CASE REPORT

Infantile Intraosseous Maxillary Hemangioma

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Received: October 09, 2015;
Initial review: October 28, 2015;
Accepted: August 06, 2016.

Background: Primary osseous hemangiomas of the facial skeleton mimic malignancy. Their location in maxillary sinus, especially in infants is extremely rare. **Case characteristics**: 1- month-old full term boy with maxillary swelling. **Observations**: Biopsy from oral route revealed hemangioma showing vascular channels lined by endothelial cells. Patient improved on oral steroids. **Message**: Hemangiomas should be considered as one of the differential diagnosis of unilateral maxillary swelling in infants. Steroids may serve as the primary mode of treatment as opposed to tumor excision.
Keywords: *Corticosteroids, Maxilla, Tumor, Vascular malformations.*

Hemangiomas are benign tumors of the capillary endothelium, and have varied clinical presentations. Intraosseous hemangioma is a rare tumor accounting for 0.2-0.7% of all bone tumors [1]. It is seen more frequently in vertebrae and calvarium, and is very rare in jaw bones. Two-thirds of the jaw lesions occur in mandible and one-third are found in maxilla [2]. Primary intraosseous maxillary hemangioma in infants is extremely rare [3,4].

CASE REPORT

A one-month-old male infant presented to us with progressive right maxillary swelling and alveolar fullness, first noted at 6 days of age. The child was born term, and had no significant perinatal history. No functional deficits due to maxillary mass were reported. Computed tomography (CT) revealed a heterogeneous, mildly enhancing and expansile lesion of the right maxilla, replacing the right maxillary sinus and alveolar ridge. Orbital contents were normal (***Fig.* 1**).

We suspected malignancy, and performed a biopsy from the swelling under general anesthesia via sublabial route. A fleshy bleeding mass was seen from which biopsy was taken. The mass bled profusely and the maxillary sinus was packed with sofragauze. The pack was subsequently removed gently under short inhalational anesthesia after two days. No bleeding was seen subsequently. Histopathological examination of the mass revealed hemangioma showing vascular channels lined by increased number of bland endothelial cells with no atypia or mitosis. We treated the child with oral steroids (2 mg/kg/day) for two months followed by gradual fortnightly tapering after the lesion regressed. Maxillary fullness resolved in four months. The patient is now well on regular follow-up.

DISCUSSION

Infantile hemangiomas appear in infancy, grow till one year of age, are more in females, and involute by adolescence. They usually do not involve the bone, and are histologically characterized by endothelial hyperplasia and increased number of mast cells with expression of glucose transporter-1 (GLUT-1). Congenital hemangiomas; however, have no postnatal proliferative phase [5]. This distinct behaviour disqualifies the possibility of congenital hemangioma in

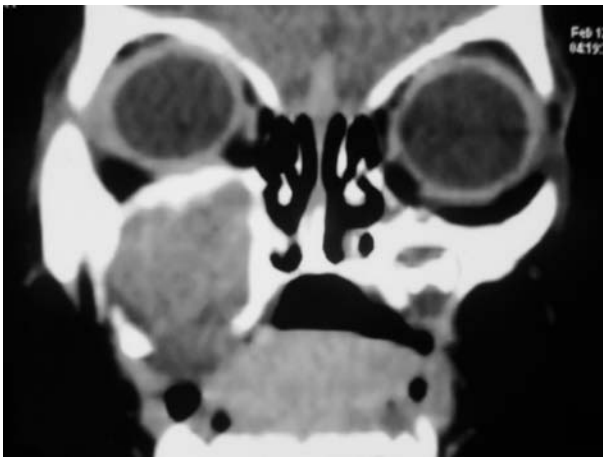

FIG. 1 *CT scan showing mildly enhancing expansile lesion of the right maxilla.*

this child, as the maxillary swelling gradually increased in size. On the other hand, vascular malformations are present at birth, have equal gender distribution and usually become more prominent by puberty because of slow progressive ectasia resulting from intraluminal flow. They do not show endothelial hyperplasia and have normal number of mast cells [4]. Venous malformations are very commonly mistaken to be hemangiomas. It was easy to confuse hemangioma with vascular malformations in this case as the presentation was congenital. We could not check GLUT-1 expression in this case because of unavailability of the marker kit but the increased number of endothelial cells on histology, relatively rapid increase in swelling size, and reversal of maxillary fullness on treatment with steroids pointed strongly towards this lesion being a hemangioma.

Intraosseous hemangiomas have a variable presentation and occur mostly in second decade of life with a 2:1 preponderance in females [6]. Incidentally, the two earlier reported cases of infantile intraosseous maxillary hemangioma were both females [3,4].

CT of such lesions may show homogenous, non-calcified masses that may remodel adjacent bone and show enhancement on contrast administration [7]. It may also cause bony destruction leading to a false impression of malignancy. Confirmation of the diagnosis requires biopsy, but that is likely to result in profuse bleeding as in our case. Hence, a very small antral window needs to be created for taking biopsy from maxilla through sublabial route so that bleeding could be minimized by pressure packing.

The treatment options of intraosseous hemangiomas include complete excision of the tumor with reconstruction using various materials. This may or may not be preceded by embolization [8]. However, these recommendations have largely been made on the basis of treatment of adult patients. Hemangiomas in infancy run growth phases of progression and involution, and there is always a possibility of the lesion resolving completely by itself with time. Moreover, any major surgical manipulation in the bony framework of an infant may result in permanent deformity. Keeping these points in consideration and with the primary aim of arresting the growth of lesion and reducing the likelihood of permanent disfigurement, we started treatment with oral steroids, which resulted in a good response. Werle, *et al*. [3] also reported complete resolution of maxillary hemangioma in an infant girl with a ten-month course of oral steroids.

We suggest that possibility of hemangiomas should always be considered in unilateral maxillary swelling in infants. Conservative treatment with oral steroids may be the first line of management in infantile intraosseous hemangiomas.

Contributors: DG and PS: case management and drafting the manuscript; IS and KG: supervision of case and revision of manuscript. All the authors approved the final version of the manuscript.
Funding: None; *Competing interests*: None stated.

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