Maxillary Labial and Left Paranasal Hemangiolymphangioma in a Female Child: A Rare Case Report and Review of Literature

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Abstract

This case report covers a rare lesion involving the left upper lip and perioral region of a 5-year-old female. The patient presented with a history of left maxillary labial and paranasal swelling for the past 4 years that increased in size during crying. It did not resolve after conservative treatment. The lesion was clinically diagnosed as a vascular anomaly. Wide surgical excision was performed after thorough investigations and planning. Excised soft tissue specimen was sent for histopathological evaluation, which confirmed the diagnosis of hemangiolymphangioma. Functional and esthetic postsurgical recovery during 4 years follow-up was uneventful. Studies reveal that hemagiolymphangioma is a rare vascular malformation that usually does not resolve with conservative treatment. Surgical intervention and long-term follow-up are essential.

Keywords

- hemangiolymphangioma
- lymphatic malformations
- vascular anomaly
- mucocutaneous lesion
- mesenchymal tumors

Introduction

Hemangiolymphangioma represents a rare lesion in the head and neck region. Perioral mucocutaneous vascular anomalies are rarely reported in literature and are seen in only 0.3 to 0.5% of the population. Lymphangiomas have a predilection for head, neck, and axilla and generally originate secondary to error in embryogenesis in areas where embryonic lymph sacs are located. They may be seen during an in utero examination on an ultrasound and clinically manifests in early childhood. Magnetic resonance imaging is considered as the imaging modality of choice. The initial few weeks up to 5 months of age is the crucial period in the growth of a child. Complications such as ulceration, distortion of vital structures, and skin disfigurement are expected to occur during this critical period.[1–4]

Typically, these lesions have a high recurrence rate and high propensity to infiltrate into the muscle and other tissues. As these lesions are extremely vascular, there are very few reports on the surgical treatment of these lesions.[4–6] Congenital low flow vascular lesions may persist throughout life and hence may require surgical excision or embolization. Localized medical care and medicines are recommended as the first line of treatment but they are usually not effective[5–8] and therefore, surgical excision is considered as the gold standard treatment.

In this report, we describe a case of a 5-year-old female with a congenital vascular lesion in the upper labial and left paranasal region.

Case Report

A 5-year-old female presented to our department with a bluish swelling on her upper lip that increased in size upon crying. The swelling extended from the philtrum to the middle one-third of the upper lip involving the skin, labial

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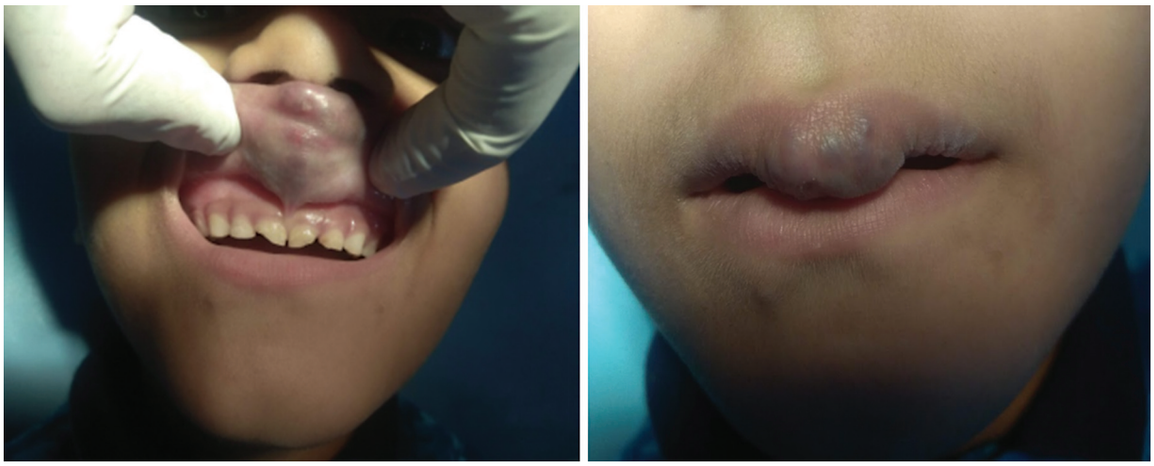

