Case Report

Low-grade fibromyxoid sarcoma: A rare case report

ABSTRACT

Low-grade fibromyxoid sarcoma (LGFMS) is a rare type of sarcoma that is characterized by benign-appearing histologic features but a paradoxically aggressive clinical course. Recognition of this lesion is important because of its indolent but metastasizing nature. These tumors generally occur in young to middle-aged adults, sometimes in children, but rarely in the high-aged adults. LGFMS typically affects the deep soft tissues of the trunk or lower extremities: however, it is rarely seen on the maxillofacial region. Here, we describe a case of LGFMS on the left lower one-third region of the face of a 35-year-old male patient with a 6-month history. On gross examination, the resected specimen consisted of an open ovoid mass of 2 cm × 2 cm × 1 cm. Light microscopy revealed well-circumscribed myxoid tumor with hypocellular areas in nodules merging to collagenized areas. Immunohistochemical examination revealed diffuse positivity to vimentin, whereas tests for desmin, S-100 protein were negative, thus confirming the diagnosis. After the initial healing of the surgical wound, the patient was advised 30 cycles of radiotherapy. Recurrence and metastasis have not been observed for 1 year of surgical excision now. Due to the notably indolent nature of LGFMS, long-term follow-up is necessary to evaluate its clinical course

Keywords: Low-grade fibromyxoid sarcoma, myxoid tumor, maxillofacial region, wide excision, recurrence

INTRODUCTION

Soft-tissue sarcoma (STS) encompasses a broad array of malignant tumors that arise in the mesenchymal soft tissues at any anatomical site. They are rare tumors, accounting for <1% of all malignancies originating in the cavity. STSs mainly arise in the extremities and trunk and only 5%–20% of STS arise in the head and neck region. STS have a varied cell of origin but are grouped together because of their similarities in clinical presentation, natural history, treatment, and outcome.[1]

Low-grade fibromyxoid sarcoma (LGFMS) is considered as a distinctive entity of fibrosarcoma with a high potential of metastasis despite the benign histologic appearance; sometimes, there is a long interval between tumor presentation and metastasis. It was first described by Evans in 1987 and was initially characterized as a spindle cell tumor with bland histologic features but paradoxically aggressive behavior, with a high rate of recurrence and metastasis.[2,3]

Its exact incidence is not well-known. Subsequent studies along with the World Health Organization classification of STTs have confirmed this relatively uncommon tumor as a discrete subtype of a fibrosarcoma. Lately, studies on genetic analysis have unraveled a characteristic underlying recurrent, balanced translocation t (7; 16) (q32-34; p11), resulting in the formation of FUS-CREB3 L2 and FUS-CREB3 L1 fusion genes in this sarcoma.[3-5] Lane *et al*.[3] identified a hyalinizing spindle cell tumor with giant rosettes as a variant of LGFMS.

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LGFMS typically presents as deep, intramuscular, soft-tissue masses of the proximal extremities and trunk.[4,5] It can be found rarely in the retroperitoneum, head, or the chest wall. The majority of cases occur in subfascial deep locations and affect typically young or middle-aged adults; but it has also been described in the pediatric population, where it tends to be smaller, superficial, and easier to resect.[4-6]

LGFMS in the oral and maxillofacial region is very rare. The literature on oral STS itself is scarce; the majority of it is in the form of isolated case reports. The aim of this paper is to report a 35-year-old male patient with LGFMS at a rare location in the maxillofacial region and to review the literature for LGFMS.

CASE REPORT

A 35-year-old male patient presented with a slow-growing painless mass on the left lower one-third region of the face. The mass was noted 6 months before the first visit. The tumor mass was hard in consistency, immobile, was fixed to the underlying tissue, approximately 3 cm × 2 cm in size, and located at the left lower border of the mandible at the body region [Figure 1]. There was no history of trauma; the mass was not tender and was deep with no skin changes. No enlarged local lymph nodes were noted and also no abdominal organ or mass was palpable. Systemic examination was unremarkable.

Routine blood investigations were normal. Chest X-ray was normal. Aspiration biopsy showed features suggestive of granulomatous inflammation. A high-resolution 12 MHz transducer ultrasound scan showed an ill-defined hyperechoic mass lesion in the left mandibular region abutting the body of the mandible. Computed tomography scan showed a well-defined encapsulated fusiform soft-tissue density mass of HU 20–30 measuring 3.0 cm × 1.7 cm × 2.8 cm noted adjacent to the left side of the body of the mandible with minimal appreciable enhancement causing scalloping of adjacent bone with thinning of the cortex. Trucut biopsy of the tumor mass suggested a diagnosis of possible benign nerve sheath tumor. The clinical and radiological impressions were those of possible intramuscular myxoma, myxoid liposarcoma, or a benign nerve sheath tumor. We performed an excisional biopsy to confirm the diagnosis [Figure 2]. A complete excision of the mass with its extension was done and at surgery, about 2 cm × 2 cm lobulated tumor mass, intramuscular and adherent to muscle fibers and underlying bone, was evident [Figure 3]. The lump was excised along with the attached muscle bundles of fibers and the wound was irrigated and closed in layers [Figure 4].

