

1 Background Rare **neural crest** tumor of infancy; only one prior Pakistani report (8-month-old girl, 2013)

~500

cases reported globally; ~70% in the maxilla

91

cranial cases in the last decade (Ren et al.)

27%

recurrence rate (up to); locally aggressive

2 Case snapshot

	CASE 1 6-month-old girl	CASE 2 3-month-old boy
Clinical	Right orbitotemporal swelling, 5 × 4 cm , 2 months	Right temporal swelling, 3 × 2 cm , 3 weeks; 5.8 kg
CT	48 × 43 × 14 mm iso- to hypodense; hyperostosis of lateral orbital wall and temporal bone; spurs at lesser sphenoid wing	29.4 × 20.1 × 30 mm extra-axial; hyperostosis; coronal suture widened; sylvian fissure obliterated; temporal lobe compressed
MRI	Not done (purely bony on CT)	3.4 × 3 × 4.2 cm ; T1 hypo, FLAIR iso, T2 hyper; avid enhancement
Initial dx	Osteoma	Meningioma vs fibrous dysplasia

Both: painless, firm, rapidly growing, non-pulsatile swelling; no seizures, no fever; normal milestones

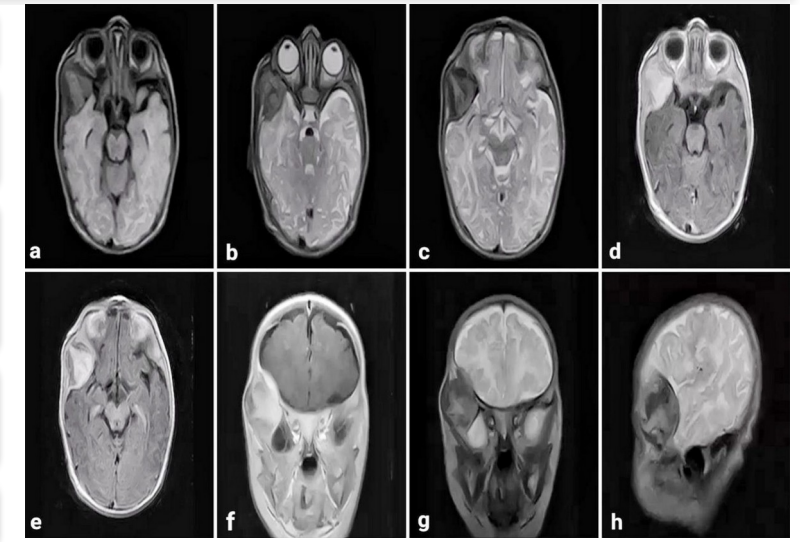


Fig. 4 Case 2 MRI: T1 (a), T2 (b, c), T1 + contrast (d-f), T2 coronal and sagittal (g, h).

3 Surgical management

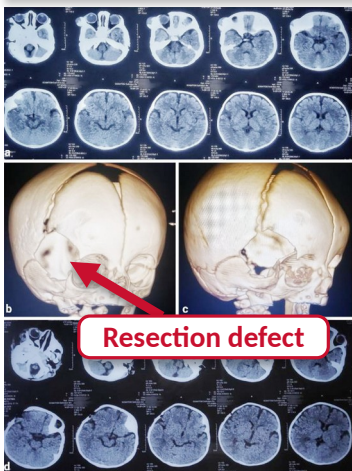


Fig. 1 (a) Pre-op CT; (b, c) post-op 3D CT; (d) post-op CT, complete resection.

