CASE REPORT AND REVIEW

Rare simultaneous occurrence of vascular malformation of tongue and odontogenic keratocyst of anterior mandible – A case report and review of literature

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Abstract

Oral cavity is mother of mysteries often revealed as an incidental finding, vascular malformation (VM) and cystic lesions to name a few. The term VM describes a variety of developmental vascular anomalies with great progress been made in the classification and understanding of these vascular lesions till date. With head and neck regions, accounting for about 60% and buccal (31%), aural (16%), nasal (11%) mandibular (5%), and maxillary (4%) regions being common site of occurrence in terms of frequency. VMs are further characterized by the predominantly affected vessel (arterial, venous, lymphatic, capillary – individually or combined) and subclassified into truncular and extratruncular vessels. Staging systems based on escalating clinical symptoms or anatomic extent have been developed by Schöbinger (for arteriovenous malformations) and has undergone several modifications in original Mulliken classification which was recently expanded and updated to incorporate new entities, genetic information, and syndromes till date. Cystic lesions of orofacial region are usually asymptomatic until secondarily infected, thereby early diagnosis, and management is of prime concern before the disease process has attained a stage of irreversibility and permanent damage. Odontogenic keratocyst (OKC) was first described by Philipsen in 1956 and has undergone numerous changes in nomenclature by the World Health Organization as primodial cyst to keratocystic odontogenic tumor, known for its typical aggressive presentation and high recurrence rate. We have reported a rare incidence of simultaneous occurrence of VM of tongue, along with OKC of anterior mandible in a young male with a brief review of the literature.

Keywords:
Odontogenic keratocyst, platelet rich fibrin, tongue, vascular malformation

Introduction

Vascular malformation (VM) is described as abnormal high or low flow lesions, abnormally developed blood vessels with varying degrees of connection with normal veins. VM although presents at birth, manifestations are presented over progression of time accounting for approximately 7% of all benign tumors with majority seen in the head and neck region of about 60%.1,2 These are associated with subcutaneous soft-tissue swelling which is produced by venous blood pooling and consist of a cluster of venous cavities with a very slow blood flow and are normally not demonstrated by conventional imaging. Cystic lesions based on its asymptomatic presentation, often is diagnosed as an incidental finding and with late manifestations of pain, swelling, pathological fracture if untreated. Based on insidious nature of presentation, simultaneous occurrence of VM of tongue, along with cystic component in anterior mandible, diagnosis, and management of lesion has significant importance, thereby prevention of late sequelae of non-intervention. The literature is suggestive of very unlikely simultaneous occurrence of both in head and neck region. We have reported a rare incidence of simultaneous occurrence of VM of tongue along with odontogenic keratocyst (OKC) of anterior mandible with a brief review of the literature.

Case Report

An 18-year-old Indian male reported to our unit with chief complaint of swelling over left lateral border of tongue since 9–10 years. He gave history of an asymptomatic swelling which

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was initially a size of about a pea, which has gradually progressed to present size with no prior history of trauma to the site (no history of pain, bleeding from the site, altered sensation, and secondary changes such as ulceration or discharge). Habitual and medical history was insignificant.

On examination, extraorally, no gross facial asymmetry was noted. Palpation was suggestive of tender, firm, mobile, and left submandibular lymph node with no fixity to overlying skin. Intraorally, on inspection, a solitary well-defined asymptomatic oval swelling was noted along left lateral border of tongue measuring approximately about 2.5*2 cm in dimensions with normal overlying surface (no ulceration/discharge). No visible pulsations were appreciable with slight reddish blue hue [Figure 1]. On palpation, further examination of the lesion suggested confirmation of above mentioned inspective findings. Depth of the lesion was about 2 cm, soft to firm in consistency, non-fluctuant, compressible, non-tender, and non-trans illuminant.

Complete examination of oral cavity revealed an incidental finding of mild buccal cortical plate expansion in 44–45 region. Hard tissue examination revealed no significant changes [Figure 1]. Based on history and clinical examination, provisional diagnosis of VM of tongue with cystic lesion over anterior mandible in 42–45 was made. Investigations were carried out to aid in definitive diagnosis which included screening hemogram, pulp vitality testing, and imaging which included conventional radiographs (OPG) and advanced imaging (Doppler study and magnetic resonance imaging [MRI] plain and contrast) which revealed following findings:

- Screening hemogram: Within normal limits.
- Pulp vitality testing: Cold test, no response (33–45).
- FNAC of lesion over tongue: No cellular elements seen, suggestive of VM.
- FNAC of lesion over anterior mandible: Straw colored aspirate.
- OPG: Well-defined unilocular translucency with sclerotic border and scalloping noted in 34 to 46 region [Figure 2].
- Doppler tongue: Well-defined lesion measuring about 18*8 mm extending to a depth of 7 mm up to transverse muscle layer, with multiple intrasional vascular channels with low resistance high-velocity flow, suggestive of VM of the left lateral border of tongue [Figure 2].
- MRI tongue (plain and contrast): Well-defined enhancing lesion appearing isointense on T1W, hyperintense on T2W in anterior one-third of tongue on the left side involving intrinsic muscles measuring 1.4*1.3*1.3 cm was noted, along with expansile lobulated unilocular lesion with thinned out cortex involving symphysis and body of mandible on right side [Figure 3].

