass was of increased attenuation with areas of low attenuation in it, suggestive of necrosis. Right optic nerve, medial and lateral recti muscles could not be defined due to the presence of the large mass. There were areas of destruction of the lateral wall of the orbit and the anterior part of the squamous temporal bone with blistering. The mass was seen extending into the right frontotemporal lobe with compression of the right ventricle and shift of the midline structures contralaterally. Another soft tissue mass was seen in the frontoparietal convexity in the midline with irregularity of the adjacent bone (*Fig. 1*). On contrast enhanced abdominal CT, the spleen, gall bladder and the left kidney were normal. Pancreas was normal but displaced ventrally. The left lobe and the upper part of the right lobe of the liver were normal. The inferior part of the right lobe of the liver was compressed by a mass lesion with obliteration of the cleavage plane between the mass and the liver. The soft tissue mass was seen extending from the inferior surface of the liver to the superior border of the fifth lumbar vertebra. The lesion was sharply defined with a thin enhancing capsule with both solid and cystic components in it. No retroperitoneal adenopathy was seen. *X*-ray abdomen taken following the contrast enhanced CT (*Fig. 2*) revealed non visualized right kidney and normal left kidney.

On ultrasonography of the abdomen, the right kidney could not be defined. A large soft tissue mass was seen in the region of the right kidney extending up to the inferior surface of the liver and could not be separately defined. The right suprarenal gland was not seen. Rest of the abdominal organs were normal.

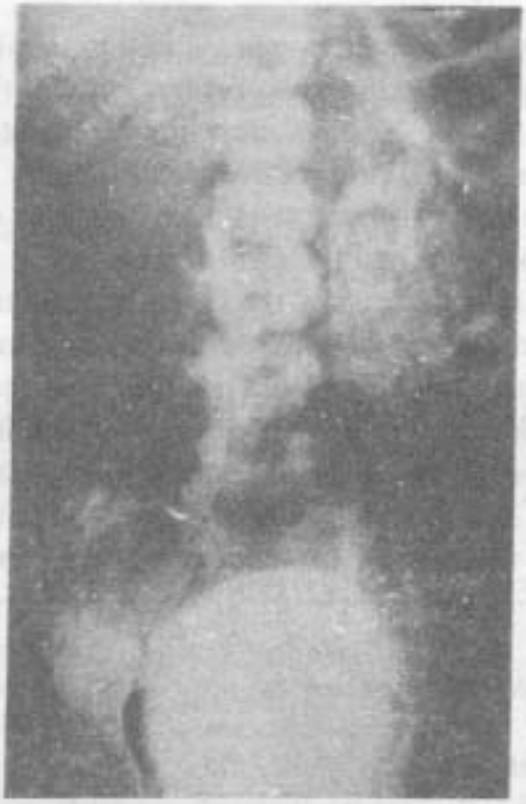

Fig. 2 X-ray of the abdomen taken following the contrast enhanced CT, showing the non-visualized right kidney and the normal left kidney.

Microscopic examination of the fine needle aspirate from the orbital mass showed highly cellular small round cell tumor with rosette formation. Biopsy from the cutaneous deposits were highly suggestive of neuroblastoma with multiple round cells having scanty cytoplasm, rosette and pseudorosette formation and multiple areas of hemorrhages and necroses (*Fig. 3*). The boy underwent palliative chemotherapy but died after three months. Postmortem examination was, however, refused by the parents.

Fig. 3. Photomicrograph showing the rosette formtion (arrow), suggestive of neuroblastoma (Hematoxylin and Eosin, 35X).

Discussion

Bone metastases are uncommon in children with renal tumors. This case with a renal lump and orbital metastasis was a diagnostic dilemma till the biopsy from the metastatic obital nodules resolved the problem. Wilms' tumor with orbital metastases has been reported in the literature (2,3). Although clinically and radiologically the abdominal mass in our patient suggested Wilms' tumor as the diagnosis, the essential features which disfavoured the diagnosis of Wilms' tumor as the primary pathology were the paucity (3.5%) of bony metastases associated with Wilms'(4) and the absence of blastema with variable, epithelial differentiation including tubule formation on histopathology. Neuroblastoma is well known to masquerade as an orbital malignancy with multiple bone deposits before the primary focus in the suprarenal gland manifests itself(5). It has also been reported to occur as a primary orbital malignancy(6,7). In our patient, the clinical features along with the non functioning kidney on intravenous pyelography done following the contrast injection for the CT scan were highly suggestive of an intrarenal origin of the mass. Though there are reports of intrarenal neuroblastoma(1,8,9), we document perhaps for the first time in the literature, a case of clinically suspected intrarenal neuroblastoma who presented to an ophthalmologist with proptosis, orbital and other skeletal deposits.

