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CASE REPORT **OPEN ACCESS**

Contemporary Diagnosis and Management of Rare Case of Angiomyolipoma of Oral Cavity

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ABSTRACT:
Angiomyolipoma is an eminently rare hamartomatous benign, soft tissue tumor. It frequently occurs in the kidney followed by the liver. One third of renal angiomyolipoma cases are associated with tuberous sclerosis whereas extrarenal angiomyolipoma cases are usually not associated with it. Till date, only 22 cases of oral angiomyolipoma have been reported. A 72-year-old woman came to our department complaining of long-term, asymptomatic swelling of her right back cheek. It was a solitary, firm, nodular, well-defined, non-tender and clinically mobile growth. The lesion was surgically respected and sent for histopathological examination, based on which the diagnosis of angiomyolipoma was made. Oral angiomyolipoma is an eminently rare histopathological lesion with no reported cases in the buccal vestibule. This case report aims to present the first case of angiomyolipoma in buccal vestibule. Although rare entity, angiomyolipoma can be considered in the differential diagnosis of exophytic soft tissue mass of the oral mucosa. Therefore, a complete resection is important for both diagnostic and therapeutic purposes.
Keywords: *Neoplasm, Renal tumor, Extra-renal tumor, Growth, Histopathology*.

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INTRODUCTION:

Angiomyolipoma is a rare begnin hamartomatous neoplasm with a mixed cellular morphology having a mesenchymal origin.(1,2) The term "Angiomyolipoma" itself depicts its specific histopathological picture which is an amalgamation of thick-walled blood vessels, aberrantly placed smooth muscle fibres with fusion of fatty tissue.(3) It broadly manifests in the kidney with uncommon extra-renal occurrence mainly in the liver followed by brain, eyes, skin, heart and lungs.(4,5) Around 30% - 40% of cases of the renal angiomyolipoma mark the presence of rare genetic disorder known as Tuberous sclerosis. (1,3,6,7) Incidence of angiomyolipoma in oral and maxillofacial region is exceptional. A total of 22 cases has been documented in the literature with 6 on the hard palate, 3 on the buccal mucosa, 3 on the upper lip, 5 on the lower lip, 5 on the tongue and 1in the parotid gland. (1-15) Hereby, we proclaim the 23rd case of oral angiomyolipoma and the very 1st case of angiomyolipoma in buccal vestibule of oral cavity.

Case Report:

A 72-year-old woman visited the Department of Oral Medicine and Radiology with the complaint of painless swelling of the right back cheek region, which had been gradually increasing for 5 years. There was a positive history of rheumatoid arthritis and dental history of multiple extractions in both the arches. Extra-orally, no facial asymmetry or any other abnormality was present. (Figure-1) Intra-oral clinical examination reveals a large solitary, pedunculated, well circumscribed growth covered with normal mucosa having white discoloration on mesial aspect of the growth due to hyperkeratosis caused from the sharp cusp of 48. The growth was present in relation to right posterior buccal vestibule. The growth was well defined, roughly oval in shape, extending from edentulous span of 47 to retromolar pad area anterio-posteriorly measuring 2cm and bucco-lingually measuring 1cm in size. (Figure-2)
On palpation, the consistency of the growth was firm in the center and soft at the periphery. There was no bleeding or discharge on provocation. The growth was non-tender and partially mobile in relation to bucco-lingual aspect. Diascopy was performed with the finger and a glass slide and no blanching of mucosa was seen. No bruit or pulse could be felt on palpation of the growth. Based on clinical findings, a diagnosis of fibroma in the right buccal vestibule was made.
Panoramic radiograph was taken. It revealed no erosion of alveolar bone. Other radiographic findings were multiple missing teeth in maxillary and mandibular arches. A well-defined homogenously radiopaque area seen in relation to the edentulous span of 36 and in proximity of roots of 37 on left side and on the right side in relation to the edentulous span of 46, 47suggestive of dense bony island. Loss of

coronal structure with remaining root wrt 13,22,23,37 suggestive of Root stumps. (Figure-3) FNAC was done which yielded negative aspiration. Following lab investigations were adviced to the patient to rule out any systemic manifestation associated with the growth such as CBC, BT, CT, RBS, LFT and KFT. CBC report revealed decreased haemoglobin levels (8.6g/dl) with increased ESR levels (54mm/hr) and all other lab findings were within normal range.

