CASE REPORT

Parapharyngeal Hemangioma with Phleboliths

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ABSTRACT

Hemangiomas are the most frequently encountered angiomatous benign tumors of head and neck region, noticed especially during infancy and early childhood. Changes in blood flow dynamics within hemangiomas result in thrombus formation and phleboliths. The tendency of hemangiomas to undergo spontaneous involution has lead to the adoption of ‘watchful neglect’ as a common practice while managing this disorder.

We describe a relatively rare case of hemangioma with numerous phleboliths involving multiple intraoral structures and orofacial spaces in a 30-year-old male patient along with a comprehensive review of literature on the condition and its most popular treatment modalities.

Keywords: Hemangioma, Phleboliths, Parapharyngeal space.

INTRODUCTION

The term hemangioma has traditionally been used to describe a variety of developmental vascular anomalies. The current concept describes hemangiomas as benign tumors of infancy having a rapid growth phase with endothelial cell proliferation followed by gradual involution whereas vascular malformations are structural anomalies of blood vessels without endothelial proliferation and persist throughout life. Clinically, hemangiomas are the most frequently encountered angiomatous benign tumors of head and neck region noticed especially during infancy and early childhood. Changes in blood flow dynamics within hemangiomas results in formation of phleboliths which is rare finding in orofacial region. It is strikingly predominant in females and has an observed left-sided preference. Due to the rarity of these tumors and unfamiliar presentation, inaccurate preoperative diagnosis and inappropriate treatment planning are common.

We report the clinical radiographic picture of a relatively rare case of hemangioma with numerous phleboliths involving multiple intraoral structures and orofacial spaces in a 30-year-old male patient, who was managed with the traditionally popular approach of ‘watchful neglect’—observation without intervention.

CASE REPORT

A 30-year-old male patient consulted the Department of Oral Medicine and Radiology, VSPM DCRC, Nagpur, with the chief complaint of recurrent mild pain resulting from food lodgment in the lower right posterior region of the jaw from last 2 months.

Clinically, a partially erupted lower right third molar with distal pocket was present. Other striking intraoral findings included bluish areas with red elevations present on soft palate of right side and right retromolar area. Similar discolored areas were also present on the tip, right lateral border and dorsal surface of tongue (Figs 1 and 2). The tongue also appeared to have a irregular and lobulated surface with limited movement and deviation toward the right side on protrusion. The uvula was enlarged and also deviated toward right side. Other intraoral structures and mucosae were normal.

Extraorally, a solitary roughly oval swelling having approximate diameter of 7 × 5 cm having a scared, hairless central area was present in the right submandibular area...
The overlying skin was elevated, flesh-colored and soft, the swelling remained same size during eating but exacerbated during movement of the involved area. Patient gave history of presence of intraoral lesion and extraoral swelling since childhood. There were no complaints or dimensional changes associated with the intraoral pigmented areas. However, a progressive enlargement was noticed with the extraoral swelling during childhood and adolescence following which the patient consulted a plastic surgeon and was administered cryotherapy. After the therapy, further enlargement of the area was arrested for a period of almost 10 years. In last 7 to 8 years, patient has observed a very slow painless increase in the area of the swelling. On palpation fluid thrill was noticed with the compressible extraoral swelling.

Panoramic and intraoral periapical radiographs in addition to impacted lower right third molar with distal pocket and carious root stumps with periapical abscess with first molars of upper right and lower left quadrants also revealed, numerous concentric radiopacities of diameters ranging from 0.5 cm to 1 cm present unilaterally on the right side. Mucosal thickening with right maxillary sinus was present. The inferior border with right angle of mandible was irregular suggesting pressure resorption of the area (Fig. 4).

