CARTILAGINOUS CHORISTOMA OF THE TONGUE: A CASE REPORT WITH IMMUNOHISTOCHEMISTRY IN A SECONDARY HEALTH CARE FACILITY

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ABSTRACT

Background: **Soft tissue cartilaginous choristoma (STC) is a mass of mature hyaline cells occurring in abnormal location. It is also referred to as extra-skeletal choristoma; a rare lesion that frequently occurs on the tongue in oro-facial region.**

The objective is to bring to the attention of Clinicians especially in secondary health care facilities the need of thorough investigations of tissue specimen to prevent misdiagnosis and avail themselves every diagnostic tool in the management of all oral swellings.

Case presentation: **A 7-year old boy presented in Dental clinic in a secondary health care facility with history of an asymptomatic, non-tender and slow-growing swelling on the dorsum of the tongue of 3 years duration. Surgical excision was done under general anaesthesia. Histopathological and immunohistochemical evaluations were the tools used to establish its diagnosis.**

Conclusion: **Soft tissue choristoma is rare benign swelling that mostly occur in the anterior two-third of the tongue and can be misdiagnosed clinically but diagnostic tools like histopathology and immunohistochemistry will help confirm it.**

Keywords: Choristoma, Tongue, Secondary health facility, Immunohistochemistry

INTRODUCTION

Soft tissue choristomas are rare mesenchymal benign neoplasms, also referred to as extraskeletal choristoma.[1] This lesion was first described by Berry in 1892, typically occurs on the lateral border of the anterior two-third of the tongue, while the ventral surface and the posterior one-third are rarely involved.[2] Cartilaginous choristoma describes an island of tumuor like mass of histologically normal hyaline tissue occurring in an abnormal location ("heterotopism"); which has been attributed to embryonic developmental abnormalities of the neural tube.[3,4] Importantly, choristoma should be categorized from entities like hamartomas and teratomas; while hamartomas are non-neoplastic developmental malformations which may be unifocal or multifocal, comprising a mixture of cytologically normal mature cells and tissues in a disorganized architectural pattern which are indigenous to the anatomic location; with one of its constituents predominating.[3] Teratomas on the other hand, are true neoplasms composed of a variety of parenchymal cell types of more than one germ layer.[3] Soft tissue cartilaginous choristoma also known as soft tissue chondroma,[4,5] constitutes about 1.5% of benign soft tissue tumours.[1,4] Orofacial choristomas are very rare but occur mostly in the tongue, masseter, preauricular region, gingiva, tonsil, and nasal cavity.[4,5] They are mostly seen between 3rd and 4th decade of life and usually asymptomatic, slow growing well defined nodules extending into the surrounding tissues.[4,6] We present our experience of soft tissue choristoma of the tongue in a Lagos secondary health facility.

CASE PRESENTATION

A 7-year old boy-child presented in the Dental Clinic, Somolu General Hospital with a painless, slow-growing and asymptomatic mass of 3 years duration. There was no history of trauma, only discomfort during swallowing.

Clinical examination revealed an elevated oval swelling on the dorsum of the left half of the tongue which was about 2x2cm in its widest diameter, extending mediolaterally at around the posterior limit of the anterior two-thirds (Figure 1) . The overlying mucosa

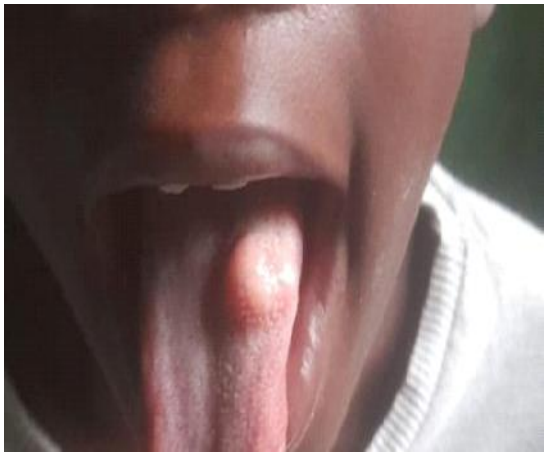


Figure 1: Pre-operative picture of the lesion.

