

CLINICAL CASE

Neuroblastoma: Incidental Craniocerebral Metastasis A Case Report

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ABSTRACT

Neuroblastoma has been attributed the term “the great imitator” due to its varied spectrum of presentations. Although neuroblastoma is a common childhood malignancy, which frequently metastasizes, involvement of the Central nervous system is rarely reported in the literature. It commonly metastasizes to the base of the skull and orbits late in the disease. Early detection and aggressive treatment of this complication may allow some patients to live longer than they would have otherwise.

KEY WORDS: Neuroblastoma; Metastasis; Skull.

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INTRODUCTION

Neuroblastoma is the third most common malignancy in children accounting for 10-15% cases of childhood tumours. The most common site of abdominal tumour is adrenal gland or along sympathetic chain in retroperitoneum. Although 90% of cases are manifested clinically in the first 8 years of age, approximately 50% of cases fall under two years of age. Neuroblastoma has diverse metastatic manifestations, often masquerading as primary neurologic disorder. Despite early metastatic potential, involvement of CNS is rarely reported in the literature. Metastases are present in upto 70% of cases at the time of diagnosis [1]. Secondary craniocerebral metastases often manifests as osseous lesions involving base of skull, orbit and calvaria [2]. Metastasis to skull is noted in approximately 25% of cases with neuroblastoma being the most common malignant metastasis to skull in children [3]. The imaging appearance varies from lytic defects, “hair-on-end” periosteal reaction to the separation of sutures. The sutural separation seen in neuroblastoma is non-uniform with indistinct margins, contrary to uniform appearance of raised intracranial pressure [3,4]. The presence of primary adrenal neuroblastoma with extensive

skeletal metastases particularly to skull, causing proptosis and bone pain has been referred to as ‘Hutchinson’s syndrome’ [5].

CASE REPORT

A nine month old male child presented to outpatient clinic with complaints of proptosis [Figure 1]. Clinical examination and laboratory investigations were normal. Further work-up with X-ray skull [Figure 2], ultrasonography and Computed tomography was done. Skull radiographs revealed separation of sutures with 'hair-on-end' periosteal reaction. USG [Figure 3] and CECT [Figure 4] demonstrated a large heterogenous retroperitoneal lesion arising from left adrenal gland, crossing the midline and encasing Aorta and Inferior vena cava. Multiple calcific foci and areas of enhancement on contrast administration are also noted. On CECT brain [Figure 5] multiple enhancing epidural soft tissue lesions were seen, representing metastasis from adrenal mass lesion. Bone window reconstructions depicted sutural separation with hair on end periosteal reaction with lytic areas [Figure 6,7]. Due to presence of distant metastasis, patient was put on intensive chemotherapy and kept on regular follow-up.

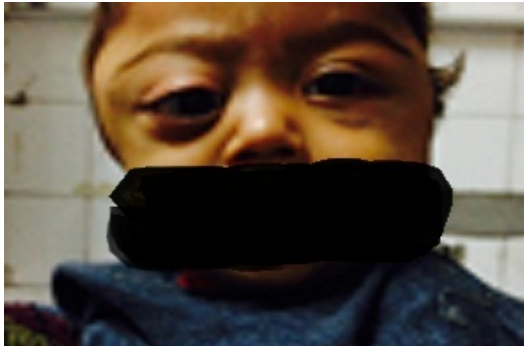


Figure 1: Pictograph of a nine month old child with right sided proptosis and periorbital ecchymosis.

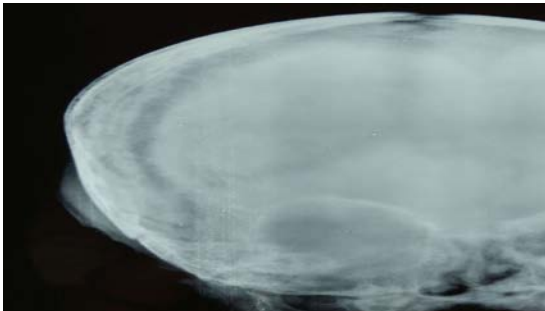


Figure 2: AP radiograph skull showing sutural separation with scalp lesion.

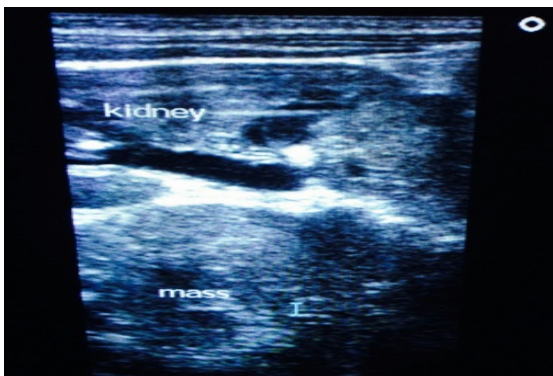


Figure 3: Sonography abdomen shows a solid heterogenous mass lesion involving right adrenal gland



Figure 4: Axial Contrast enhanced abdominal CT scan showing a heterogenous lesion with foci of calcification

