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A SPECTRUM OF RARE LESIONS OF ORAL CAVITY- AN EXPERIENCE IN A TERTIARY CARE HOSPITAL.

Oral Pathology

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ABSTRACT

Introduction Oral rare lesions included various groups of diseases like inflammatory, benign and malignant conditions. Because of its rarity, it is difficult to diagnose clinically so the definitive diagnosis of oral cavity lesions can be established by proper histopathological examination and it is the gold standard method. **Aims and objectives** The present study was carried out to study the histopathological spectrum of rare lesions of the oral cavity. **Material and methods** A total of 27 rare cases out of 180 cases of oral cavity lesions, studied from 1st October 2020 to 30th September 2022 (2 years duration) were included in the present study. **Results** A total of 27 cases were included in this study. Among them, 14 were males and 13 were females. Age ranged from 2 days to 72 years. The most common involved site was tongue followed by tonsil, lip, and buccal mucosa, jaw and palate. Out of 27 cases, 1 was inflammatory, 12 were benign, and 3 were malignant. The various lesions included actinomycosis, chondroid choristoma, epidermoid cyst, angioleiomyoma, fibroma, lingual/ectopic thyroid, congenital granular cell myoblastoma, pindborg tumour, schwannoma, ameloblastoma, sebaceoma, chondroid syringoma, mucoepidermal carcinoma ex pleomorphic adenoma, non-hodgkin's lymphoma, metastasis, myxoma, rhinoscleroma tonsil, keratoacanthoma, condyloma acumilatum, histiocytosis x, neuriloma, nevus spongiosus albus, bowen's disease, lentigo nevi, verrucous hemangioma and melanoacanthoma. **Conclusion** Knowledge of oral lesions is important for early diagnosis and management. Clinically rare diseases are very difficult to diagnose, hence histopathological analysis is an important tool to establish a definitive diagnosis.

KEYWORDS

Rare oral lesion, premalignant lesion, metastatic lesion.

INTRODUCTION

Oral cavity rare diseases include various groups of uncommon morbid conditions. The most effective and minimally intrusive procedure for the diagnosis of uncommon diseases is a biopsy. It is a common procedure in surgery, consisting in taking a tissue fragment from a living organism, to get a microscopic examination. This operation is frequently used in the oral cavity, for purpose of histopathologic diagnosis of an unknown lesion. Furthermore, a biopsy can be used to evaluate a wide benign lesion, when the patient refuses its complete removal for aesthetic and functional consequences of the surgical operation. [1]

India ranks 2nd in the consumption of tobacco and 3rd largest producer of tobacco in the World as per Global Adult Tobacco Survey Report 2009-2010. The association between tobacco chewing in the form of gutka and cigarette smoking with pathological, pre-cancerous and cancerous lesions has already been proven. Oral cavity lesions are usually asymptomatic. Lesions in the oral cavity generally present as ulcerations, red-white lesions, pigmentations, and exophytic lesions. Clinical classification of oral lesions is of great importance in the diagnostic process. Histopathology is still the gold standard method for diagnosis. Different sites in the oral cavity show a propensity for different types of lesions. The present retrospective study was carried out to assess various rare oral cavity lesions. [2]

In the present study, 1 case of each total of 27 different cases was included in this study and the diagnosis was confirmed histopathologically with clinicopathological correlation.

MATERIALS AND METHODS

This is a retrospective study of a total of 27 rare cases out of 180 cases of oral cavity lesions, from 1st October 2020 to 30th September 2022 (2-year duration) were included. All specimens were processed, sectioned, and stained with Haematoxylin and eosin stain and reviewed by pathologists. Histopathology reports were acquired from the database of the register.

