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Case Report

Benign extra skeletal chondroma of the tongue in a 16-year-old female

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ABSTRACT

Chondromas are tumors that result from chondroid differentiation most frequently occurring in areas where chondrocytes are present. Rarely, they can present in soft tissues, such as the tongue. This case report will review the extra skeletal chondroma found on the posterior tongue of an asymptomatic 16-year-old female. Discussed within this case report will include the importance of histopathological analysis in distinguishing enchondroma versus chondrosarcoma and review the malignant transformation rate. Additionally, it will compare the trends of chondromas found in the oral cavity that have been previously published. This case will highlight the importance of recognizing chondromas in unusual places, such as the origin of soft tissues, and emphasize the importance of early diagnosis, surgical intervention and providing optimal outcomes for young patients.

Keywords: Chondroma, Enchondroma, Chondrosarcoma, Oral lesion, Chondrocytes

INTRODUCTION

Chondromas are benign solitary central tumors composed of mature well differentiated hyaline cartilage. They generally exhibit limited growth potential, are not locally aggressive, and have a low probability of malignant transformation. Chondromas develop from nests of growth-plate cartilage that may become entrapped in the medullary canal of the metaphysis or in the metaphyseal diaphyseal junction. Such hamartomatous proliferations persist as islands in the bone and develop via endochondral ossification influenced by growth factors. Chondromas are classified as enchondromas when they occur in the medullary canal, periosteal or juxtacortical chondromas when they occur on the surface of the bone, and synovial chondromas when they arise from the synovial sheaths of tendons.[1] A rare form of a chondroma, an extra-skeletal soft tissue chondroma, is a benign tumor of mature hyaline cartilage occurring outside of bone in extra-synovial locations without attachment to the periosteum. Although the majority of extra-skeletal soft tissue chondromas occur in the hands and feet, previous case reports highlight the rare manifestations of such lesions in the head and neck region. This case report serves to elucidate the unique presentation of an extra-skeletal soft tissue chondroma in the posterior tongue region of an asymptomatic 16-year-old female.

Extra skeletal soft tissue chondromas are extraosseous and extra synovial chondromas most commonly identified in the hands and feet. Epidemiologically, extra-skeletal soft tissue chondromas (STC's) may occur at any age but are most prevalent between 30-60 years of age with a male preponderance. The most frequent clinical presentation is a slow-growing firm painless or occasionally tender soft tissue mass. While the etiology is unknown, STC's harbor the FN1/FGFR1 gene fusion.[2] Macroscopically, this benign tumor appears well-demarcated and nodular that is grey-white-bluish in color. The lesions are usually less than 3 cm in diameter.[2] Histologically, the lesion displays a mature type hyaline cartilage and myxoid matrix outlined by fibrous septa. Clusters of large chondrocytes are noted floating in myxoid along with fine calcifications or

endochondral ossification. Pathology reveals the lesions displays limited mitotic activity and no abnormal mitotic figures with variable infiltrate of histiocytes and stain positive for S100 protein.[2,3] Although magnetic resonance imaging (MRI) imaging feature a well-demarcated lobular and heterogenous appearance with T1 signal hypo to isointense compared to muscle and T2 signal hyperintensity the definitive diagnosis is established histologically upon surgical resection.[3]

Establishing the risk of malignant transformation of extra-skeletal STC's is difficult due to its clinical rarity. To date, malignant transformation of extra-skeletal STC's has not been described in the literature; however, chondrosarcomas have been described and whether precursor lesion transformation occurred cannot be ascertained. Therefore, the risk cannot be definitively concluded due to the limited presentations available. Understanding the paucity and limitations of the literature, examining the risk of malignant transformation seen in osseous chondromas, such as enchondromas and periosteal chondromas may provide insight into prognostics and risk stratification. The risk of malignant transformation in a solitary lesion is controversial.[1] However, the presence of multiple chondromatous lesions dramatically increases the risk of transformation into secondary chondrosarcoma.

