Hutchison's syndrome: suspecting metastatic neuroblastoma

Krishna Sindhu Nadella, Devjyoti Tripathy

Department of Ophthalmic
Plastics, Orbit and Ocular
Oncology, LV Prasad Eye
Institute Bhubaneswar Campus,
Bhubaneswar, Odisha, India

Correspondence to
Dr Devjyoti Tripathy;
drdtripathy@gmail.com

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DESCRIPTION

A 2-year-old male child presented with periocular ecchymosis (more noticeable on the right side), left eye proptosis with severe chemosis and exposure keratitis, left eyelid and periorbital swelling extending on to the temporal fossa (figure 1A), a distended abdomen and bipedal oedema of about 3 weeks' duration. He had facial swelling involving the maxillary bones and the mandible. The child was severely cachectic, febrile, irritable and had an unstable, limping gait with major difficulty in walking. CT imaging of the orbits showed multiple lytic bony lesions involving several of the skull bones—the frontal bone (figures 1B and 2A), the left maxillary bone (figure 1B), and, the mandible (figure 1C). Ultrasonography of the abdomen revealed a large heteroechoic mass centred on the right suprarenal region (figure 2B) that encased the inferior vena cava (figure 2D) and the abdominal aorta (not shown). There was a coexisting severe anaemia (a haemoglobin level of 2.8 gm/dL). Clinico-radiologically, this was highly suggestive of a metastatic neuroblastoma. The patient succumbed

Figure 2 (A) An upper axial orbital CT scan shows a heterogeneous soft-tissue mass on the left side with intraorbital and extraorbital temporal fossa components (arrows) and a middle cranial fossa component (solid arrowhead). (A) The adjacent bone shows erosive changes. (B) Ultrasonography of the abdomen shows a heteroechoic suprarenal soft-tissue mass on the right side radiologically suggestive of a neuroblastoma. The mass appears to be displacing the (C) liver anteriorly and encasing the (D) inferior vena cava (IVC).

Figure 1 A 2-year-old child presenting with right periocular ecchymosis and left orbital mass with proptosis, severe chemosis, inferior globe displacement and severe exposure keratopathy (A) Eyelid oedema with temporal periorbital swelling extending onto the infratemporal fossa is also noted. (B) 3-D reconstructed CT of the skull shows lytic bone lesions involving the frontal bone and the left maxillary bone (hollow arrowheads). (C) The skull radiograph shows lytic bone lesions with associated soft-tissue masses in the left maxilla (solid arrowhead) and the mandible (arrows).

to the disease just short of 3 weeks from the time of diagnosis.

The presentation of skull bone metastases in primary adrenal neuroblastoma was first reported in a series of 10 children in 1907 by Robert Hutchison¹ and is thus known by the name of 'Hutchison's Syndrome'. Associated severe anaemia was also a prominent feature in all the patients in this series. A limping gait and irritability have also been described as accompanying features of this syndrome.² The common ocular signs of metastatic neuroblastoma are periocular ecchymosis, proptosis, unilateral Horner's syndrome and opsoclonus.³ Nearly 48% of patients with neuroblastoma have metastatic disease at presentation.⁴ While involvement of the skull and orbital bones is

Learning points

- ▶ In the presence of periocular ecchymosis, proptosis and bony facial swelling in children less than 10 years old, metastatic neuroblastoma should be strongly suspected.
- ▶ Abdominal ultrasonography can be an extremely cost-effective method for quick screening and a preliminary diagnosis of the primary tumour since the adrenal glands are the most common site of origin.
- ▶ Multiple metastases in neuroblastoma usually portend a very poor prognostic outcome.

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reported in up to 25% of cases with metastatic neuroblastoma, involvement of the mandible is uncommon, and that of the maxilla is extremely rare.³ Our patient presented with involvement of both the mandible as well as the maxilla.

Twitter Devjyoti Tripathy @devtripathyNU

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