- 1 Incision:** right curvilinear frontotemporal incision
- 2 En bloc excision:** single burr hole; hyperostotic bone cut with drill; dura coagulated at the lesion attachment
- 3 Specimen:** C1 50 × 45 × 15 mm; C2 ~4.5 × 4 × 4.5 cm of bone
- 4 Closure:** no cranioplasty (growing infant skull)

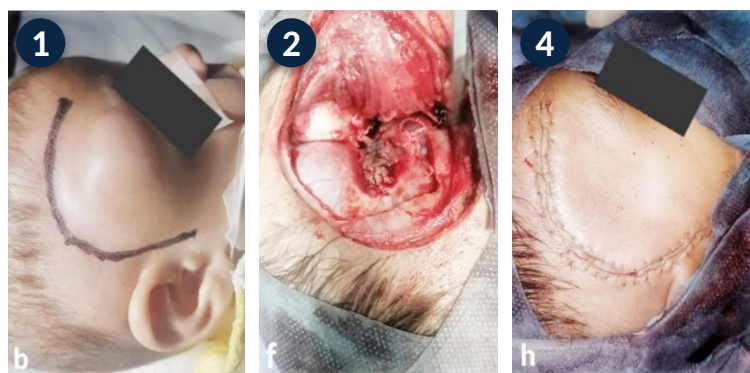


Fig. 2 Intraoperative, Case 1: incision marking (b); complete en bloc bone excision (f); skin closure (h). Faces masked.

4 Histopathology

Biphasic: small round blue cells + large melanin-pigmented epithelioid nests.

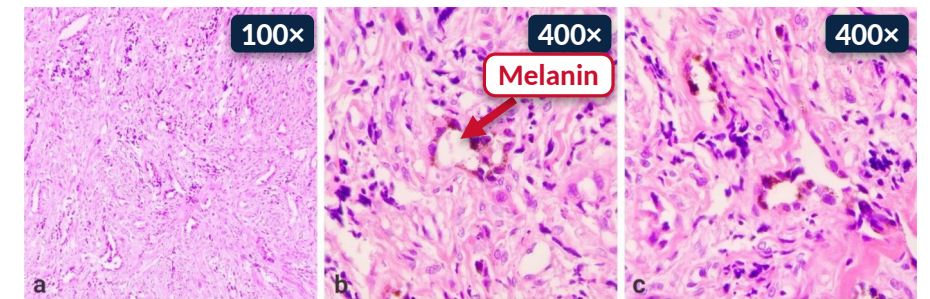
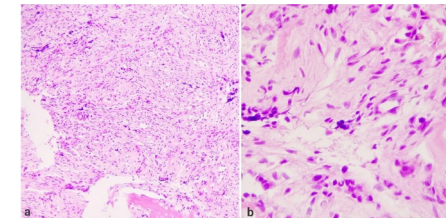


Fig. 3 (a) 100x; (b, c) 400x.



IHC and VMA not done: resource limits; no preoperative suspicion.

Fig. 5 Round blue cells and melanin-pigmented epithelioid cells.

5 KEY RESULTS

100%
gross total resection

0

intraoperative complications

0

postoperative complications

0

recurrence at 6-month follow-up

2/2

normal developmental milestones

88.7% of skull MNTI: painless, rapidly growing swelling

~70%

diagnosed within 6 months of life

2/18

VMA positive when tested preoperatively

3:1

male:female in literature; here 1 boy, 1 girl

6 Clinical pearls

Suspect MNTI in any infant under 1 year with a hyperostotic skull lesion.

Do not label it osteoma or fibrous dysplasia without considering MNTI.

Clear-margin excision is definitive; avoid immediate cranioplasty.

Long-term surveillance is essential; adjuvant therapy is controversial.

7 Conclusion

MNTI is a rare, locally aggressive pediatric tumor. Early, complete resection gave excellent short-term outcomes in both cases; structured long-term follow-up is needed.

8 References

- Ren Q et al. MNTI of skull and brain: systematic review. World Neurosurg. 2019;130:170-178.
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- Krawczyk MA et al. Focus on MNTI. EJC Paediatr Oncol. 2024;3:100141.
- Khan MB et al. MNTI: a rare brain tumour of childhood. JCPSP. 2013;23:367-369.

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