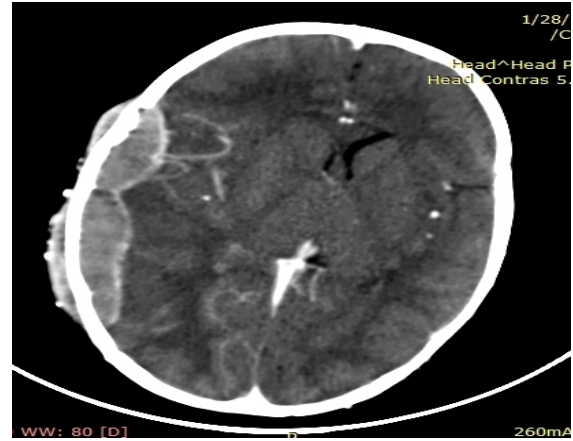


Figure 5: Axial Contrast enhanced CT Brain showing enhancing epidural and scalp lesion, most likely metastasis

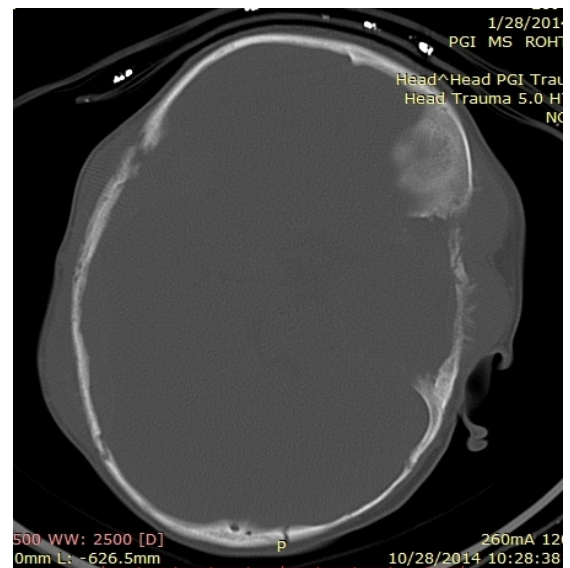


Figure 6: Axial Bone CT showing split sutures with periosteal reaction involving bilateral temporal bones.

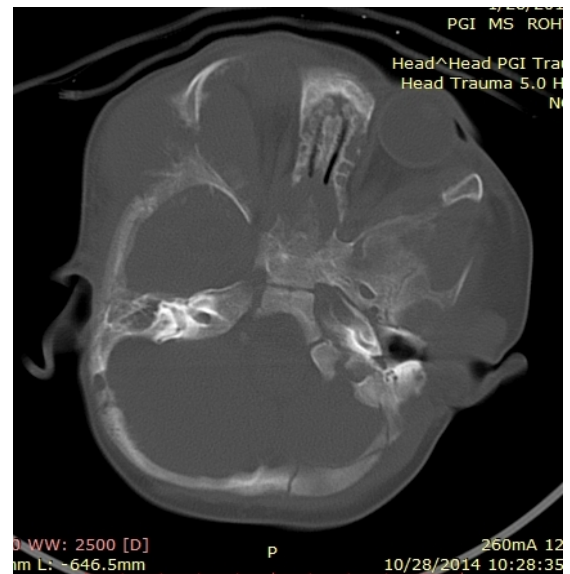


Figure 7: Axial Bone window reconstructed CT showing osseous metastasis at the junction of frontal bone and greater wing of sphenoid with hair on end periosteal reaction.

DISCUSSION

Neuroblastoma arises from the primordial neural crest cells that form the sympathetic nervous system. The exact etiology of this disease is not yet known. It usually occurs sporadically, only 1-2% of the cases being familial.[6,7] Approximately 60-70% of the cases are metastatic at presentation.[8] The median age at diagnosis is 22 months.[9] Symptoms caused by skeletal metastases lead to bony pain and orbital wall metastases presenting as Panda sign or Raccoon eyes (due to orbital ecchymoses causing darkening of periorbital tissues) [9]. On CT, the tumour typically is heterogeneous with calcifications seen in 80-90% of cases [5]. CNS metastasis in cases of primary adrenal neuroblastoma clinically occult in most of the cases and symptoms of visual disturbances and headaches often go unnoticed in very young children. Early diagnosis and treatment is imperative in such cases to improve longevity and prevent further dissemination of disease. The reported frequency of CNS metastasis (parenchymal or leptomeningeal) in disseminated neuroblastoma is 2-16%. It has an inherent predilection to metastasize to dura, limited to its external surface spreading over the

AUTHORS' CONTRIBUTIONS

The participation of each author corresponds to the criteria of authorship and contributorship emphasized in the [Recommendations for the Conduct, Reporting, Editing, and Publication of Scholarly work in Medical Journals of the International Committee of Medical Journal Editors](#). Indeed, all the authors have actively participated in the redaction, the revision of the manuscript and provided approval for this final revised version.

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CONCLUSION

Imaging plays a crucial role in diagnosis, staging, pre-operative evaluation, management and surveillance of recurrence in cases of neuroblastoma. Any subtle sign of neurologic impairment in paediatric age warrants the active search for neuroblastoma using various imaging techniques.

ACKNOWLEDGEMENT

Declared none.

PATIENT CONSENT

Written informed consent was obtained from the patient for publication of this case report.

COMPETING INTERESTS

The authors declare no competing interests.