

ADRENAL NEUROBLASTOMA WITH CALVARIAL INVOLVEMENT SPARING ORBIT

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ABSTRACT

Neuroblastoma is third commonest childhood tumor. It is the most common extracranial pediatric neoplasm. It is a tumour of neuroblastic origin. Intra abdominal occurrence is common with adrenals (35%) being the most common site of origin. Other sites may be retroperitoneum: 30-35%, posterior mediastinum: 20%, neck: 1-5%, pelvis: 2-3%. They have been associated with a number of disorders, such as Hirschsprung disease, fetal alcohol syndrome, DiGeorge syndrome, Von Recklinghausen disease, and Beckwith-Wiedemann syndrome. Approximately 70-80% of patients older than 18 months present with metastatic disease in liver, lymph nodes, bones, and bone marrow. Skull vault metastases are generally more common in adults and infrequent in children. Calvarial metastases are characteristically seen with simultaneous involvement of the orbits and isolated metastasis rarely occurs.^{1,2,3} We report a case of five year old male who was diagnosed as a case of neuroblastoma which initially presented with extensive calvarial metastasis with no involvement of the orbits. CT showed cortical irregularity and thinning of the skull bones and hair on end periosteal reaction which was involving the fronto parieto temporal bones bilaterally. The coronal and sagittal sutures were also widened. Intracranial soft tissue component was noted along the fronto-parieto-temporal bones bilaterally. Concave convex extracranial soft tissue component was also noted in both fronto parietal regions. The differentials included lymphoproliferative disorders. Patient ultrasound abdomen showed a well-defined hyperechoic mass in right adrenal gland. It showed no internal calcifications. Abdominal lymphadenopathy was also noted. Patient was further referred to oncologist.

Key words: Neuroblastoma, calvarial metastasis, chemotherapy.

Introduction

Neuroblastoma is third commonest childhood tumor. It is the most common extracranial pediatric neoplasm. It is a tumour of neuroblastic origin. Intra abdominal occurrence is common with adrenals (35%) being the most common site of origin. Other sites may be retroperitoneum: 30-35%, posterior mediastinum: 20%, neck: 1-5%, pelvis: 2-3%. They have been associated with a number of disorders, such as Hirschsprung disease, fetal alcohol syndrome, DiGeorge syndrome, Von Recklinghausen disease,

and Beckwith-Wiedemann syndrome. Approximately 70-80% of patients older than 18 months present with metastatic disease in liver, lymph nodes, bones, and bone marrow. Skull vault metastases are generally more common in adults and infrequent in children. Calvarial metastases are characteristically seen with simultaneous involvement of the orbits and isolated metastasis rarely occurs.^{1,2,3} We report a case of neuroblastoma which initially presented with extensive calvarial metastasis with no involvement of the orbits.

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Case Report

A 5 years old male child presented to us with anaemia, weakness, lethargy, reduced appetite and irritability. Upon general physical examination child had normal built, his vitals were within normal limits, and however his skull was enlarged and was lumpy bumpy on palpation. There was no tenderness or signs of inflammation over the swelling of the scalp. His labs show reduced hemoglobin and MCHC and MCV, x-ray chest was normal. His previous documents showed workup for thalassemia major which was negative. He was advised CT scan brain which showed cortical irregularity and thinning of the skull bones and hair on end periosteal reaction which was involving the fronto-parieto-temporal bones bilaterally. The coronal and sagittal sutures were also widened. Intracranial soft tissue component was noted along the fronto-parieto-temporal bones bilaterally. Concavo convex extracranial soft tissue component was also noted in both fronto parietal regions. Patient was given the differentials of metastasis of neuroblastoma/ lymphoproliferative disorder.

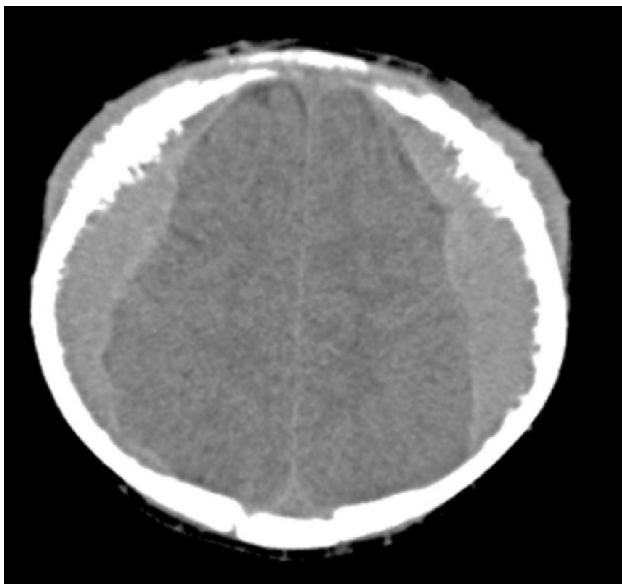


Figure 1: Extra axial soft tissue component along the frontoparietal bones along with concavo convex soft tissue extra cranial component

