Fibromyxolipoma: A Palatal Mass

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Abstract: Fibromyxolipoma is a rare tumor that is considered as a variant of spindle cell lipoma, it is characterized by extensive myxoid changes and the presence of stellate cells with dentritic process. This report presents a rare case of fibromyxolipoma arising from the palate, which is easily misdiagnosed as soft tissue malignancy. Here we are going to discuss the clinical presentation, diagnosis, and management of fibromyxolipoma.

Keywords: Fibromyxolipoma, palatal mass, excision, biopsy

Date of Submission: 06-09-2021

Date of Acceptance: 20-09-2021

I. Introduction

Fibromyxolipoma is a rare entity and its presence in the palate is very uncommon. It is a variant of spindle cell lipoma usually present as a mass in the head and neck, shoulder, and extremities between the age of 30 to 50 years, and males are affected more. Fibromyxolipoma is a benign soft tissue tumor with features of solitary fibrous tumor and spindle cell lipoma. Macroscopically it present as a well-defined, encapsulated lesion with a focal mucinous or gelatinous cutting surface. Microscopically it appears as well vascularised with matured adipose tissue, spindle, and stellate cells and myxoid stroma with abundant collagen.

Case Presentation

A 30-year-old female came to our OP with complaints of mass in the oral cavity for 1 year which is increasing in size without pain or bleeding. She had difficulty in swallowing. She does not have the habit of betel nut chewing. And also there is no similar mass in her body anywhere.

On examination her general condition and vitals are normal. Examination of oral cavity shows 4cm x 3cm pedunculated mass which is pale in color, firm in consistency, arising from the junction between soft palate and hard palate on the left side which is partially obstructing the oropharynx.



Fig 1&2: showing a mass in the oral cavity.

She was admitted in the ward where she was advised for routine blood tests and MRI of the Head and Neck. Her blood test was normal and MRI shows a well-defined altered signal intensity lesion noted at the posterior aspect of the oral cavity. The lesion shows internal T2 hyperintensity with peripheral T2 hypointense rim and internal areas of T1 hyperintensity with possibility of soft tissue/salivary gland tumor.

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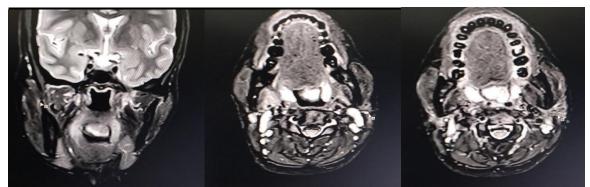


Fig 3,4,&5: shows T2 hyperintensity with peripheral T2 hypointense rim and internal areas of T1 hyperintensity in the oral cavity.

We plan for excision of the mass and further histopathological examination. Under general anesthesia, mass was excised and removed then the base was cauterized. Specimen sample sent for biopsy.



Fig 6,7&8: shows a specimen sample of the mass

After 2 weeks histopathology report came, on microscopic examination abundant myxoid background with vascularized tissue was noted with mature adipocyte scattered around the lesion. Also, have fibrotic tissue with collagenous bundles. No lipoblast and mitotic activity were noted in the lesion. Which is suggestive of Fibromyxolipoma.

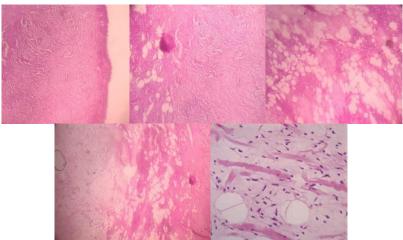


Fig 9,10,11,& 12: showing histopathological slides with features of adipose tissue with myxoidareas and fibrous tissues.

Immunohistochemistry was not done due to limited resources in our hospital.