Futher on, complete surgical excision was performed for the patient. Since patient was elderly and the site the growth was posteriorly placed, we planned for excision with Zolar diode laser at 980nm and the resected specimen which was sent for histopathological examination. (Figure-4) The histopathology report under (4X) magnification of microscope revealed the presence of epithelium and connective tissue stroma. The epithelium was stratified squamous with increased thickness in stratum spinosum. The epithelium was ulcerated at one focus. The underlying connective tissue exhibited numerous thick-walled blood vessels with engorged RBCs and adipose tissue, interspersed in between parallel arranged smooth muscle fibres. The smooth muscle fibres are arranged radially to the blood vessels. Connective tissue was infiltrated by chronic inflammatory cells. (Figure-5)

Based on histopathological report of surgical specimen, the final diagnosis of angiomyolipoma was made. After the final diagnosis, the patient was sent to a General physician for complete systemic evaluation to rule out involvement of any other viscera and no systemic involvement was noted in the patient.

This article highlights a rare case at a very rare site in oral cavity i.e. buccal vestibule.

DISCUSSION

Angiomyolipoma is an exceptionally rare hamartoma especially in context of the oral cavity, initially reported in 1975 in a 39-year old male patient on hard palate followed by 21 more cases of angiomyolipoma at various sites of oral cavity involving the tongue, hard palate, upper and lower lip, buccal mucosa and the parotid gland. (1-15) The latest angiomyolipoma case was reported in the year of 2020 in a 28-year old male on buccal mucosa. (8) Hence, in reference to all the articles listed, a varied range of age from 2nd to 7th decade with slight male predilection can be appreciated. (1-15) (Table-1)

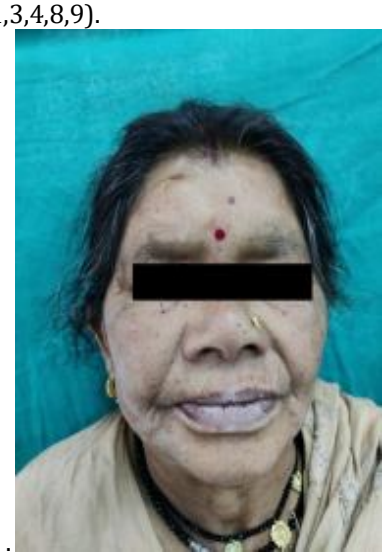
Table-1: Depicting a series of angiomyolipoma cases reported in literature in relation to oral cavity

S. N.	Author	Year	Age	Gender	Site	Treatment	Tuberous sclerosis
1.	Gutmann et al.	1975	39	M	Hard palate	Surgical excision	Not associated
2.	Komiya	1983	74	F	Buccal mucosa	Surgical excision	Not associated
3.	Iwai	1991	66	F	Upper lip	Surgical excision	Not associated
4.	Miyazaki	1994	66	M	Hard palate	Surgical excision	Not associated
5.	Yamamoto et al.	1995	62	F	Hard palate	Surgical excision	Not associated
6.			69	F	Lower lip	Surgical excision	Not associated
7.	Ide et al.	1998	60	F	Border of tongue	Surgical excision	Not associated
8.	Foschini et al	1999	68	F	Parotid gland	Surgical excision	Not associated
9.	Piattelli et al	2001	43	M	Hard palate	Surgical excision	Not associated
10.	Redman		71	M	Lower lip	Surgical excision	
11.	Lopez et al	2004	55	F	Buccal mucosa and lower lip	-	Associated
12.	Farah	2006	54	F	Hard palate	Surgical excision	Not associated
13.	Alvarez Alvarez et al	2007	52	M	Hard palate	Surgical excision	Not associated
14.	Silva et al	2007	43	F	Upper lip	Surgical excision	Not associated
15.	Koizumi et al	2008	23	M	Tongue	Surgical excision	Not associated
16.	Konstantinos	2010	78	M	Upper lip	Surgical excision	Not associated
17.	Kim et al	2011	54	M	Lower lip	Surgical excision	Not associated
18.	Yura et al	2011	61	F	Tongue	Surgical excision	Not associated
19.	Nakabayashi et al	2013	50s	M	Lower lip	Surgical excision	Not associated