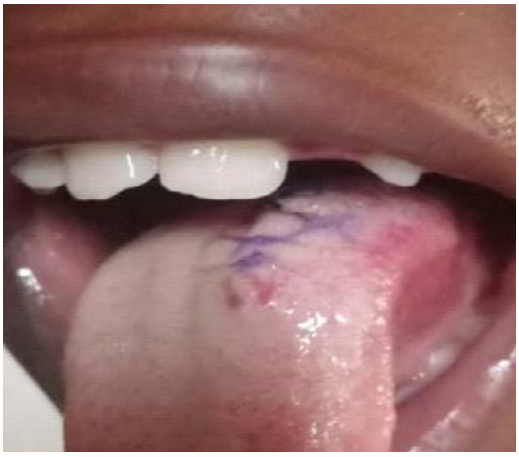


Figure 2: Post-operative picture

was normal. No tenderness was elicited and the lesion was firm in consistency.

The following differentials were made: Fibroma, Rhabdomyoma and Neuroma. Patient was admitted into the hospital and scheduled for surgical excision under general anaesthesia having secured both the parental consent and assent of the child. The excision was uneventful and patient was discharge five days post-operative after making full recovery (Figure 2). The excised specimen was sent for histopathologic review (Figures A,B&F) and subsequently

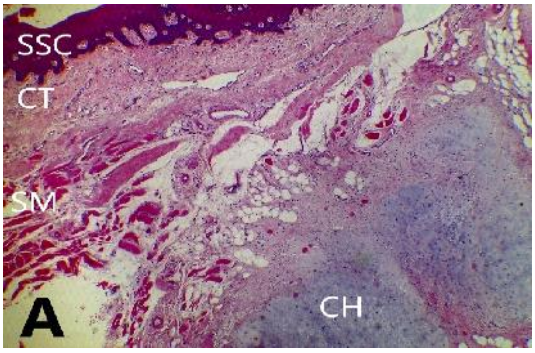


Figure A

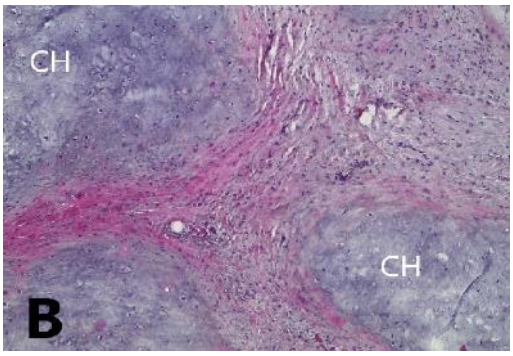
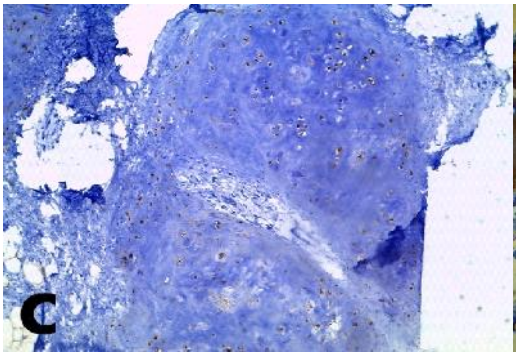


Figure B

H&E stains of the tongue tissue displaying a mucosa comprising stratified squamous epithelium (SSC), submucosa (connective tissue - CT) and an underlying muscularis made up of skeletal muscle fibres (SM).
Figure A (x 4 obj): Shows lobules of mature chondroid tissue (CH) surrounded by bundles of skeletal muscle fibres (SM).
Figure B (x 10 obj): Shows higher magnification of same tissue displaying lobules of chondroid tissue separated by fibrous septae



Figures C

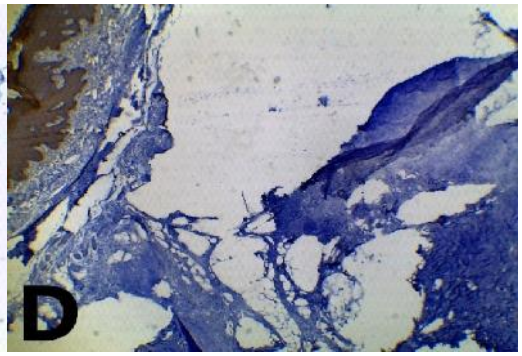


Figure D

Figure C (x 10 obj): Shows prominent cytoplasmic and nuclear positivity of the chondrocytes for S100 IHC stain.
Figure D (x 4 obj):Shows the negative AE1/AE3 IHC stain in the cartilage in the lower right corner in this micrograph (note the retraction artifact seen in the middle of the micrograph). There is a diffuse cytoplasmic staining of the overlying squamous epithelium (upper left) which is expected.