Ultrasonography, angiography and contrast-enhanced CT scan revealed large relatively well-defined mass lesion of $6 \times 8 \times 10$ cm in transverse, anterioposterior and craniocaudal diameters respectively, involving right parapharyngeal, masticator, lingual and submandibular space on right side with multiple rounded calcifications within the lesion. The lesion was supplied by branches of right external carotid artery and drained into right internal jugular vein. Widening of pterygoid fossa, crossing of the midline and involvement of left side of tongue, displacement and narrowing of nasopharynx and oropharynx were caused by the extension of the lesions. Parotid and submandibular salivary glands were not involved in the pathology. Imaging features suggested right parapharyngeal hemangioma with multiple phleboliths (Fig. 5).

Present complaint of pain was found to be associated with the distal pocket associated with impacted lower right third molar and had no connection to the hemangioma lesion.

**DISCUSSION**

Vascular malformations and hemangiomas can cause significant morbidity and even mortality in both children and adults. For a number of reasons, physicians often confuse these lesions and often use the nomenclature for classifying these lesions interchangeably and inappropriately. Latest classification of vascular anomalies has classified hemangiomas as vascular tumors which involute during childhood and noninvoluting or rapidly involuting congenital hemangiomas. Hemangiomas include dilation, proliferation and degeneration and malformations of the blood vessels. From the history and presentation of the above case, it was categorized as noninvolving congenital hemangioma.

Also, the said pathology is commonly described as occurring in Caucasian females mostly with the left side whereas the patient under consideration was an Indian male having right side pathology. Most frequently involved orofacial structures include lips, tongue followed by gingiva, mandible, palate, buccal mucosa and floor of the mouth and salivary glands with almost 80% of hemangiomas occurring as single tumor. The reported case had simultaneous involvement of multiple intraoral structures but did not have any clinical or radiographic features suggestive of salivary gland involvement, which are otherwise most common site for hemangiomas in adults.
Phleboliths are a common finding in the hemangiomas but rare in orofacial lesions. They are usually multiple in number, small and ovoid in shape with radiologic hallmark of a laminated concentric ring (onion-like) appearance.

Differential diagnosis for orofacial calcifications includes sialoliths, calcified lymph nodes, phleboliths, other vascular calcifications, cysticercoses and tuberculosis of lymph nodes or of the salivary gland itself. Phleboliths and sialoliths can be easily differentiated; sialoliths cause an obstructive sialadenitis with glandular swelling and pain, especially with meals and suppurative saliva. Also, sialoliths tend to occur singly, and are frequently larger than phleboliths; they have an elliptical shape resulting from confinement within a salivary duct. Phleboliths are distributed in a randomized pattern, while sialoliths, particularly when several are present, anatomically trace the configuration of the involved salivary duct.

As to pathogenesis of phlebolith formation, Ribbert advocated the theory suggesting that thrombi produced by slowing of peripheral blood flow may be organized and mineralized and become the core of a phleboliths. Anneroth suggested that intravascular thrombosis might be formed, become organized and secondarily mineralize under the influence of proliferating fibroblasts and contains a mineral pattern of pure apatite, calcium phosphate and calcium carbonate.

**INVESTIGATIONS**

Usually, no laboratory studies are useful in the diagnosis or management of oral hemangiomas. Several radiological and imaging techniques, such as X-ray, ultrasonography, computed tomography, magnetic resonance imaging (MRI) and angiography, are helpful in the diagnosis of hemangiomas mainly to determine their extent and flow characteristics. Contrast-enhanced CT and MRI are extremely helpful in tracing the extent of the lesion. Phleboliths can be seen inside the lesion as low-intensity areas on CT and MRI and might be considered as a specific feature of hemangiomas. Over 90% of hemangiomas are misdiagnosed radiologically as they can have unusual manifestation at unusual site especially when they contain an excessive amount of fat or fibrous tissue. Sialography is opted for conditions where salivary gland involvement and sialoliths are suspected. Panoramic view, posteroanterior view, contrast-enhanced CT scan, USG and angiography were used in present case to diagnose the condition as parapharyngeal cavernous hemangioma with multiple phleboliths.