Fig. 1 Clinical preoperative views (intraoral & extraoral) showing the size and extent of swelling.

mucosa, and the muscle. She had a history of fall when she was one and a half years old. Few years later, she was given local sclerosing injections in the swelling by her general physician and was advised pharmacological management as further treatment. However, the swelling did not regress even after conservative management. On inspection, the overlying skin was stretched with bluish engorged vasculature present below the swelling. The lesion appeared as a functional and esthetic deformity on the upper lip. Clinical differential diagnosis included hemangioma, lymphangioma, and hemangiolymphangioma. Preoperative complete blood count, chest X-ray, and echocardiogram were normal and serology results for human immunodeficiency virus and severe acute respiratory syndrome coronavirus 2 reverse-transcriptase polymerase chain reaction test were also negative. On examination diascopy and turkey wattle sign were positive (►**Fig. 1**). Color Doppler scan revealed moderate vascularity in a small well-defined 21×7 mm multicystic lesion on the upper lip and underneath the skin (►**Fig. 2A**). Plain and contrast magnetic resonance imaging of face confirmed a well-defined vascular lesion involving the midline and left paramedian region involving the upper lip and philtrum predominantly on the left side up to the nasolabial fold resulting in subcutaneous bulge superficially reaching up to left nostril. A well-defined altered signal intensity t1 and t2 hyperintense lobulated, intensely enhancing lesion measuring approximately 16 mm x7.5mm was noted involving the midline, left paramedian region, upper lip, and philtrum in the superficial and deep subcutaneous planes. No obvious cortical irregularity or destruction was evident (►**Fig. 2B**).

After investigations, surgical intervention was planned. Under strict aseptic conditions, intravenous sedation with 0.5 mL Diazepam, 0.5 mL Phenergan, 0.5 mL Ketamine, and 100% Oxygen through nasal catheter was performed. Bilateral infraorbital nerve blocks were given along with infiltration in the upper nasolabial region with 2% lignocaine and 1:200,000 epinephrine. Incision was made on the vermillion side of the upper lip lesion (►**Fig. 3A**). Dissection was performed with a hemostat to reach the stalk of the lesion. Further excision was performed by dissection with bipolar cautery and the base of lesion was cauterized to control bleeding from the potential vessels. The feeder vessel in the upper labial mucosa was ligated and the wound was closed with 3-0 Vicryl sutures. After adequate hemostasis, patient left the operation theater in a satisfactory condition. Histological analysis showed stratified squamous epithelium overlying loose, fibrovascular stroma. Dilated blood vessels filled with eosinophilic material and blood were seen. Deeper stroma showed normal appearing blood vessels and muscle tissue (►**Fig. 3B**). These features were suggestive of hemangiolymphangioma. During the follow-up period of 4 years, the healing was uneventful (►**Figs. 3C–D, –4**).

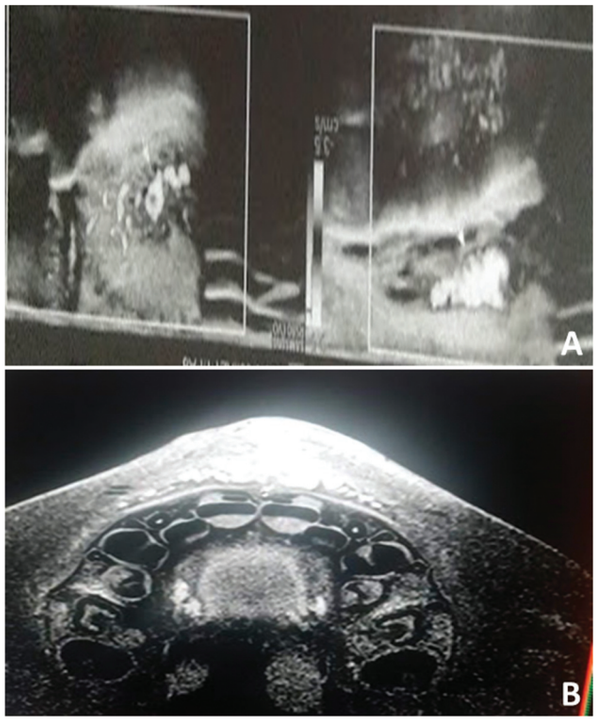

Fig. 2 Color Doppler (A) and magnetic resonance imaging scan (B).