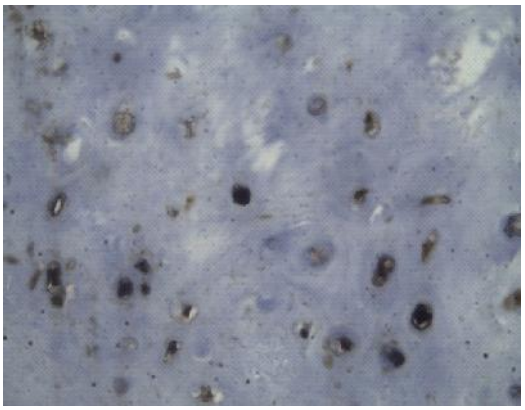


Figure E

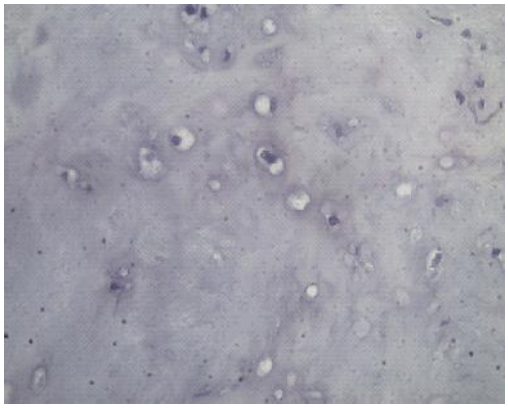


Figure F

Figure E(x 40 obj): Showing higher magnification of prominent cytoplasmic and nuclear positivity of the chondrocytes for S100 IHC stain.
Figure F(x 40 obj): Showing higher magnification of H&E stain for chondrocytes.

immunohistochemical evaluation (Figures C,D&E) and a definitive diagnosis of Cartilagenous Choristoma of the tongue was made. There was no recurrence after a year follow-up.

DISCUSSION

Soft tissue choristomas (STCs) of the oral cavity are rare benign tumours and possible differentials are neurofibroma, pleomorphic adenoma, ectomesenchymal chondromyxoid tumour, lingual thyroid, thyroglossal duct cyst and peripheral ossifying fibroma.[7,8] About one hundred cases have been reported in the English literature;[8] one described at the Lagos University Teaching Hospital, Nigeria, by Effiom *et al.*[4] and this present case will be second to be recorded in Nigeria to the best of our knowledge. The tongue is the commonest intra-oral site of its occurrence (78-85%) and other sites may include the buccal mucosa, alveolar mucosa, submandibular region, submental region, masseter and hard palate in descending order.[5,8] This present case agrees with observations from other studies[5,6,7,8,9] that tongue is the commonest site of its occurrence, and particularly the middle third of the dorsum of the tongue.[10,11]

STC is known to occur between ages 1month to 80 years,[6,12] but are commonest in the third and fourth decades of life with mean age of 31.97±18.04.[12] Sex predilection remains controversial. Kim *et al.*[5] and Sakrikar *et al.*[7] both reported almost equal distribution in sexes, Vescovi *et al.*[6] reported male preponderance while other studies[10,11,12] noted a higher female predilection.

The pathogenesis of the origin of STC is not clear, but theories about this include undifferentiated multipotent mesenchymal cells that eventually differentiate to its cartilaginous component. Also, the possibility of incomplete resorption of embryonic cartilaginous lingual septum during foetal development persisting post-natally may explain the chondromatous proliferation along the midline of the tongue.[9,10] This theory seems to be consistent with the present case due to the fact that the lesion occurred in a seven year old boy, extending from the midline to the lateral border of the tongue which was noticeable 3years before visiting the clinic. Furthermore, this present case occurred in the middle third of the tongue almost at the junction of the anterior two-thirds with posterior third, seemingly pointing to the hypothesis of entrapment embryonic cartilaginous remnant of the lingual septum during development of the tongue from 1st, 2nd and 3rd branchial arches.[11] STC may present with discomfort, swelling, gagging and dysphagia.[5,8,11] Our case presented with discomfort during swallowing.

