

Role of Preoperative Embolization for Hypervascular Cherubism

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Cherubism is an autosomal dominant condition, first described by Jones in three siblings who presented with a fullness of the cheeks and enlargement of the jaws.¹ The characteristic appearance closely resembled the Renaissance cherubs inspiring Jones to name it as cherubism.^{2,3} The condition is a fibro-osseous dysplasia characterized by painless symmetrical enlargement of the jaws. The mean age at the time of presentation is 7 years with a range of 12 months to 14 years.⁴ The condition has a 100% penetrance in males and 50 to 70% in females.⁵ In this article, we describe the role of angiography and embolization for a rare hypervascular variant of cherubism prior to surgical therapy.

Case Report

A 9-year-old girl presented with progressive, painless enlargement of both the cheeks. The symptoms were first observed at the age of 4 years. There was no associated

difficulty in breathing or swallowing. However, the parents observed snoring in the last year of presentation. There was no history of similar symptoms in any of the family members. On examination, the swelling was noted to involve the bilateral maxilla and mandible. There was proptosis. Intraoral examination showed displaced and missing teeth. Laboratory evaluation was unremarkable with normal coagulation profile. A computed tomographic (CT) angiography of the head and neck demonstrated a diffuse expansile bone lesion involving the bilateral maxilla and mandible with a soap bubble pattern and cortical thinning. The lesion involved the mandibular condyles and pushed the orbital floor superiorly with associated narrowing of the superior orbital fissure and pterygopalatine fossa (►Fig. 1). On the angiographic phase, the soft tissue component within the lesion showed avid enhancement with multiple arterial feeders from bilateral facial and internal maxillary arteries. No arterial feeders were seen arising from the internal carotid arteries (►Fig. 2).

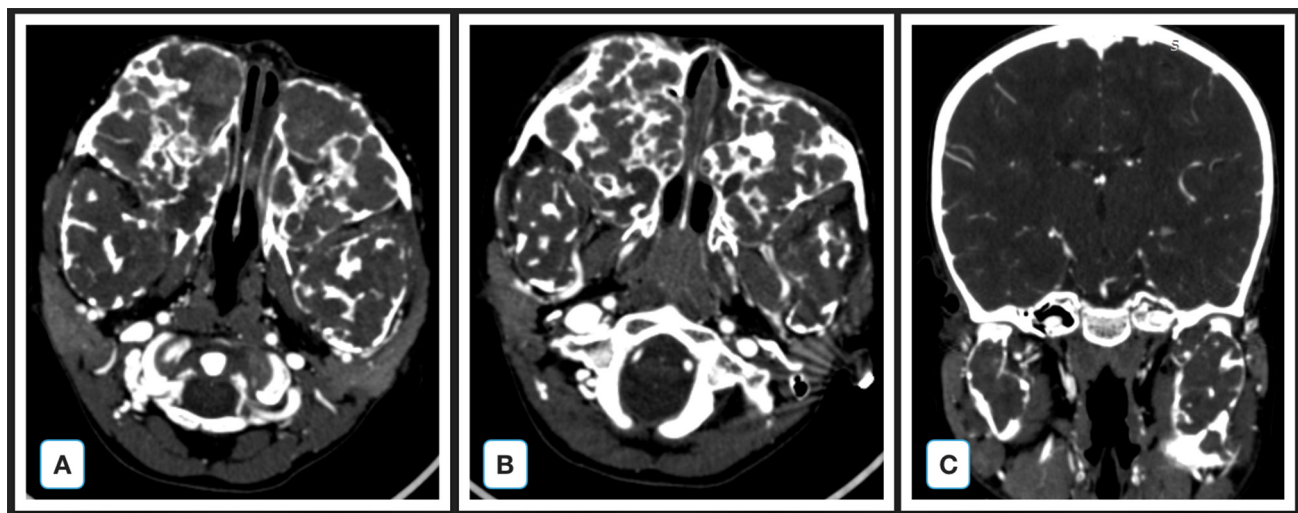


Fig. 1 Contrast-enhanced CT. (A, B) Axial sections. (C) Coronal reformat image showing classic expansile lesion with soap bubble pattern involving both the maxilla and mandible with soft tissue enhancement. (C) Bilateral condylar involvement.