20.	Morisaki et al	2016	72	M	Base of tongue	Surgical excision	Not associated
21.	Zarei et al	2019	59	M	Tongue	Surgical excision	Not associated
22.	Cheah et al	2020	28	M	Buccal mucosa	Surgical excision	Not associated

Renal angiomyolipoma differs from oral angiomyolipoma as renal angiomyolipoma is a large solitary or multiple masses which can be locally invasive in behavior, involve regional lymph nodes and due to presence of epitheloid cells, chances of the malignant transformation increases and recurrence may occur after excision. (1, 2) Whereas, oral angiomyolipoma is usually well defined with encapsulation hence, referred as benign and can be excised with ease. Recurrence or malignant transformation has not been reported for oral angiomyolipoma after excision. Chronic lymphocytic infiltrate can be present in oral angiomyolipoma with occasional presence in case of renal angiomyolipoma. (9)

Existence of the angiomyolipoma can be isolated or with a genetic disorder known as tuberous sclerosis. 80% of tuberous sclerosis patients have angiomyolipoma whereas 30%- 40% of renal angiomyolipoma patients have tuberous sclerosis; with a single listed case of oral angiomyolipoma with tuberous sclerosis by Lopez at al. (3,9) To rule out angiomyolipoma as a manifestation of tuberous sclerosis, thorough family history of skin lesions (angiofibromas referred as adenoma sebaceum), vision problems (phakoma of retina), seizures (intracranial tumor) should be taken from the patient. (9) Complications namely nephropathy, epilepsy, blindness, pulmonary hypertension, oro-facial problems such as trismus, facial asymmetry, dysarthria, dysphagia may result due to failure of diagnosisng the tuberous sclerosis with renal or extra-renal angiomyolipoma. (6,8,10) The differential diagnosis in case of such typical exophytic growth will include fibroma, fibrolipomatous hyperplasia, lipoma, angiolipoma, angioleiomyoma, hemangioma, inflammatory or reactive lesions and the definitive diagnosis will require a thorough histopathological examination. Usually a complete surgical excision in such cases is an absolute cure. (1,3,4,8,9).



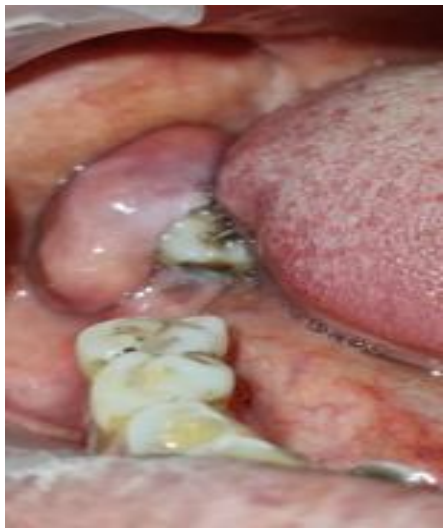
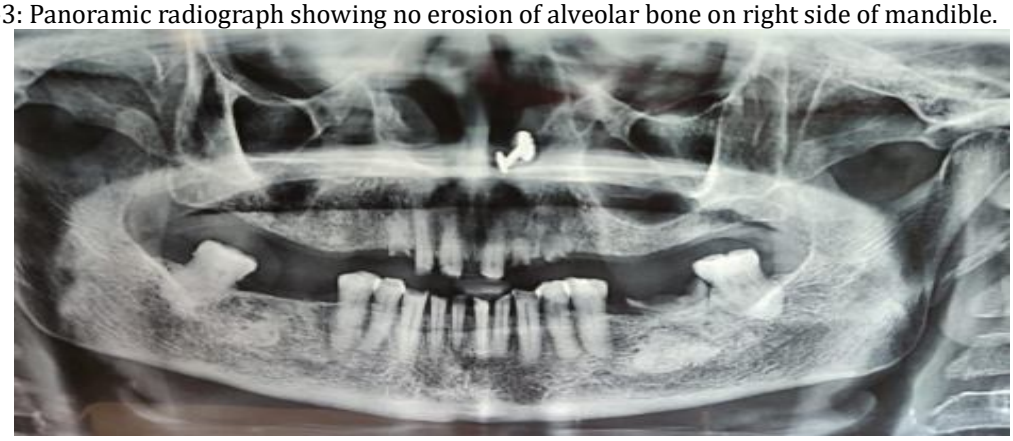


Figure-1: Extra-oral picture of the patient. Figure-2: Intra-oral picture of the growth in right buccal vestibule

Figure-3: Panoramic radiograph showing no erosion of alveolar bone on right side of mandible.