Patient further underwent ultrasound abdomen which showed a well-defined hyperechoic mass in right adrenal gland. No internal calcification/ cystic areas were

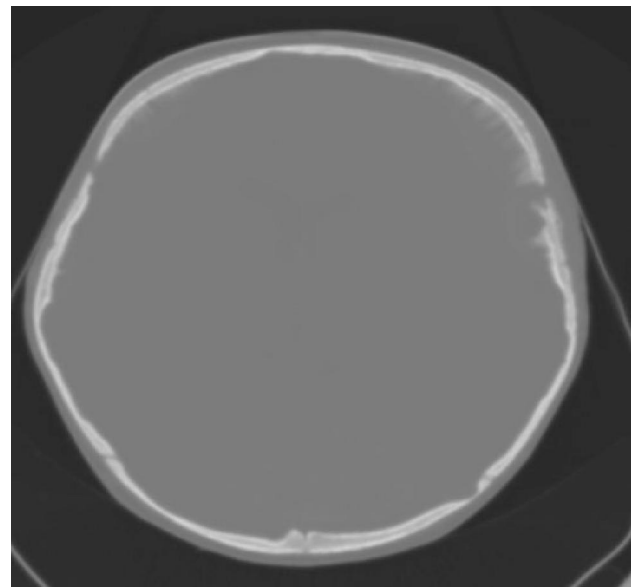


Figure 2: Axial CT scan bone window showing hair on end periosteal reaction and diastasis of coronal suture.

seen. Abdominal lymphadenopathy was also noted. CECT abdomen showed a mildly enhancing right adrenal mass with lymphadenopathy at the para aortic and retrocaval region which was pushing the right renal vessels anteriorly. CT abdomen further revealed involvement of both the iliac bones showing hair on end periosteal reaction.

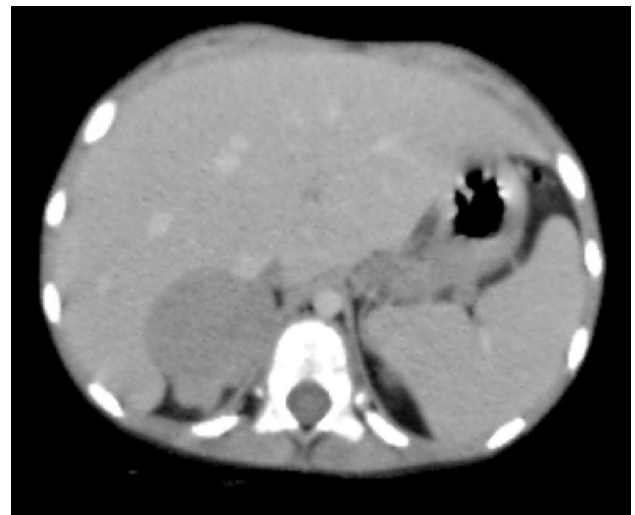


Figure 3: CECT Abdomen showing mildly enhancing right adrenal mass

I-¹²³ MIBG scan was in plan but was not performed due to unavailability of the test. Patient was referred to oncologist.

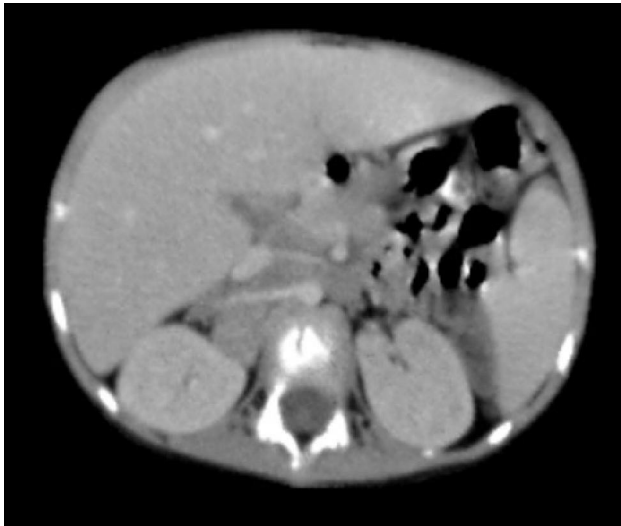


Figure 4: Para aortic lymphadenopathy encasing and displacing the right renal artery and left renal vein