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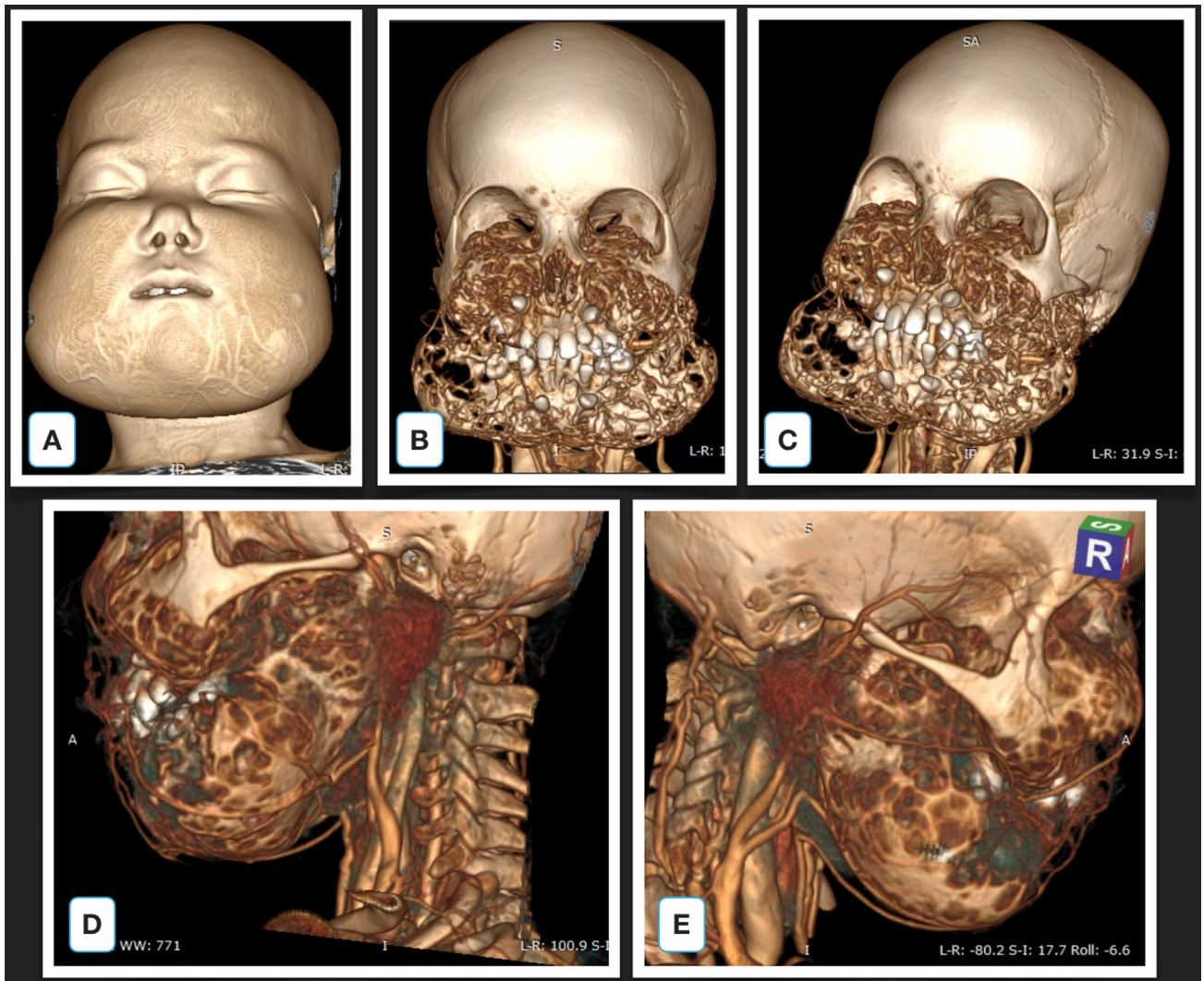


Fig. 2 Volume-rendered images. **(A–C)** Expansile lesion with soap bubble pattern involving both maxilla and mandible. **(D, E)** Arterial feeders to the lesion.