RESULTS

A total of 27 cases were included in this study. Among those 14 males and 13 females. Age ranged from 2 days to 72 years. The most common site in the present study was the tongue 6 cases followed by tonsil, lip, and buccal mucosa 5 cases of each, the jaw with 4 cases and 2 cases in the palate. Of the 27 cases, 2 was infective, 22 cases of benign neoplasm, and 3 were malignant. The various lesions included actinomycosis, rhinoscleroma tonsil, chondroid choristoma, infected squamous papilloma, angioleiomyoma, fibroma, lingual/ectopic thyroid, congenital granular cell myoblastoma, pindborgtumourr, schwannoma, ameloblastoma, sebaceoma, chondroid syringoma, myxoma, , keratoacanthoma, condyloma acumilatum, histiocytosis x, neuriloma, nevus spongiosus albus, bowen's disease, lentigo nevi, verrucous hemangioma and melanoacanthoma, mucoepider macarcinoma ex pleomorphic adenoma, non-hodgkin's lymphoma, metastasis.

Table:1 Site distribution

Site	Number of	Percentages
Tonsil	5	18.5%
Tongue	6	22.2%
Jaw	4	14.8%
Lip	5	18.5%
Palate	2	7.4%
Buccal mucosa	5	18.5%

Table: 2: Infective lesions

Sl no.	Age (years)	Sex	Site	Diagnosis
1	11	Female	Tonsil	Actinomycosis
2	43	Male	Tonsil	Rhinoscleroma

Table 3: Benign lesions

Sl no.	Age (years)	Sex	Site	Diagnosis
3	22	Female	Tonsil	Chondroid Choristoma
4	44	Male	Tongue	Angioleiomyoma
5	20	Female	Tongue	Fibroma
6	33	Female	Tongue	Histiocytosis x
7	60	Female	Tongue	Neuriloma
8	64	Female	Tongue	Nevus spongiosus albus
9	11	Female	Base of tongue	Lingual/Ectopic Thyroid
10	2 days	Female	Upper jaw lesion	Congenital Granular cell myoblastoma
11	72	Male	Posterior aspect of the Jaw	Pindborg tumour

12	38	Male	The left lower region of the jaw	Schwannoma
13	42	Male	Posterior aspect of the jaw	Ameloblastoma
14	62	Male	Upper lip	Lentigo nevi
15	45	Male	Upper lip	Condyloma acumilatum
16	40	Female	Upper lip	Keratoacanthoma
17	58	Female	Upper lip	Sebaceoma
18	42	Male	Lower lip	Chondroid Syringoma
19	34	Female	Palate	Melanoacanthoma
20	35	Male	Upper left buccal vestibule	Verrucous hemangioma
21	24	Female	Upper left buccal vestibule	Fibrous dysplasia
22	38	Female	Buccal mucosa	Fibrolipoma
23	41	Male	Buccal mucosa	Myxoma
24	36	Female	Buccal mucosa	Bowen's disease

Table 4: Malignant lesions

Sl no.	Age (years)	Sex	Site	Diagnosis
25	67	Male	Tonsil	Non-Hodgkin's Lymphoma
26	60	Male	Tonsil	Metastasis
27	50	Male	Palate	Mucoepidermal carcinoma ex pleomorphic adenoma

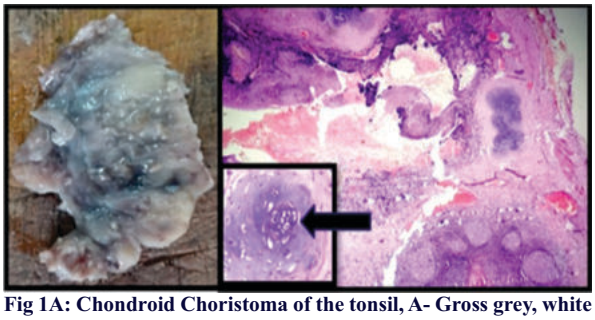

Fig 1A: Chondroid Choristoma of the tonsil, A- Gross grey, white irregular tissue, measuring 4x2x1 cm.
1B- Microscopy H&E 10X: Variable-sized lymphoid follicles with islands of mature hyaline cartilage in tonsillar tissue.