With working diagnosis of VM and cystic lesion over anterior mandible, staged intervention was planned. Firstly, before addressing cystic lesion and lesion over tongue, intentional root canal treatment of 33–44 was carried out followed by surgical intervention of the lesion.

Informed consent was obtained after attaining physician’s clearance for procedure. Under local hemostatic measures and monitored anesthetic conditions, stay sutures were placed around the lesion over the tongue and excision was carried out in total with clearance of 2 cm provided, followed by primary closure using resorbable sutures [Figure 4]. The cystic lesion in the mandible was addressed by placement of crevicular incision from 34 to 46 region and reflection of full thickness mucoperiosteal flap for visualization of defect. On exposure, buccal expansion was evident. Osseous window was created and enucleation was carried out for removal of pathology in total. Following enucleation, chemical cauterization was carried out using carnoys solution and packing with platelet-rich fibrin (PRF) and primary closure with resorbable sutures [Figure 5]. Final histopathological reports were suggestive of VM over left lateral border of tongue with infected odontogenic cyst in 34–47 region. Post-operative period was uneventful and the patient was on regular follow-up with no recurrence [Figure 6].

Discussion

“VM” is a generalized term used to describe a group of lesions, present at birth, formed by an anomaly of angiovascular or lymphovascular structures occurring in approximately 1% of births, but majority of these patients do not present for treatment, based on occult nature of presentation. Venous malformations are common VM that can occur in any location, although they have a propensity to occur in the head and neck region, including the tongue. With head and neck regions, accounting for about 60% and buccal (31%), aural (16%), nasal (11%), mandibular (5%), and maxillary (4%) regions being common site of occurrence in terms of frequency. VM is similar to hemangiomas that they are composed of abnormal vascular channels lined with a single layer of dysplastic endothelium however, do not regress the way hemangiomas do thereby causing diagnostic dilemma to clinician.
Vascular anomalies are among the most common congenital and neonatal dysmorphogenesis, which are separated into hemangiomas and VM (ISSVA classification). VM is further being subdivided into two groups: Slow/low flow and fast/high flow malformations.[5] VM predominantly though may present at birth, but manifestation seen in later years. By contrast, hemangioma are usually not present at birth, grow rapidly during infancy, and typically involute during childhood.

VM do not have increased endothelial cell turnover instead, is structural abnormalities of the capillary, venous, lymphatic, and arterial system that grow in proportion of the individual. VM can occur as primary lesion or as a part of a regional or diffuse syndrome such as Kasabach-Merritt syndrome, Trenaunay and Parkes-Weber syndromes, blue rubber bleb nevus syndrome, Proteus syndrome, and Klippel-Trénaunay syndrome.[4,5]

Presenting initially as bluish, compressible, and nonpulsating masses, involving various superficial or deep anatomic areas, usually are asymptomatic although swelling, pain, reduced range of motion, and cosmetic issues are not uncommon. Peak incidence is seen in terms of local trauma, puberty, and pregnancy in response to altered hormonal changes.[6] Clinical presentation, along with use of advanced imaging, including MRI and ultrasonography, aids in accurate diagnosis, prompt treatment planning, and execution. With
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Treatment ranging from non-intervention to use of sclerotherapy, surgery, chemotherapeutic stabilization of lesion to recent advances in form of use of lasers, and interferon’s huge research and advances are being proposed till date; however, treatment of choice still remains controversial based on nature of presentation[7].

Ultrasonography, Doppler and MRI are the most widely used modalities of choice for assessment and demonstration of the extent of lesion in total and for planning medical, interventional, and/or surgical therapy. Sclerosing agents used to destroy the vascular endothelium through different mechanisms: Chemical agents (iodine or alcohol), osmotic agents (salicylates or hypertonic saline), detergents (morrhuate sodium, sodium tetradecyl sulfate, polidocanol, and diatrizoate sodium), and anti-cancer drugs which change the surface tension of the cell, producing direct destruction of the endothelial cell, inflammatory reaction, formation of a thrombus and fibrosis, leading to obstruction of the vessels, and shrinkage of the lesions, pre-operatively as a support to surgery or as a post-operative complement.[6,7]