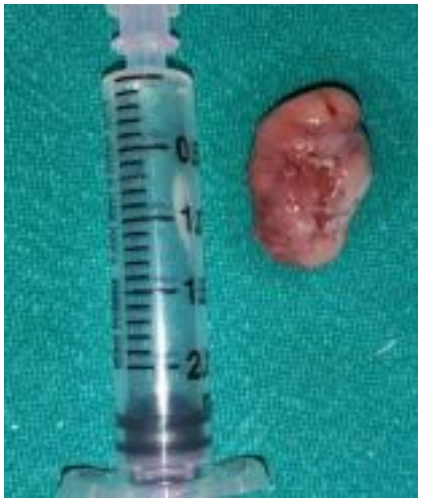


Figure-4: Excised gross specimen of approximately 1.5x1x1cm in size.

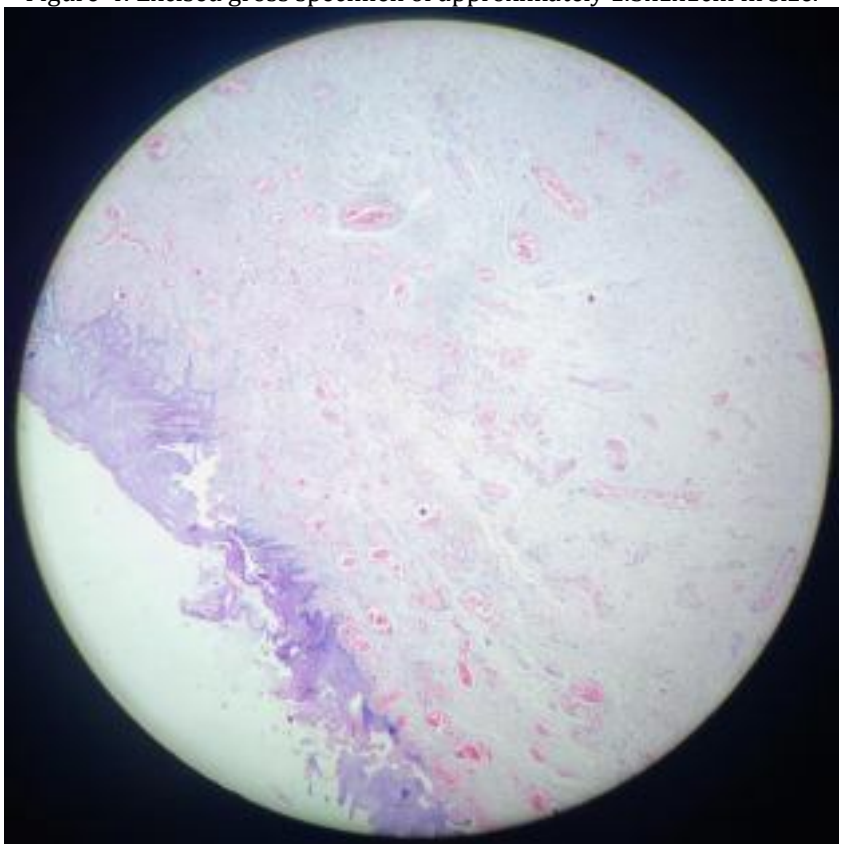


Figure-5: Histopathological picture under (4X) magnification under microscope.

It is an eminently rare and challenging case to diagnose as it is a neoplasm having mixed tissue variant of blood vessels, smooth muscle fibres and fatty tissue constitute the lesion, making a difference in elasticity or color of the lesion therefore, clinical appearance of the lesion may vary in each case. In most of the cases reported, the clinical diagnosis mentioned were lipoma, fibroma, angioma, hemangioma with the final diagnosis turn out as angiomyolipoma when studied histopathologically. Hence, histopathology is a gold standard to diagnose such cases but as an Oral medicine physician, we must have a thorough knowledge about the existence of such lesions in oral cavity. So that, we can advice a complete excision of this type of growth or lesion followed by complete histopathlogical examination and systemic review which will help in prompt diagnosis and management of such case. (1,11)

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