Notably, the risk of malignant transformation in patients with enchondromatosis such as Ollier disease (multiple enchondromas with a propensity for continuous slow growth) or Maffucci syndrome (a syndrome characterized by numerous enchondromas and associated extra skeletal soft-tissue angiomatosis involving the skin, soft tissues, and visceral organs) is documented at 10-30% and as high as 100%, respectively.[1]

The treatment of extra skeletal soft tissue chondromas centers on alleviation of symptoms if present and on histological confirmation of the lesion post resection. In contrast to osseous chondromas where impending pathologic fracture or transformation into chondrosarcoma must be considered, STC's are absent of such risks.[1] Nevertheless, resection carries inherent risks and complications such as compromising critical anatomic structures and/or the risk of lesion recurrence estimated at 15-20%.[2]

The curious development of a chondroma on the tongue is derived from the fact that the tongue is composed of striated muscle and is glandular, lacking any native chondrocytic mesodermally destined cells. This case report will explore the development of a chondroma located on the posterior tongue of an asymptomatic patient and review current cases published. Chondromas rarely manifest in the oral cavity, and more rarely involving the tongue, so this comprehensive report will review the literature in aim to enhance understanding clinically, and provide additional insights into management strategies regarding this infrequent occurrence.

CASE REPORT

A 16-year-old female with a past medical history significant for only sinusitis presented to the pediatrician for a small marble sized mass on the posterior tongue that she had for greater than 10 years. She denied any change in size or pain. She has never previously had treatment or biopsy with no imaging of the lesion. She denied otalgia, loss of sensation, or dysphasia. Referral was placed to ENT office for surgical excision and workup.

No imaging was performed given the size and lack of symptoms described by the patient. The mass was surgically excised and gross anatomy revealed a gray-tan consistency lesion measuring 1×0.9 cm to the depth of 0.5 cm. Histological examination revealed tissue that was mildly acanthotic with benign stratified squamous mucosal epithelium with a slightly lobulated but well circumscribed submucosal nodule composed of mature cartilaginous tissue characterized by mature chondrocytes within a cartilaginous stroma. The tissue was surrounded by a very thin rim of fibroadipose tissue. There was no evidence of inflammation, atypia or malignancy identified.

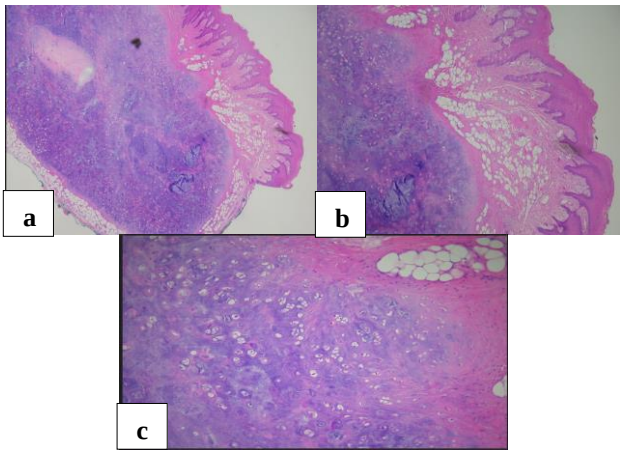

Figure 1: (a) Low, (b) medium and (c) high power view of lesion.

Low, mid, and high-power examination demonstrates squamous mucosa with a proliferation of benign, well circumscribed appearing cartilage involving the subepithelial tissue. The left half of the images demonstrates the proliferation of chondrocytes while the right side demonstrates benign oral tissue for orientation. There is no evidence of overlying squamous atypia or findings to suggest reactive metaplasia. The cartilaginous component does not demonstrate any evidence of significant cytologic atypia or evidence of malignancy. These findings are compatible with an extra skeletal chondroma involving the oral tongue.

The patient will be followed by primary care. There is no need for ENT follow up, as this tissue was benign and not causing symptoms.

DISCUSSION

Chondromas are often found in the pelvis and ribs, and they rarely occur in soft tissues. Soft tissue chondromas occur mainly in the fingers, with head and neck chondromas accounting for only 10% of all cases.[4] In a review of the 33 cases documented worldwide published in 2011 of 46 chondroma lesions isolated the to the oral cavity, the tongue was the most frequently occurring place of this rare entity.[4,5] Between this case series in 2011 aforementioned and 2019, one more tongue lesion had been reported until this case report of this 16-year-old. To our knowledge since 2019, the case of this 16-year-old girl is the latest chondroma of the tongue documented worldwide. Reported cases indicated this lesion in both include males and females ranging from all ages. These reports have identified soft tissue chondromas of the tongue localized to the lateral tongue, ventral tongue and dorsum of the tongue. In these cases, many came to clinical attention due to patients presenting with symptoms of dysphagia, speech impairment, and foreign body sensation. The average time for individuals to present to the clinic with this condition is approximately 7 years based on published data and symptoms typically arise several years after the lesion develops. This case involves a 16-year-old who self-reports a lesion persisting for 10 years, showing no growth and remaining asymptomatic, adding an intriguing aspect to the presentation.[5]

