Case Report

Pediatric extramammary myofibroblastoma in the head and neck region: a case report

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ABSTRACT

A rare form of mesenchymal neoplasm that typically presents in older men and women is mammary myofibroblastoma. Even so rarely, these benign tumours may occur outside of the breasts in soft tissues; which then are called mammary-type myofibroblastoma. There can be marked variability in the histological composition of these benign spindle cell tumours. Along the milk line is the commonest presentation of these extramammary myofibroblastoma. Here we have an 8 years old female patient presented to the department of otorhinolaryngology with a painless, fusiform swelling over the right lateral aspect of tongue in the past 6 months. Intra oral examination revealed a non-tender, non-ulcerated fusiform shaped mass approximately 3×2 cm firm in consistency arising from the right lateral border of the tongue. FNAC was done which showed spindle cell tumour tongue. The lesion was diagnosed as mammary type myofibroblastoma on immunohistochemistry.

Keywords: Mammary type myofibroblastoma, Head and neck myofibroblastoma, Pediatric myofibroblastoma

INTRODUCTION

Mammary-type myofibroblastoma (MFB) was first described by Wargotz et al in 1987.1 It was described as a benign spindle cell neoplasm with myofibroblastic differentiation. It is similar to MFB of the breast histologically. It is rare in occurrence and when found, reported among older men and postmenopausal women. Commonly, it is found along the embryonic milk line; which extends from mid-axilla to medial groin. Painless, slow growing mass is the most common form of presentation in such tumours.2,3 The age at presentation ranges from 35 to 85 years.2,4 Bland spindle cells with myofibroblastic differentiation, prominent mast cells, fatty component, and hyalinized stroma is the presentation on histology. Diffuse positivity for CD34 and desmin is the commonest immunohistochemical profile in these tumours.2 Variable positivity has also been reported.5 Surgical removal is the standard management for such tumours as malignant behavior or recurrence has not been described. Only a few cases have been reported so far in the literature, most of them located along the embryonic milk-line.2,4,5 We describe a very rare case of extramammary MFB in the head and neck area and the first case to be reported in the tongue.

CASE REPORT

An 8 years old female presented to the department of otorhinolaryngology with a painless, fusiform swelling over the right lateral aspect of tongue in the past 6 months. Initially, the swelling was small in size and then gradually progressed to the present size (Figure 1). Movement of the tongue was normal and mouth opening was adequate. Intra oral examination revealed a non-tender, non-ulcerated fusiform shaped mass...
approximately 3×2 cm, firm in consistency, arising from the right lateral border of the tongue. The patient’s medical history was unremarkable. Fine needle aspiration from the lesion over the tongue showed spindle cell tumour. Wide local excision of the tumour was done under general anesthesia after which the specimen was sent for histopathological examination. Postoperative course was uneventful and the patient was discharged from hospital. She was followed up closely on outpatient basis.

Histopathological microscopic examination revealed a cellular tumour composed of oval to spindle cells with high nuclear to cytoplasmic ratio. Nuclei showed moderate atypia. Tumour cells were mainly in sheets and short fascicles. No necrosis was observed. Edges were infiltrative with tumour invasion to adjacent skeletal muscle. Features were suggestive of malignant spindle cell tumour (Figure 2 and 3). Hence, immunohistochemistry was performed.

Immunohistochemistry (Figure 4) revealed the tumour cells express SMA (focal), CD34 (very focal) EMA (weak/focal) and PR and immune negative for desmin, S-100 protein, myogenin, Myo D1, cytokeratin, SOX 10, ALK-1 and ERG. The tumour cells showed loss of expression of IHC marker retinoblastoma protein. The lesion was diagnosed as mammary type myofibroblastoma on immunohistochemistry.

**Ethical approval**

Written informed consent from the patient for the publication of clinical details and/or clinical images was obtained.

**DISCUSSION**

Myofibroblastoma is a rare, benign, mesenchymal tumor of breast. An extra-mammary location is rare, and usually occurs along the embryonic milk-line, which extends from the mid-axilla to medial groin. The mammary-type myofibroblastomas described in the literature are well-
circumscribed, un-encapsulated masses, consisting of spindle-shaped cells haphazardly arranged in fascicles of various sizes, along with bands of hyalinized thick collagen in the background. Intralobular fat lobules are also seen, and intranuclear grooves have been described.

So far, five cases have been reported outside the pelvis, including cases in the mandibular region, popliteal fossa, abdominal wall, liver, and back. Hox et al has described a case of head and neck MF in the infraauricular region. In our case, we describe a case of pediatric MF in the tongue of the head and neck region. According to the literature search, this is the first such case to be reported from liguinal region and second such case in head and neck.

Clinically, these tumors present as slow growing, painless masses, without evidence of local lymphadenopathy. In our case too, we had a similar presentation.

In the review of the 20 previously reported cases by Ghafar et al, showed that mammary-type myofibroblastoma occurs at a relatively old age, with an average reported age of 52.5 years (range 35 to 85 years), and slight male predominance (M:F ratio=1:0.7). In our case, the presentation was in an 8 year old girl.

The most common immunohistochemical profile seen in these tumors is diffuse positivity for CD34 and desmin. Morphologic variation in mixture tuned matched filtering (MTMF) can cause diagnostic confusion, and unusual immunophenotypes such as CD34 and/or desmin negativity can also occur, albeit infrequently. Loss of Rb expression may be a valuable aid in supporting the diagnosis.

In our case, the Immunohistochemistry revealed the tumour cells express SMA, CD34, EMA and PR and immune negative for Desmin, S-100 protein, Myogenin, Myo D1, Cytokeratin, SOX 10, ALK-1 and ERG. The tumour cells showed loss of expression of IHC marker Retinoblastoma protein. The lesion was diagnosed as mammary type myofibroblastoma on immuno-histochemistry.

Regardless of the histologic appearance, immunophenotype, and anatomic location, MTMF has virtually no potential for recurrence or metastasis, even with positive excision margins.

CONCLUSION

Mammary type myofibroblastoma is a rare and a benign entity which is commonly encountered along the milk line. IHC is commonly used modality in the aid of its diagnostics. Herein, we report a case of head and neck MTMF; only the second such case in this region and the first case in oral cavity. Its occurrence in a young girl also makes it a unique case to report. Further studies are required to study the occurrence of MTMF outside the milk line.

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