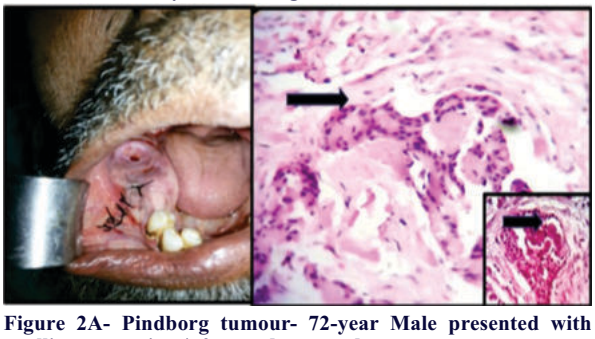

Figure 2A- Pindborg tumour- 72-year Male presented with swelling measuring 4x3cm and non-tender.
2B- Microscopy H&E 10X: Sheets of polygonal, eosinophilic squamous epithelial cells encircling homogenous hyaline material and concentric calcified areas called Liesegang rings.

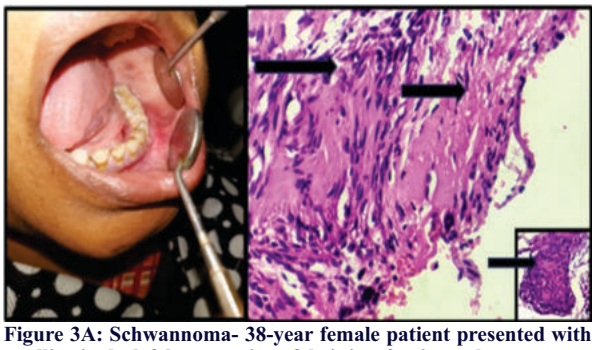

Figure 3A: Schwannoma- 38-year female patient presented with swelling in the left lower region of their jaw for 6 months
3B- Both hypercellular and hypocellular areas are noted. Bundles of spindle cells are arranged in a palisading pattern with homogenous acellular material called verrocay bodies (inset).

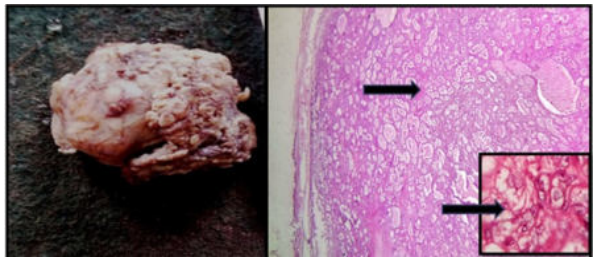

Figure 4: Mucoepidermoid carcinoma ex pleomorphic adenoma,
4A-Gross- large irregular grey-brown mass with uneven surface and engorged veins measuring 2x3 cm.
4B-Microscopy: Thin capsule with tubular proliferation filled with eosinophilic material, PAS-positive for mucin-secreting cells(inset).

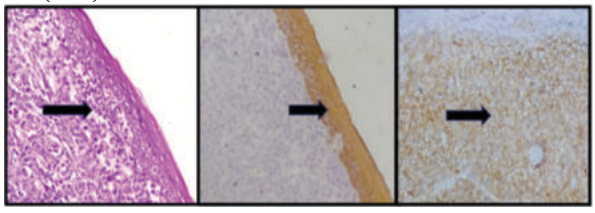

Figure 5: Non-Hodgkin's Lymphoma of the tonsil,
5A- H&E: 40X- Stratified squamous epithelium below which sheets of The monotonous population of round to oval cells is seen.
5B IHC: Cytokeratin- negative and
5C: IHC CD45- Positive in tumour cells.

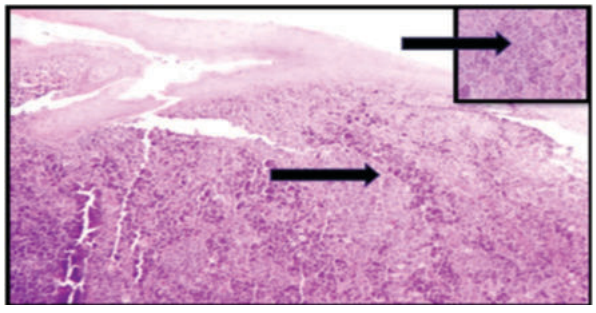

Figure 6: Metastasis in tonsil, H&E 10x: poorly differentiated metastatic carcinoma

DISCUSSION

The study included 27 cases of rare oral cavity lesions, of that only 2 case was infective etiology, 22 benign cases, and 3 malignant cases. In the present study, the age range was 2 days to 72 years. The most common site in the present study was the tongue followed by the tonsil, lip, buccal mucosa, jaw and palate.