Literature reports that the oral cavity soft tissue chondromas are most commonly seen in adults, with the female sex more prevalent and the common areas being the lateral tongue and dorsum of the tongue.[6,7] While the lateral and dorsum of the tongue are the most common, the anterior ⅔ is generally where the lesions are found, making this posterior lesion a rare documented occurrence.[8] When comparing pediatric specific population, it appears additionally there is predilection towards female sex, like in the case report presented above.[7]

There are two theories for the etiology of soft tissue chondromas: metaplastic theory, which states trauma, and chronic irritation, which is most commonly seen in the elderly population. The second theory is the embryonal theory, which states that the branchial cleft cartilage remnants can get dislocated during development and may become placed within the tongue.[9] In the 16-year-old female reported in this case report, while embryonal theory or metaplastic theory could be plausible, given no inflammatory process visible on histology slides, metaplastic theory is likely the origin of the lesion in this case report. While most oral lesions documented have occurred in non-pediatric patient populations adding to the novelty of this case, given the young age of self-reported lesion, the brachial cleft cartilage remnant is a highly probable theory. When looking at documentation of patients who presented as young as 3 whose parents reported lesion time of one year, it is fair to assume that this lesion was most likely not second to inflammatory causes.[7] When reviewing the literature in pediatric patient, the variability in whether or not symptoms were present is evident, as well as lack of reported trauma to the area.

A review of limited pediatric lingual chondromas available, a previously reported case of a 10-year old female, conversely to the patient described above, had a speech impairment that was attributed to a 3-month swelling on the left side of the tongue with no visible physical changes other than a palpable mass.[5] Many pediatric patients had no symptoms and a long period of indolence prior to presentation, with average of pediatric presentation being 2.5 years with the very limited case reports available.[4-8] When looking at adult population who notated growth starting in pediatric years, the average time to presentation was found to be 22.5 years ranging in adults in the early 20s to early 40s.[7] When reviewing the available data for pediatric oral chondromas in general, of the 47 cases known to date excluding this report above, to our knowledge it appears, less than a quarter of those cases are attributed to pediatric causes, and the overwhelming majority is located on the tongue followed by the gingiva.[5,7,10-11]

The importance of this report lies in its revelation that chondromas, despite their potential for growth, can persist in a small and asymptomatic state for an extended period—specifically, ten years, as in the context of this case report above. This finding is particularly noteworthy when contrasted with the aforementioned average, chondromas tend to manifest symptoms and become noticeable within a span of 7 years from onset.

This extended period of asymptomatic growth challenges the conventional understanding of chondromas, emphasizing that they can remain clinically silent and evade detection for a significant duration. While the majority of cases that had symptomatology presented earlier, this demonstrates challenges of management of chondroma and early detection of the very low risk chondrosarcoma. This is demonstrated by the case of a 54-year-old female who had a 2-year history of a posterior, nontender, non-symptom causing tongue swelling that grew within a year prior to presentation histology revealed to be chondrosarcoma.[12]

Treatment for chondromas has preferentially been both diagnostic and therapeutic via surgical removal. While the necessity of removing chondromas for histological staining to ensure correct identification of the lesion, it is also important due to the risk of local recurrence occurring around 10-15%.[5]

While historically, documentation does not reveal malignant transformation to be a prominent concern and has been theorized to approximately 1%, follow up should continue as chondrosarcomas have been reported to occur in the same region.[13] Given the tongue lacks chondrocytic cells, histopathologically a nodular lesion consumed by small chondrocytes within lacunar pockets is indicative of a chondromal lesion present on the tongue.

CONCLUSION

This case stands out for its distinctive features, marked by the onset of soft tissue chondroma during the patient's pediatric years. The condition's enduring and indolent nature, coupled with lack of symptomatology preceding presentation, further distinguishes it. Notably, the tumor's location on the posterior aspect of the tongue adds a layer of complexity. Emphasizing the significance of proactive education, early removal to prevent symptoms, and the consistent adoption of wide surgical excision as routine treatment. Surgical intervention, while imperative, introduces inherent risks, particularly the potential for damage to local blood vessels and nerves. The documentation of this case report serves as a valuable contribution, offering insights that can inform future approaches to pediatric chondromas of the tongue.

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