Other hypotheses of occurrence of STCs include the following: (i) Originating from cartilaginous embryonic rests, (ii) Metaplasticchondroid tissue, (iii) Derivation from pluripotent cells, (iv) Neoplasm or teratoma with a preponderance of cartilage and (v) Mixed salivary gland tumour with a predominance of cartilage.[1,7]

The embryonic rests theory suggests that this lesion originates from heterotopic cartilage remnants from any of the first four branchial arches and it is believed that these multipotential cells are misplaced during development and then sequestered in the tongue.[10,11] This theory explains the wide distribution of cartilaginous choristoma within the tongue and the possible embryonic origin from remnants of Meckel's cartilage. This seems to agree with the histology of this present case showing areas of well circumscribed multilobulated chondrocytes with intertwining fibrous band and adipose tissue (Figures A,B&F) which was also the findings by Sakrikar *et al.*[7]

The "metaplastic theory" suggests that trauma, and chronic irritation may stimulate metaplastic development of STC as reported by Jeyasivanesm *et al.*[1] and Effiom *et al.*[4] in which the patients had history of chronic irritation and trauma. This conflicts with this present case, as there was neither trauma nor any source of chronic irritation found in this 7-year old boy.

Immunohistochemically, this present case revealed high cytoplasmic immunoreactivity in chondrocytes to S-100 protein stain (Figures C&E), which consistent with observations by Kim *et al.*[5] and Pereira *et al.*[9] The negative immunoreaction to Pan-cytokeratin AE1/AE3 stain (Figures D) ruled out tumour of epithelial cells,[9] this is consistent with observations from other studies.[1,10] This present case with no history of trauma nor chronic irritation; with patient's age and anatomical site seems to suggest foetal proliferation of embryonic cartilaginous developmental aberration.

Surgical excision is the treatment of choice.[5,11,12] Recurrence is rare, but has been reported.[12] In this present case, however, there has not been recurrence after follow-up period of one year.

CONCLUSION

Cartilaginous choristoma is a slow growing and rare benign lesion which is commonest on the tongue in the orofacial region. The clinical, histological and immunohistochemical evaluations provide the definitive diagnosis of this present case. .

Conflict of Interest Statement

The authors affirm that they have no conflict of interests to declare.

REFERENCES

1. **Jeyasivanesm DL,** Muhamed SP. Soft tissue Chondroma of anterior Gingiva: A rare Entity. Case Rep Dent. 2018 Mar; 2018:3642827
2. **Min KO,** Lee KY, Kang CS, Shim SI *et al.* Extraskeletal Chondroma Arising from the Tongue and Tonsil: Three cases report. Korea J Pathol 1993; 27(4):421-432.
3. **Nezam S,** Nishat R, Khan SA, Shukla JN. Chondroid choristoma of the tongue: A rare case report. Natl J Maxillofac Surg 2022; 13: 121-126.
4. **Effiom OA,** Umeizudike KA, Olawuyi AB, *et al.* Choristoma of the gingivae following trauma: An unusual case in a Nigerian with review of relevant literature. Nig J Dent Res 2018; 3(1):45-49.
5. **Kim Y,** Moses M, Zegarelli DJ, Yoon AJ. Cartilage Choristoma (Soft Tissue Chondroma): A rare presentation in the Lower. J Clin Pediatr Dent 2009, 33(3):253-254.
6. **Vescovi P,** Meleti M, Merigo E, Manfredi M *et al.* Soft tissue Chondroma of the Oral Cavity: An Extremely Rare Tumour Localized on the Hard Palate. Case Rep Med. 2014; 2014:414861.
7. **Sakrikar A,** Agrawal U, Pimpale G, Agrawal P. Cartilaginous choristoma of tongue- A case report and review. J Oral MaxillofacPathol 2022;26:395-398.
8. **Te BC,** Ahmad MA, Yunus MRM, Paiman NM. Osseous choristoma of the tongue: A rare disease. Bali Med J 2020; 9(2): 413-415. DOI:10.15562/bmj.v9i2.1771.
9. **Pereira GW,** Pereira VD, Pereira Junior J de C, da Silva RM. Cartilaginous choristoma of the tongue with immunohistochemical study. BMJ case reports 2012; 10.1136/bcr-2012-006752.
10. **Semwal S,** Joshi D, Gupta V, Kapoor N. Cartilaginous choristoma of tongue: A rare case report. J Oral Maxillofac 2019; 23:S40-2.
11. **Gkouveris I**, Kapranos N, Mitrou GG. Lingual Osseous Choristoma: Case report and literature review of this rare entity. J ClinExp Dent. 2023 Jan; 15(1):e79-e81.
12. **Shamloo N,** Modanloo K, Khaleghi A. Osseous choristoma: Report of a case on a palate and literature review. Clin Case Rep. 2023;11:e8355.