The patient was planned for a two-stage surgical correction. Stage 1 included a multidisciplinary approach with the interventional radiology team for presurgical embolization followed by surgical curettage. Stage 2 was cosmetic recontouring of the jaw after 6 months of stage 1 treatment.

Preoperative embolization was performed under general anesthesia. Through a right femoral arterial access, bilateral common carotid and selective external carotid angiography was performed. The bony lesion was supplied by multiple arterial feeders from the branches of the right

internal maxillary artery (both the superior and inferior alveolar arteries), left internal maxillary artery (superior alveolar branches), and bilateral facial arteries. There were no arterial feeders from the internal carotid artery. Angiographic findings included enlarged arterial branches and a tumor-like contrast retaining blush in the late capillary phase. Each arterial branch supplying the hypervascular areas of the lesion was superselectively catheterized with a 2.7F microcatheter (Progreat, Terumo) and embolized to stasis using 300 to 500 μ m polyvinyl alcohol microspheres

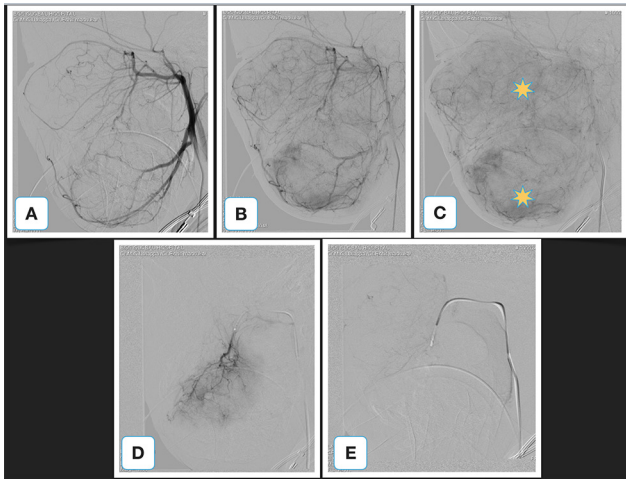


Fig. 3 External carotid angiography. (A–C) Contrast blush of the bony lesion (best seen on C [yellow star]). (D) Selective injection of superior alveolar artery with contrast blush. (E) Postembolization image showing absence of contrast blush.

(Cook Medical). Postembolization angiography showed stasis of flow to the lesion with no new feeders from either the internal or external carotid arteries (►**Fig. 3**). Postembolization, the patient underwent surgical curettage on day 2 (►**Fig. 4**). The surgery went well with no hemodynamic complications and very minimal blood loss (50 mL). The patient continues to do well (►**Fig. 5**) and is on schedule for cosmetic recontouring surgery.

Discussion

Cherubism is a rare hereditary fibro-osseous dysplasia characterized by bone degradation and fibrous replacement. The World Health Organization (WHO) classifies cherubism as a non-neoplastic tumor of the mandible.^{6,7} Based on the radiographic location of the lesion in the jaw, Seward and Hankey has proposed a grading system⁸: grade I—involvement of bilateral mandibular molar regions and ascending rami, mandible body, or mentis; grade II—involvement of bilateral maxillary tuberosities (in addition to grade 1 lesions) and diffuse mandibular involvement; grade III—massive involvement of the entire maxilla and mandible, except the condyles; grade IV—involvement of both the jaws, including the condyles.⁸ The patient in our study had a grade IV lesion that is considered to be a rare presentation. The lesion is even rarer because of its high vascularity, and according to our knowledge, only four cases of such similar highly vascular cherubism have been reported.^{9–12}



Fig. 4 Postcurettage material.