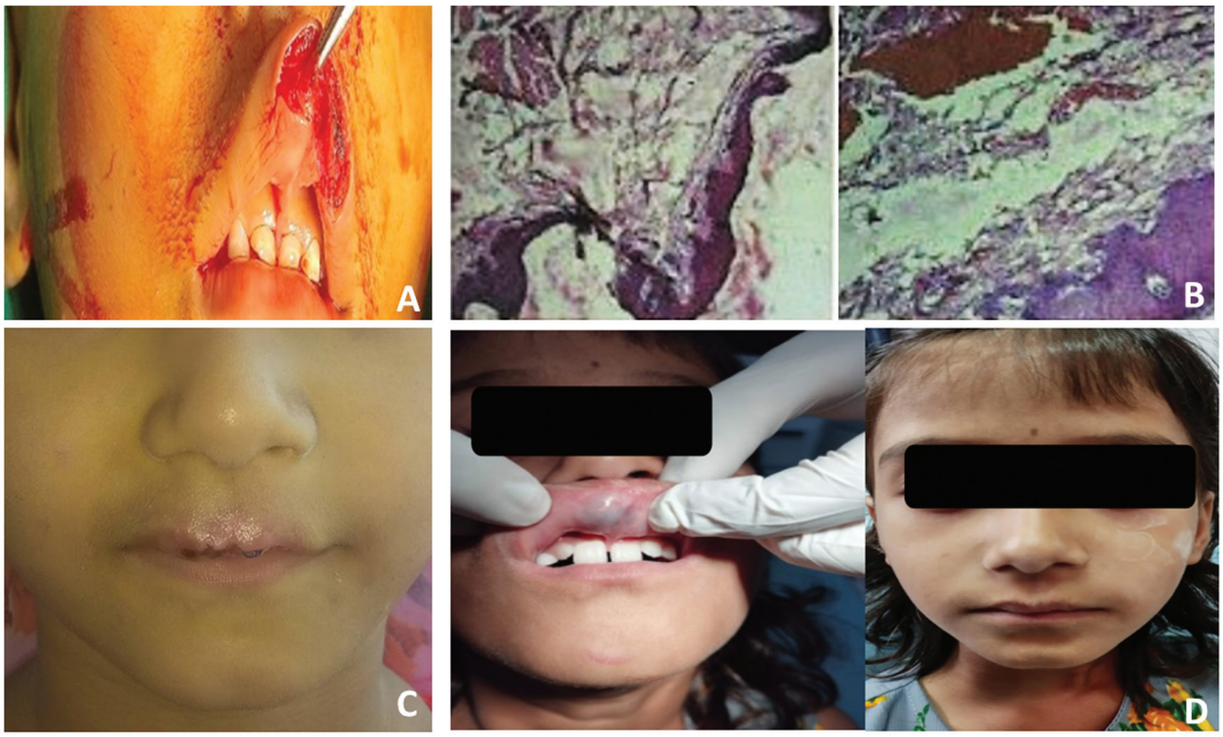

Fig. 3 Perioperative (**A**), hematoxylin and eosin stained section of biopsy (**B**), appearance of the lesion 14 days (**C**), and 2 years (**D**) postoperatively.