According to data from the international literature review, unhealthy conditions are not very common in rare diseases. They are also called orphan diseases, because not very interesting for experimental research. The rarity of diseases causes greater difficulty in describing the natural history of pathology with the possible variants and planning some valid clinical research. In other words, the low prevalence causes less knowledge about not very examined diseases and frequently without adequate therapy. Therefore, patients experience two types of harm: first, a pathology that is almost always severe, and second, a lack of proper recognition, diagnosis, and treatment. These factors have led them to start providing resources to aid these disorders. All Western nations and WHO were participating in this effort, which was initiated in the USA. Because of this system, it's necessary to know exactly how much all rare diseases affect the population and their interest in health services. The rare diseases group includes several different diseases because these pathologies aren't a well-known nomologic group, but a heterogeneous group characterized only by low prevalence. Rare diseases are difficult to define precisely. Congress USA gives the only official definition of rare diseases which is a disease that affects 200.000 persons at the most, therefore one person every 1200. The European Community's Action Program for Rare Diseases (1999–2003) defines rare diseases as those with a prevalence rate of fewer than 5 per 10,000 people.[1]Among this one inflammatory lesion was found which is actinomycosis of the tonsil, which is

commonly found in young female O/E we have seen tonsils were enlarged & inflamed and the incidence is 1.3-13%. Minor infections, especially on the surface of your skin, usually heal on their own without treatment. [3]

In the present study, we found 20 years old female presented with a lesion on the tongue Fibroma. It is the most frequently occurring benign soft tissue neoplasm of the oral cavity with a 1-2% incidence. Histologically, fibroma consists of bundles of interlacing collagen fibres interspersed with varying numbers of fibroblasts and small blood vessels. [4]

Angioleiomyoma (vascular variant) accounts for up to 65% of oral leiomyomas and incidence is 0.027-0.06%. The oral angioleiomyoma is mainly found on the labial mucosa followed by the tongue. The patient's age ranges from 40 to 59 years old with a mean age of 46 years and is rarely seen in infants.[5]

A 20-year female presented with a recurrent episode of sore throat diagnosed as choristomas. we received a grey-white irregular tissue measuring 4x2x1 cm. Lesions known as choristomas are characterized by the presence of cells that are not local to the area. These have been reported in patients ranging between 3 and 80 years of age, with the tongue being the most common site and the incidence being <3%. Sixty percent of the tongue lesions are seen on the dorsum of the tongue, 32% on the lateral border, and only 8% on the ventral surface.[6]

Sebaceoma is a new term proposed to designate a distinctive rare benign neoplasm of adnexal epithelium with differentiation toward sebaceous cells. A 58 years old female presented with swelling over the upper lip from 7 years. Sebaceous carcinoma is rare with an incidence is 0.01%, aggressive, malignant tumour that is derived from the adnexal epithelium of sebaceous glands and complete surgical excision is required.[7]

A 72-year-old male presented with swelling over the posterior aspect of the jaw from 1 year. O/E size of swelling was 4*3 cm & nontender and diagnosed as pindborg tumour otherwise also known as a calcifying epithelial odontogenic tumour. First recognized by Jens Jorgen Pindborg in 1955 but was already previously described as "adenoid adamtoblastoma" by Thoma; "unusual ameloblastoma" by Tuy and "cystic odontoma" by Stoppack. The tumour is thought to arise from the epithelial element of the enamel origin. Rare primary tumours of the jaw with incidence are <1% of all odontogenic tumours. Most often located at the posterior mandible. About 50% are associated with unerupted or embedded teeth. Patient ages ranged from 8 to 92 years with a mean age of 40 years. The diagnosis is based on histology revealing areas of polyhedral neoplastic cells, amyloid and calcific deposits.[8]