The treatment options for cherubism include conservative management and surgical management. Surgical management includes vigorous and repeated curettage and aesthetic correction surgery.¹³ Removal of the fibrotic component usually initiates ossification¹⁴; however, it can provoke postoperative enlargement, rapid recurrence, and diffuse bleeding.^{5,15} Because of the vascular nature of the lesion, bleeding can be profuse ranging from approximately 600 to 1,300 cc.^{10,15,16} Surgical management is necessary in more aggressive cases to reduce facial deformities and progression of condition after puberty.¹⁷ Preoperative embolization of hypervascular cherubism reduces significant blood loss during surgical curettage. Polyvinyl alcohol (PVA) particles provide permanent effect due to adhesion to the vessel walls, causing stagnation of flow. This results in secondary focal angioneclerosis and inflammatory reaction; 300 to 500 μm are considered to prevent nontargeted embolization. According to our knowledge, preoperative



Fig. 5 Photographs comparing pre- and postembolization + curettage taken 2 months apart.

embolization has been reported only twice.^{12,18} Our case showed a significantly reduced blood loss (~ 50 mL) when compared with that reported in the literature.¹²

Conclusion

Cherubism is a rare disease affecting the mandible and maxillary bones. Treatment is mainly surgical with curettage and reconstructive surgery. Preoperative embolization of hypervascular variants of cherubism helps reduce blood loss during surgical curettage.

References

- 1 Jones WA. Familial multilocular cystic disease of the jaws. *Am J Cancer* 1933;17(4):946–950
- 2 Waldron CA. Bone pathology. In: Neville BW, Damm DD, Allen CM, et al, eds. *Oral and Maxillofacial Pathology*. Philadelphia, PA; Saunders; 1995:456–458
- 3 Kalantar Motamedi MH. Treatment of cherubism with locally aggressive behavior presenting in adulthood: report of four cases and a proposed new grading system. *J Oral Maxillofac Surg* 1998;56(11):1336–1342
- 4 Riefkohl R, Georgiade GS, Georgiade NG. Cherubism. *Ann Plast Surg* 1985;14(1):85–90

- 5 Zachariades N, Papanicolaou S, Xypolyta A, Constantinidis I. Cherubism. *Int J Oral Surg* 1985;14(2):138–145
- 6 Yamaguchi T, Dorfman HD, Eising S. Cherubism: clinicopathologic features. *Skeletal Radiol* 1999;28(6):350–353
- 7 Regezi JA, Sciubba J. *Oral Pathology: Clinical-Pathologic Correlations*. 2nd ed. Philadelphia, PA: Saunders; 1993;345–354
- 8 Seward GR, Hankey GT. Cherubism. *Oral Surg Oral Med Oral Pathol* 1957;10(9):952–974
- 9 Brannin DE, Christensen RO. Bilateral giant cell tumors of the mandible in siblings; report of cases. *J Oral Surg (Chic)* 1954;12(3):247–251
- 10 Hamner JE III, Ketcham AS. Cherubism: an analysis of treatment. *Cancer* 1969;23(5):1133–1143
- 11 Khosla VM, Korobkin M. Cherubism. *Am J Dis Child* 1970;120(5):458–461
- 12 Koury ME, Stella JP, Epker BN. Vascular transformation in cherubism. *Oral Surg Oral Med Oral Pathol* 1993;76(1):20–27
- 13 Meng XM, Yu SF, Yu GY. Clinicopathologic study of 24 cases of cherubism. *Int J Oral Maxillofac Surg* 2005;34(4):350–356
- 14 Kaugars GE, Niamtu J III, Svirsky JA. Cherubism: diagnosis, treatment, and comparison with central giant cell granulomas and giant cell tumors. *Oral Surg Oral Med Oral Pathol* 1992;73(3):369–374
- 15 Topazian RG, Costich ER. Familial fibrous dysplasia of the jaws (cherubism): report of a case. *J Oral Surg* 1965;23:559–568
- 16 Lannon DA, Earley MJ. Cherubism and its charlatans. *Br J Plast Surg* 2001;54(8):708–711
- 17 Hitomi G, Nishide N, Mitsui K. Cherubism: diagnostic imaging and review of the literature in Japan. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1996;81(5):623–628
- 18 Henry F, Testelin S, Gauvin AC, Poirier J, Henry E. [Cherubism: the value of imaging and preoperative embolization] [in French] *J Radiol* 2003;84(11 Pt 1):1774–1778