**TREATMENT**

Management of vascular lesions is guided by location, flow characteristics, symptoms, functional disability and cosmetic deformity. As majority of the lesions undergo spontaneous involution and about 50% of hemangiomas show complete resolution by 5 years of age no treatment is advocated when discovered in childhood. Hemangiomas are usually managed conservatively, a number of preferred methods include simple observation, irradiation and injection of sclerosing agents, corticosteroid treatment, embolization, circumferential ligation of tongue lesions, lasers, cryotherapy and surgical excision. Currently, the hemangiomatous lesions considered for treatment are only the more serious types threatening vital or functional prognostics; that entail severe complications and that result in esthetic or functional after-effects. Although, based on the clinical history and physical examination, some authors argue that the cavernous type can be differentiated from the capillary type and recommend simple follow-up of these lesions unless they cause cosmetic, neurological or functional deficits, many others claim that surgery is the best way to exclude any impending risk of malignancy and report good outcomes and low recurrence rates with surgical excision.

Complications of orofacial hemangiomas can be related to the disease process or result from treatment. Complications from the disease process include hemorrhage, infection, function problems (e.g. airway, vision and hearing), thrombocytopenia and ulceration. Kasabach-Merritt phenomenon is a coagulopathy condition associated with subtype, tuffted hemangioma characterized by severe thrombocytopenia and hemorrhage because of platelet trapping within the tumor and resulting in 20 to 30% mortality. Bleeding is one of the most common reasons that patients with oral hemangiomas and vascular malformations seek care. Almost 40% of affected individuals show permanent changes such as atrophy, scarring, wrinkling or telangiectasias, also seen in the described case. As lesions become larger and, more importantly, as the flow in the lesions becomes greater, the complications increase.

Complications from treatment of hemangiomas are many, and because of this, many lesions are left untreated. Treatment of the nonproliferating lesion with minimal functional impairment becomes a risk-to-benefit decision. With all treatments, a common complication is persistence or recurrence of disease. With the therapy involving steroids and interferons, the complications are the well-known adverse effects of the drugs. Vasoformative tumors are also found to be nonresponsive to treatment.
to treatment. With embolotherapy, complications can range from minor to life-threatening which include pain, infections, fever, organ infarction and abscesses related to the introduction of external agents; exclusion of a vital segment of the blood supply; release of pyogenic materials into the circulation; migration of emboli into other parts of the body; and general effects of drugs, such as thrombin and ethanol, depending on specific kinds of embolizing devices and techniques. Use of sclerosing agents result in minor ulcerations. Complications related to sclerotherapy include skin necrosis, temporary myoglobinuria and airway compromise. Irradiation is not recommended as the amount of radiation needed is very high and has severe potential complications especially in children. Complications from treatment can be summarized as immediate complications, such as hemorrhage, airway obstruction, hematoma formation, skin necrosis and coagulopathy. Late complications include reduced mouth opening, malocclusion, drooling and dysphagia. For the present case, local irrigation and analgesics were administered to the patient for the chief complaint of distal pocket with the impacted third molar and instruction for maintenance of proper oral hygiene were given to avoid further development of periodontal problems. Surgical removal of the impacted tooth was ruled out due to potential risk of complications as the tooth was located in close proximity of vascular lesion with unusually large dimensions. Extremely slow growing: asymptomatic nature of the lesion warranted long-term observation without any further treatment at present. Carious root stumps with first molars of upper right and lower left quadrants were extracted successfully without complications.

In our review of literature, we did not come across any reported case of hemangiomas located chiefly in the parapharyngeal space with such unusually large dimensional extensions involving intraoral structures as well. Clinically significant vascular malformations are uncommon and patients with these malformations are rarely encountered in routine, primary dental facilities, rendering most dentists inexperienced in providing optimal care. Diagnosticians and radiologists may become involved in the care of these patients when tailor-made imaging or imaging-guided therapy and treatment plan is necessitated for successful management; therefore, knowledge of the imaging and pros and cons of different treatment modalities of these patients is essential.

REFERENCES