Schwannoma is a benign tumour of nerve sheath origin. In the present study, we found a case of schwannoma in 38 years old female who presented with swelling in the left lower region of the jaw for 6 months. Approximately 25–45% of all schwannomas occur in the head and neck with the parapharyngeal space being the most common site but the oral cavity is very rare. Histologically tumours are encapsulated with Antoni type A hypercellular area and Antoni B Hypocellular area. Free bands of amorphous substance between rows of nuclei constitute the Verrocay bodies.[9]

In the present case study 11-year-old female presented with a swelling behind the tongue diagnosed as lingual/ectopic thyroid. The first person to note the existence of thyroid gland lingual ectopia was Hickman in 1869. Incidence is 1:4000-1:10000. The exact pathogenesis regarding the lingual thyroid is not known, but the main cause lies in aberrant embryogenesis. This abnormal development, known as lingual thyroid, usually doesn't become apparent until late adolescence or early adulthood and results from the medial anlage of the thyroid gland failing to descend from the base of the tongue to its normal location in front of the trachea. The lingual thyroid resembles the normal thyroid parenchyma histologically. Other histological patterns often seen in this ectopic tissue include fetal adenoma, colloid, and hyperplastic. [10]

The congenital gingival granular cell tumour is most frequently seen on the maxilla's alveolar median ridge than the mandible's alveolar ridge (1:3). The presentation is unique at birth as a fibrous mass arising from the gingival mucosa or alveolar ridge of the maxilla or mandible. An ultrasound can confirm a diagnosis as early as 26 weeks of gestation or in the third trimester of pregnancy. Though spontaneous regression of the lesion has been reported [11, 12], surgical excision is the only valuable therapeutic option that can benefit both mother and newborn.[11]

In the present study 42 years old male presented with the posterior aspect of the jaw which was diagnosed as Ameloblastoma. Ameloblastoma or adamantinoma is a benign odontogenic tumour of epithelial origin It was first described by Cusack in 1827, given the name ameloblastoma by Ivey and Churchill in 1930, and classified as an adamantinoma by French physician Louis-Charles Malassez in 1885. It's usually diagnosed between the 4th and 5th decades of life except in the unicystic variety (20-30 years). Approximately, 80% of the tumours are found in the mandible and the incidence is 1% of jaw cysts & tumours. Diagnosis is usually made by fine needle aspiration, which reveals palisaded basal cell layer with stellate reticulum-like epithelium.[12]

A 50-year-old male patient presented with a mass on the hard palate diagnosed as Mucoepidermoid CEPA. It is a very rare occurrence; the incidence is 2% of all salivary tumours. Within two to three years of treatment, these tumours frequently recur. A delay in treatment would adversely affect survival. When detected as confined or non-invasive carcinoma, the management of CEPA is similar to that of PA and involves surgical excision.[13]

In the present study, 67 years old male presented recurrent episodes of sore throat in the past 3 months diagnosed with non-Hodgkin's lymphoma of the tonsil. Clinical presentation includes nodal & extranodal variety with an incidence is 0.1-0.2%. Depending on its histologic form, non-Hodgkin lymphoma can be either nodular or diffuse. Large cell clusters are visible in the nodular pattern because the neoplastic cells tend to aggregate in that way. The diffuse pattern is characterized by a monotonous distribution of cells with no evidence of nodularity or germinal centre formation, with a complete effacement of lymph node architecture. (Reference Rohit Singh, Shabana Shaik).[14]

A 60 years male confirmed metastatic poorly differentiated carcinoma in the tonsil with a very poor prognosis due to a very late diagnosis, he died within 1 year of diagnosis.[15]

CONCLUSION

We believe that rare diseases will become more prevalent in the future due to the rise in average life expectancy and population heterogeneity brought on by emigration. A reasonable biological cost and accurate differential diagnosis are made possible by biopsies. We can state conclusively that biopsy is a clinical-diagnostic methodology crucial for the diagnosis of rare diseases of the oral cavity.

