Ectomesenchymal chondromyxoid tumor: A rare case report

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Abstract

Ectomesenchymal chondromyxoid tumor (ECMT) is a rare benign neoplasm of uncertain histogenesis, which exclusively involves the oral cavity, particularly the tongue. Clinically, it presents as slow growing, painless, firm, submucosal swelling exclusively occurring on the anterior dorsum of the tongue. Histopathologically, it comprised well circumscribed, unencapsulated lobular proliferation of fusiform and polygonal cells, with varying degree of cellularity, with neoplastic cells often seen in a myxoid, chondroid or hyalinized background. Until date, only 40 cases have been reported in the literature. Most documented lesions involve anterior tongue, however 2 cases in posterior tongue and one palatal tumor has been described. Here, we present a rare case diagnosed clinically and histopathologically as (ECMT) in a 7-year-old girl with the size of the lesion 5.0 cm making this case even rarer and throwing some light on this distinct entity.

Keywords: Benign, chondromyxoid, ectomesenchymal, tongue, tumor

Introduction

Ectomesenchymal chondromyxoid tumor (ECMT) is a benign tumor, a rare entity which has recently been added to the group of lesions affecting the tongue. The uniqueness of this lesion lies in the fact that it mostly affects the anterior dorsum of the tongue usually occurring as a submucosal swelling on the anterior third of the dorsum of the tongue. “ECMT” was first described by Smith et al. following review of all myxoid, chondromyxoid, and myoepithelial tongue lesions from the files of the Armed Forces Institute of Pathology in 24-year period and only 40 cases have been reported until date. The reason for the limited reported cases of ECMT is probably because it is being misinterpreted as other similar chondromyxoid lesions such as focal mucinosis, ossifying fibromyxoid tumor, pleomorphic adenoma, myxoma, myoepithelioma etc.[1,2]

Clinically, ECMT presents as a slow growing asymptomatic swelling usually seen on the anterior dorsum of the tongue, however, 2 cases presenting on the posterior tongue have also been documented. In addition, a case of ECMT on the hard palate has been reported but due to lack of appropriate supporting documentation, its diagnosis has been the subject of controversy. The size of the lesion usually varies from 0.3 cm to 2.0 cm and the age ranges from 9 years to 78 years, with a mean age of 39 years. No sex predilection is seen. The diagnosis of ECMT is based on the clinical as well as histopathological and immunohistochemical confirmations.[2,3]

Case Report

A 7-year-old female came to the Cancer Research Institute, Ahmedabad, Gujarat with a painless swelling on the anterior dorsum of the tongue. Her medical history was non relevant. On oral examination, a solitary well-defined nodule, since birth and currently measuring 5.0 cm × 3.0 cm × 3.0 cm was located on the left dorsum of the anterior two-third of the tongue, involving the tip of the tongue. There were no secondary changes on the surface except for the partial depapillation. On palpation, the nodule was sessile, firm in consistency, mobile and nontender. Her extra-oral examination was non relevant and lymph nodes were non palpable. Provisional diagnosis of granular cell tumor/fibroma of tongue was given [Figure 1].

Following the clinical examination, fine needle aspiration cytology was done which was suggestive of a granular cell tumor. Subsequently, surgical excision was performed, following which it was submitted for further histopathological examination.

On gross examination, the specimen was smooth surfaced, firm in consistency measuring 5.0 cm × 3.0 cm × 3.0 cm. The cut surface was non glistening and white in color.
Histopathologically, the lesion was well circumscribed and unencapsulated showing a lobular growth pattern. The lesional cells were arranged in the form of cords and strands in a net-like pattern within a myxoid background. The nuclei were round to oval with hyperchromatism. Few multinucleated cells were also seen. The lesional cells were seen infiltrating into the skeletal muscle. The connective tissue stroma showed few chronic inflammatory cells. Thin bands of collagen fibers separated the lesional tissue giving it a lobular appearance [Figure 2]. For the conclusion, immunohistochemistry was performed, and the tumor cells were found to be positive for vimentin confirming the mesenchymal origin of the lesional cells. Therefore, a final diagnosis of ECMT was given on the basis of histopathological and immunohistochemical evaluation with the correlavancy to the clinical history of the patient[4,3] [Figure 3].

Discussion

Ectomesenchymal chondromyxoid tumor of the tongue is a rare benign neoplasm that was initially described by Smith et al. Clinically, ECMT presents as a slow growing, painless, firm, well-circumscribed submucosal swelling and size of the lesion usually varies from 0.3 cm to 2.0 cm. Its occurrence has been noted in a wide range from 9 years to 78 years; However, most cases have been reported in adults with no gender predilection. Histopathologically, the lesion usually presented as a well circumscribed and unencapsulated lesion showing a lobular growth pattern and the neoplastic cells were separated from the overlying epithelium by a thin layer of loosely compressed connective tissue. The cells were arranged in the form of cords and strands in a net-like pattern against a chondroid/chondromyxoid background with round to oval nuclei, showing vesiculation to hyperchromasia. Few multinucleated cells were also noted. The connective tissue stroma showed few chronic inflammatory cells.[2,5]

Microscopic features of this case paralleled to the finding of other reported cases of ECMTs. The presence of myxoid areas always raise the suspicion of the other myxoid lesions such as myoepithelioma, oral focal mucinosis, soft tissue myxoma, glial choristoma, myxolipoma and nerve sheath myxoma, which need to be considered in the differential diagnosis. However, these lesions were excluded on the basis of histopathological and immunohistochemical findings.[6,7]

The uniqueness of our case lies in the fact that it is presented in a 7 year old girl which is a rare finding as most of the cases of ECMT are reported until date in adults, with only 5/40 cases of ECMT reported in first two decades of life[8] and secondarily about the size of the lesion, as the usual size is found to be 0.3- 2.0 cm, while in our patient the size of the lesion was 5.0 cm, which is furthermore rare.[9]

Treatment of choice is conservative surgical excision.[10] Only 2 cases have been reported to recur suggesting only 5% recurrence rate. However, they were successfully managed in the 2nd surgical intervention. Our patient was also treated by surgical excision and is on follow-up for the prognosis of the surgically treated site [Figure 4].
Conclusion

Ectomesenchymal chondromyxoid tumor is an uncommon intraoral asymptomatic mesenchymal soft tissue neoplasm located on the dorsum of the tongue. This case adds to the limited literature on ECMT and emphasizes the need for this rarely occurring tumor to be considered in the differential diagnosis of nodular lesions affecting the tongue. The occurrence of this lesion in children is even rarer. This emphasizes the importance of recognizing this entity when presented with a similar clinical scenario. Immunohistochemical expression of vimentin and smooth muscle actin is very helpful in confirming diagnosis, suggesting a probable mesenchymal and neural origin of this rare entity.

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