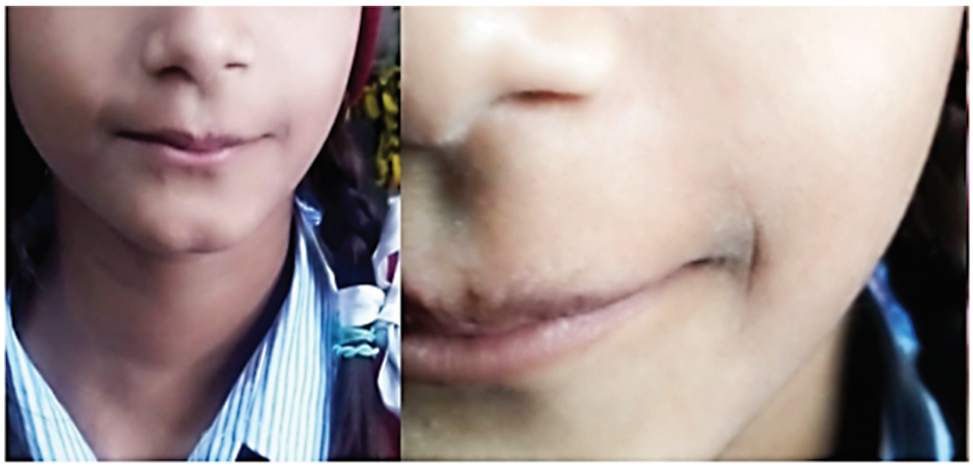

Fig. 4 Postoperative image at 4 years.

Discussion

Hemangiolymphangioma of oral and perioral region is a very rare vascular anomaly. In this case, we report a 5-year-old female with hemangiolymphangioma of upper labial region with a positive diascopy test and turkey wattle sign that is pathognomonic manifestation of vascular malformations of the head and neck region.[9,10] Spontaneous regression of hemangiolymphangiomas is rarely observed.[1,2] Various therapeutic approaches have been proposed based on the size, type, and location of lesion, as well as its association with anatomic structures and infiltration into the surrounding tissues.[5–9]

Vascular lesions can be either macrocystic or microcystic. Macrocystic disease is relatively easily treated with percutaneous image-guided sclerotherapy. It is less invasive and has an acceptable complication rate with a short recovery time. Doxycycline (100–200 mg in infants) is usually the first-line sclerosing agent because of its ease of availability and high safety profile. Other agents are bleomycin, STS, Picibanil (OK-432), Ethibloc, and ethanol. Intralesional bleomycin under sonographic guidance is generally believed to be the best

agent to achieve a bulk reduction in microcystic type. Its instillation into the stroma or solid component of the lesion leads to effective results. Rapamycin (mammalian target of rapamycin) inhibitors such as sirolimus have been reported to be effective for microcystic lymphatic malformations. Surgical resection is usually reserved as second-line therapy.[1] With the advent of propranolol therapy, local sclerosing agents, surgery, and laser treatment are no longer commonly employed.[5–7,9–12] Despite being the greatest treatment modality, propranolol still has side effects such as bradycardia and hypoglycemia. Nevertheless, complete surgical excision is still the usual treatment option for these lesions whenever possible.

Wide local excision is still considered the gold standard method for the treatment of low flow vascular malformations that compromise the function and esthetics of face.[9] In the literature, studies have demonstrated that in untreated hemangiolymphangioma, involution is usually complete by 4 years of age. However, in this case we did not see self-involution of the lesion and was therefore treated surgically. The 3-year follow-up of the patient revealed optimum functional recovery. A bluish colored engorged vasculature can still be seen in the latest follow-up visits under the labial vasculature along with a restrictive maxillary labial frenum attached to the interdental papilla between the upper central incisors. Surgical correction of the restrictive maxillary labial frenum is not recommended before the permanent canines erupt and even before any necessary orthodontic intervention.

Conclusion

Vascular malformations are usually congenital and may follow a traumatic episode. Usually, these malformations are self-limiting. However, in rare cases patient may report with persistent lesion warranting surgical intervention. Pain, mass effect, and oozing from the lesion are the common associated symptoms in persistent lesions affecting the quality of life of patient.

Conflict of Interest
None declared.

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