REFERENCES

1. F.N. Bartuli, F. Luciani, G. Cardon, F. Muzzi, F. Caddeo, L. Ottri, C. Arcuri, oral cavity rare lesions: 15 years case histories, oral & Implantology,2011;3-4:18-22.
2. Ranjan agrawal, ashok chauhan , parbodh kumar, Spectrum of Oral Lesions in A Tertiary Care Hospital, Journal of Clinical and Diagnostic Research. 2015 Jun, Vol-9(6): Ec11 EC13.
3. Mahboob Hasan, Amit Kumar, a case report of Actinomycosis and tonsillar disease, BMJ Case Reports, 2011;1-5.
4. Sakthivel VS, Krishnan CG, Manishaa V, Sumithra K, Mukesh S, Et Al. Traumatic Fibroma of The Tongue – A Treatment Modality Study. J Med Case Rep Case Series 2(4):1-3
5. Apostolos Matiakis, Panagiotis Karakostas, Achilleia-Maria Pavlou, Eleftherios Anagnostou, Athanasios Poulopoulos, Angioleiomyoma of the oral cavity a case report and brief review of the literature, J Korean Assoc Oral Maxillofac Surg 2018;44:136139.
6. Nezam S, Nishat R, Khan SA, Shukla JN. Chondroid choristoma of the tongue: A rare case report. Natl J Maxillofac Surg 2022;13;1:121-6
7. Heba Al-Khashnam, Hisham Burezq, Ibrahim Al-Aradi, Humoud Al-Sabah and Khalid Al-Abdulhadi, Unusual Malignant Transformation of Recurrent Sebaceoma A case report, Clinical Medicine: Oncology 2008;2:389–392.
8. Ida Marie, Tabangay-LimRaymund Noel C. MallariNoelito M. LacsamanaDexter Domingo Z. PazArthur Rico R. VillafuertePeter Raymund M. Quilendrino, Recurrent calcifying epithelial odontogenic tumour (Pindborg tumour): A case study, Oral Oncology EXTRA (2005) 41, 259–266.
9. Marc cohen, Marilene B. Wang, Schwannoma of the tongue: two case reports, Eur Arch Otorhinolaryngol (2009) 266:1823-29.
10. L. K. Surej Kumar, Nikhil Mathew Kurien, M. M. Jacob1, P. Varun Menon, Sherin A. Khalam, cade report developmental defect lingual thyroid, Annals of Maxillofacial Surgery |January - June 2015 | Volume 5 | Issue 1;104-7.
11. Monal Yuwanati, Shubhangi Mhaske, Ashok Mhaske, Review article, Congenital Granular Cell Tumour – A Rare Entity, Journal of Neonatal Surgery 2015; 4(2):17;1-6
12. Hariram, Shadab Mohammad, Laxman R. Malkunje Nimisha Singh Sugata Das Gagan Maheta,A case report of Ameloblastoma of the anterior mandible, National Journal of Maxillofacial Surgery | Vol 5 | Issue 1 | Jan-Jun 2014 |47-50.
13. Lily Daniel, Shalinee Rao1, Radhakrishnan Muthusamy, Deepika Yerramsetti, Mucoepidermoid carcinoma ex Pleomorphic adenoma of Lacrimal gland: A rare

presentation, Indian Journal of Ophthalmology 2014;62:743-6.
14. Singh R, Shaik S, Negi BS, Rajguru JP, Patil PB, Parihar AS, et al. Non-Hodgkin's lymphoma: A review. J Family Med Prim Care 2020;9:1834-40.
15. Shujhat Khan1 Giulio Anichini2 Areeb Mian1 Haider Kareem2 Nelofer Syed2 Kevin O'Neill, Tonsillar Carcinoma Spreading Metastases to Central Nervous System: Case Report And Literature Review, Journal of Neurological Surgery Report 2021;82:e11-27.