II. Discussion

Spindle cell lipoma (SCL) is a benign adipocytictumor which not uncommon. Certain SCLs show extensive myxoid changes and spindle cells with dendritic cytoplasmic processes (7–9). As first described by Suster *et al* in 1998, DFML(DentriticFibromyxolipoma) was distinguished from SCL by the presence of dendritic cytoplasmic processes, a plexiform vascular pattern and abundant keloidal collagen (1). With further study, these features of DFML have also been observed in certain cases of SCL. Karim *et al* (2) speculated that DFML possibly represented an unusual variant of myxoid SCL, due to the similarities in their clinical and pathological features. By contrast, Tan and Wen (3) inferred that DFML occurred on a morphologic continuum and represented a transitional form between spindle cell lipoma and SFT (solitary fibrous tumor), and SFT was on the end of the spectrum. DFML represents a transitional form between spindle cell lipoma and SFT, and the latter is at the end stage of the transition.

From literature, the age of the patients ranged between 24 and 81 years, and the median age at diagnosis was 66.5 years. Among the cases, there was a higher prevalence of males. The majority of tumors were located in the superficial soft tissue of the head and neck region, chest, back, shoulder and lower leg. The masses ranged in size between 2 and 24 cm. The tumors were characterized by a proliferation of small spindle or stellate cells, prominent abundant myxoid stroma with ropey collagen bundles, and an admixture of mature adipose tissues, which is similar to the morphology of SCL.

In clinical practice, DFML has abundant myxoid stroma and a prominent plexiform vascular pattern reminiscent of that observed in myxoidliposarcoma, therefore, the tumor may be mistaken for a myxoidliposarcoma, which may include myxofibrosarcoma, low-grade fibromyxoid sarcoma and myxoid synovial sarcoma. Suster et al described 12 cases of DFML, of which three were initially misdiagnosed as myxoidliposarcoma and one as myxoid malignant fibrous histiocytoma (1). The case reported by Karim et al was initially diagnosed as low-grade liposarcoma on an incisional biopsy (2). The present case was also mistaken for myxoidliposarcoma at the preliminary diagnosis. Myxoidliposarcoma often occurs between the ages of 45-60 years, and ~75% of cases develop in the deep muscles of the lower extremities. Welldifferentiated myxoidliposarcoma in such locations requires careful consideration. Immunohistochemically, the tumor cells of the overwhelming majority cases stain for S-100 protein, while CD34 staining is negative. In addition, >90% of myxoidliposarcomas present with a reciprocal translocation between chromosomes 12 and 16: t(12;16)(q13; p11) (13,14). The study by Zhang et al (15) showed that molecular testing was useful for all deep-seated and larger lipomatoustumors (>15 cm). Myxofibrosarcoma is generally characterized by a significant degree of nuclear pleomorphism and occasional mitotic figures. The particular features are vacuoles of these pseudolipoblasts and a vascular pattern characterized by large curvilinear vessels. Low-grade fibromyxoid sarcoma exhibits myxoid areas and cytologically bland spindle cells. The spindle cells are characterized by whorled or swirling growth patterns. Myxoid chondrosarcomas are composed of small, distinctly eosinophilic cells typically arranged in small clusters, cords or pseudoacini, unlike the single cell arrangement in pure myxoidliposarcomas (13,16). Overall, as a result, the distinction between the tumors is occasionally difficult. It is therefore important to conduct long-term follow-up in order to remain alert to the correct diagnosis.

The differential diagnosis for DFML includes other myxoid spindle cell neoplasms of the soft tissue, including superficial angiomyxoma, superficial acralfibromyxoma and myxoidperineuroma (1-6,8). When a myxoid spindle cell tumor is diagnosed, DFML should be considered; it is important to avoid misdiagnosis of more aggressive neoplasms.

III. Conclusion

In summary, the present study reported a case of an unusual fibromyxolipoma on palate. This case was unusual in its location and size, and was diagnosed based on the typical morphological criteria of DFML and the exclusion of malignant features. Further studies with more cases will be required to determine whether the tumor belongs to a myxoid variant of SCLs, an unusual form of myxoid SFT or an intermediate type between the two.

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Dr.V.P.Selvakumar, et. al. "Fibromyxolipoma: A Palatal Mass." *IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*, 20(09), 2021, pp. 43-46.