Table 1: Vascular malformation-treatment modalities

<table>
<thead>
<tr>
<th>No.</th>
<th>Treatment Modalities</th>
<th>Description</th>
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<tbody>
<tr>
<td>1.</td>
<td>Observation/Wait and watch policy</td>
<td>-Asymptomatic lesion</td>
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<td>-No cosmetic and functional compromise</td>
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<td>2.</td>
<td>Surgical excision</td>
<td>-Smaller lesions</td>
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<td>-No approximation to vital structures</td>
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<td></td>
<td></td>
<td>-Isolated, symptomatic venous malformations or following sclerotherapy to improve form or function</td>
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<td>3.</td>
<td>Cryosurgery</td>
<td>-Easily collapsible lesions that are accessible</td>
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<td></td>
<td>-Minimal scar contracture, good hemostasis</td>
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<td>4.</td>
<td>Corticosteroids</td>
<td>-To inhibit VEGF-A (vascular endothelial growth factor) expression and subsequent proliferation slowing the growth thereby decreasing the size</td>
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<td></td>
<td>Prednisone at a dose of 20–30 mg/d given for 2 weeks–4 months</td>
<td>-Performed in cases with risk of bleeding and compromise of esthetic and/or physiological functions (such as speech and chewing)</td>
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<td></td>
<td>Intralesional triamcinolone acetoneide (4 mg/mL)</td>
<td>Contraindications: uncontrolled diabetic patients, pregnant patients, lactating women and regions with secondary infection in which treatment could cause edema and bleeding</td>
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<td>5.</td>
<td>Sclerotherapy (Ethanolamine oleate (EO), Sagg sodium morrhuate Sodium tetradecyl sulfate, sodium psylliate, hypertonic solution, 75% glucose, absolute alcohol, bleomycin)</td>
<td>Based on hypothesis that local vasoconstriction may cause early color change and softening of the lesion</td>
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<td></td>
<td></td>
<td>-High-flow lesions</td>
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<td>6.</td>
<td>Beta blockers (Propranolol at a dose of 2–3 mg/kg/d in 2–3 divided doses for 2–10 months)</td>
<td>-Non-specific and beta-2-selective blockers triggers apoptosis of capillary endothelial cells</td>
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<td>7.</td>
<td>Pre-surgical embolization</td>
<td>High-flow lesions</td>
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<td>8.</td>
<td>Laser therapy (Neodymium–yttrium–aluminum–garnet (Nd:YAG) laser (1064 nm), potassium–titanium–phosphate (KTP) laser (532 nm), diode laser (800–980 nm), pulsed dye lasers (585 and 595 nm), argon laser (514 nm), and carbon dioxide (CO2) laser (10,600 nm)</td>
<td>Based on photocoagulation, vaporization, or excision procedures Advantages:</td>
</tr>
<tr>
<td>9.</td>
<td>Circumferential (intratumoral) ligature</td>
<td>High-flow lesions</td>
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Table 2: Diagnostic criteria for OKC

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<tr>
<th>A.</th>
<th>Demographics</th>
<th>2–3rd decade of life</th>
<th>Posterior mandible</th>
<th>Male&gt;Female</th>
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<td>B.</td>
<td>Clinical presentation</td>
<td>Usually asymptomatic</td>
<td>Anteroposterior growth within medullary cavity</td>
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(Contd...)
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Table 2: (Continued)

| C. | Aspirate | Dirty white cheesy material
| | | Total protein content is <4 g/100 ml (Toller)
| | | Albumin: 2–4 g/dl
| | | Globulin: 0.5–2.5 g/dl
| | | Cholesterol crystals
| D. | Histopathology | - Tombstone/palisaded appearance of basal epithelial layer.
| | | - Uniformly thin epithelial lining ranging from 8 to 10 cell layers.
| | | - Uniform nuclei and parakeratinized/ orthokeratinized luminal epithelial cells.
| | | - Microcysts/satellite cysts
| | | - Relatively thin fibrous cyst wall and usually lacks inflammatory cell infiltrate
| E. | Immunohistochemistry | CK13, CK17, CK10, CK19, Ki67, pCNA, p63
| F. | Imaging | OPG: Multilocular/unilocular radiolucency
| | | Well defined sclerotic margins
| | | Root resorption
| | | Association with impacted teeth: 25–40%
| | | CT: Expansile with cystic lesion (about 30 HU) with scalloped, well corticated borders
| | | MRI: T1: High signal due to cholesterol and keratin contents
| | | T2: Heterogeneous signal
| | | DWI: Restricts due to presence of keratin
| | | T1 C+: peripheral enhancement but unlike ameloblastomas no enhancing nodular component
| G. | Associated syndrome | Nevoid basal cell carcinoma syndrome
| E. | Recurrence rate | 5–62%

same in form of decompression, marsupialization, enucleation with primary closure, chemical cauterization, bone grafting, placement of PRF, closure with buccal pad of cad as an intermediate material, peripheral ostectomy to en bloc resection and reconstruction.[9,10]

Conclusion

Early identification and classification of occult pathologies such as vascular anomalies and cystic lesions were hampered historically by the use of confusing nomenclature and categorization. Treatment range from no or minimal intervention, while others require a cohesive multidisciplinary approach. With advances in imaging and diagnostic tools, unveiling these mysteries and timely prompt intervention has improved quality of care in terms of management and prevention of late sequelae including recurrence. Understanding these nuances and complexities is essential for the most comprehensive care and optimal clinical outcomes.

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