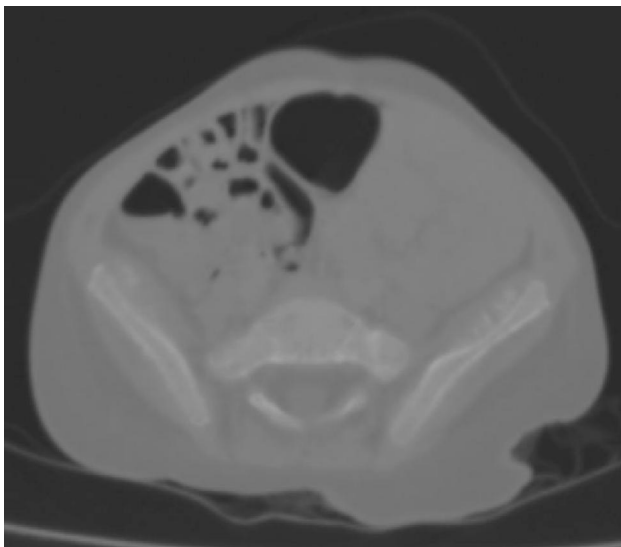


Figure 5: Axial CT scan bone window showing hair on end periosteal reaction involving both iliac bones

Discussion

The incidence of neuroblastomas is 9.5 cases per million children. The incidence is highest in high income countries and lower in low income countries as in Asia and Africa. Incidence is also high in white children, males and under the age of 10 years. The differentials of abdominal mass includes Wilms tumour and Rhabdomyosarcoma whereas that of calvarial metastasis include osteomyelitis, eosinophilic granuloma and lymphoproliferative disorders.⁴

Metastatic neuroblastoma has a predilection to metastasize to the dura. The typical pattern of metastasis was first observed by Hutchinson in 1907 to have strong preference for skull bones and bony orbit. Dural metastases tend to favor the external surface of the dura, spreading diffusely over both the convexities and base of the skull. The dura acts as a barrier to direct invasion, so involvement of the brain parenchyma is rarely seen. Dural metastases are almost always associated with osseous metastases and can be hemorrhagic. Tumour often extends to the scalp with palpable nodules as in our case. Dural metastases may respond favorably to treatment.^{5,6} Keating and Cromwell⁵ reported the remote effects of neuroblastoma in a five year old, who presented with lumps of scalp, decreased appetite, anemia, widening of sutures with hair on end periosteal reaction very much like our patient, but they could not trace the primary tumour.

Prognostic factors for neuroblastoma are age at onset of disease, the site of origin of lesion, presence or absence of osseous metastasis and the spontaneous disappearance or maturation of tumour. A case of spontaneous cure was reported by Carvalho L,⁷ in which the mother refused treatment for her child over a period of almost twenty years. Although there were bony metastasis but patient survived without therapy whatsoever.

Neuroblastomas are not hereditary, do not result from injury and are not infectious. Only 1-2% have family history.

Egelhoff and Zelles⁶ are the only ones to report an isolated calvarial deposit with a large soft tissue component and sun ray spiculation as a leading presentation. Our patient presented with extensive calvarial deposits which had a spiculated bone reaction.

Neuroblastomas is a chemosensitive tumor and in combination with surgery has good prognosis. According to Kim et al⁸ neoadjuvant chemotherapy is very effective. They used it for calvarial metastasis for five months followed by surgery. No recurrence was observed.

Conclusion

Neuroblastoma can be confused with other neoplastic or non neoplastic disease of childhood. The diagnosis is challenging in 10% of cases. Awareness of the

confusing presentation of this disease is important for early diagnosis. It presents with vague signs and symptoms as anorexia, weight loss, fatigue, bone pain, chronic diarrhea and irritability. Two thirds present with abdominal mass which may be asymptomatic. Anemia and cytopenias suggest hematological disorders. A multidisciplinary approach helps in early diagnosis. Ultrasound abdomen, CECT abdomen, CT brain, MRI orbit, MIBG, T⁹⁹ bone scan and skeletal survey are all important diagnostic modalities in their own way.⁸

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