Bone metastasising renal tumor of childhood (BMRTC), a recently established entity, mimicked our case closely with its well-known propensity(10) and multiple bone deposits. However, the absence of prominent capillary network, a hallmark of BMRTC, was characteristically absent in our biopsy study(11).

This case differed from typical neuroblastoma in being intrarenal and presenting to an ophthalmogist with orbital and multiple skeletal metastases.

REFERENCES

1. Shende A, Wind ES, Lanzkowsky P. Intrarenal neuroblastoma mimicking Wilms' tumor. NY State J Med 1979 79: 93.
2. Fratkin JD, Purcell JJ, Krachmer JH, Taylor JC Wilms' tumor metastatic to the orbit. JAMA 1977, 238: 1841-1842.

3. Apple DJ. Wilms' tumor metastatic to the orbit. Arch Ophthalmol 1968, 80: 480-483.
4. Bond JV, Martin EC. Bone metastasis in Wilms' tumour. Clin Radiol 1975, 26: 103-106.
5. Mortada A. Clinical characteristics of early orbital metastatic neuroblastoma. Am J Ophthalmol 1967, 63: 1787-1793.
6. Levy WJ. Neuroblastoma. Br J Ophthalmol 1957, 41: 48.
7. Bullock JD. Primary Orbital Neuroblastoma. Arch Ophthalmol 1989, 107: 1031-1033.
8. Third National Cancer Survey:Incidence data publication (NIH) 75-787, US Department of Health, Education, Education and Welfare, Washington, DC. March, 1975, p 420.
9. Gross RE, Farber S, Martin LW. Neuroblastoma sympatheticum: A study and report of 217 cases. Pediatrics 1959 23: 1179-1182.
10. Marsden HB, Lennox EL, Lawler W. Kinnier-Wilson LM. Bone metastases in childhood renal tumours. Br J Cancer 1980:41: 875-879.
11. Marsden HB, Lawler W, Kumar PM. Bone metastasising renal tumor of childhood. Cancer 1978, 42:1922-1928.

Cystic Tuberculosis of Bones

A.S. Kher
B.A. Bharucha
J.R. Kamat
P. Kurkure

Multiple osteolytic lesions involving the axial and extra axial skeleton are a rare form of tuberculosis (TB) in children(1). It is seen in patients with rapidly acquired immunity and in those with good resistance(2). There are scanty reports of such lesions in pediatric patients and therefore we wish to present this unusual form of tuberculous osteomyelitis which showed a favorable response to treatment.

Case Report

A 5-year-old boy presented with fever, slowly progressive and painless soft swellings over the forehead, left cheek, right mid-axillary region, both legs and right thumb of 6 months duration. Systemic examination revealed a soft hepatomegaly of 2 cm. Investigations included a positive Mantoux test (20 × 20 mm) and an ESR of 55 mm at 1 hour. Hemogram, liver function tests and abdominal sonography were normal. Skeletal survey revealed fifteen lesions. Well defined rounded osteolytic lesions with sclerotic margin were seen in the right frontal and parietal bones, body of left mandible, right 8th and 9th ribs and without sclerosis on both tibiae and fibulae (*Fig. 1*). Lateral margins of the D_2 and D_3 vertebral bodies and their left pedicles were eroded. Osteolytic lesions were also seen in the left ischial bones, distal half of the right humeral diaphysis and the left radial shaft. Characteristic fusiform expansion described as spina ventosa was noted in the left first metacarpal.

A ^{99m}Tc MDP bone scan showed uniformly increased tracer concentrations in

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

Reprint requests: Dr. J.R. Kamat, Department of Pediatrics, K.E.M. Hospital, and Seth GSM College, Parel, Bombay 400 012.

Received for publication: June 16, 1992; Accepted: December 4, 1992