On gross examination, the resected specimen consisted of an open ovoid mass of 2 cm × 2 × cm 1 cm. Light microscopy revealed well-circumscribed myxoid tumor with hypocellular areas in nodules merging to collagenized areas [Figure 5]. Tongues of myxoid areas were insinuating between muscle and fascial planes. Bland spindle cells with no atypia were seen in short fascicles and whorls in the myxoid zone. The tumor cells contained oval or short spindle-shaped nuclei without a degree of atypism or pleomorphism. Based on the histologic findings, including the absence of nuclear atypism or pleomorphism, and the presence of predominant fibrous components, the diagnosis was given as LGFMS [Figures 6 and 7]. Immunohistochemical examination revealed diffuse positivity to vimentin, whereas tests for desmin, S-100 protein were negative, thus confirming the diagnosis. After the initial healing of the surgical wound, the patient was advised 30 cycles of radiotherapy.

DISCUSSION

LGFMS is a rare tumor type first described by Evans in 1987. In the original description, LGFMS was histologically

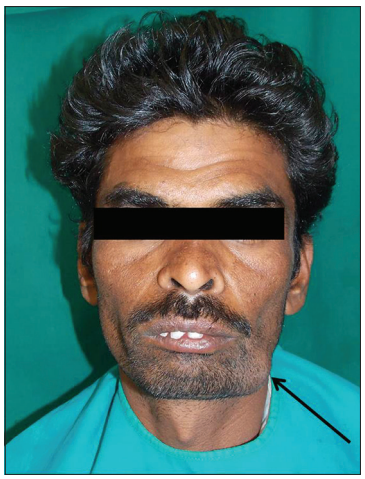

Figure 1: Preoperative profile photograph

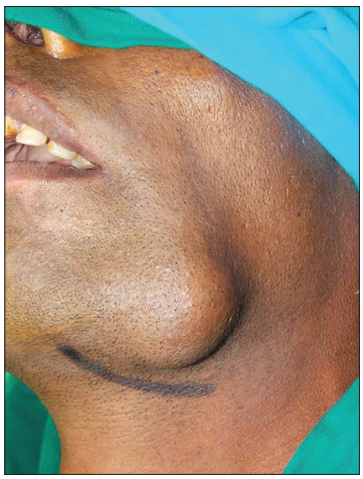

Figure 2: Extraoral incision

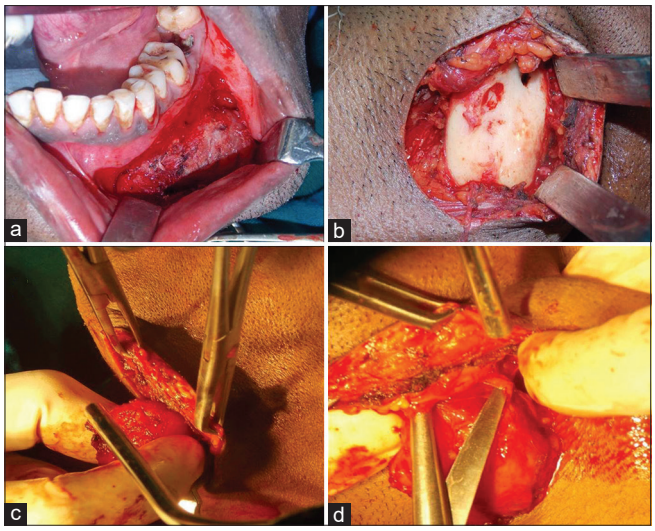

Figure 3: Intraoperative dissection

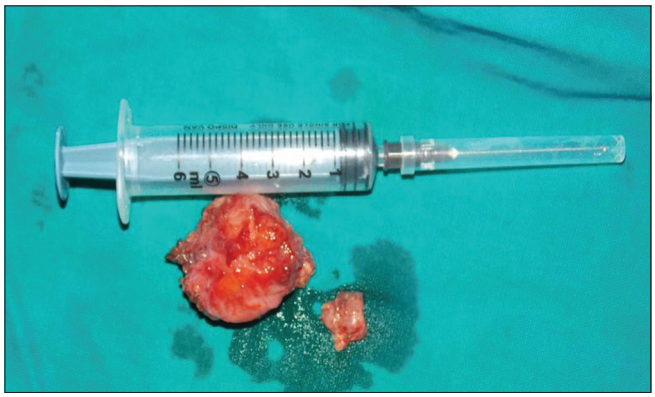

Figure 5: Excised lesion

characterized as a bland appearing soft-tissue neoplasm, but the tumor was found to be associated with aggressive behavior and a high degree of local recurrence or distant metastasis.[4,5] In a report by Folpe *et al*,[7] local recurrence and distant metastasis were seen to be 9% and 3%, respectively. The authors highlighted that early surgery was one of the reasons why the rates of local recurrence and distant metastasis in their study were lower than other reports.[7,8]

LGFMS is a variant of fibrosarcoma with distinctive histopathological features. Immunohistochemical staining (IHC) is usually positive for vimentin, while it is negative for a variety of antibodies such as desmin, keratin, S100 protein, epithelial membrane antigen, CD34, and CD316.[2]

The most frequently reported location of LGFMS is the lower extremity, especially the thigh, followed by the trunk and groin, upper extremity, and buttock. The head and neck region is a very rare location for this sarcoma, as based on previous reports.[1-5] Hence, this case of LGFMS in the

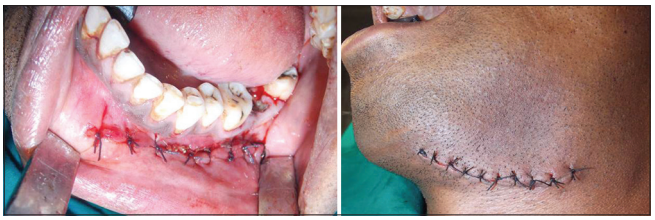

Figure 4: Closure

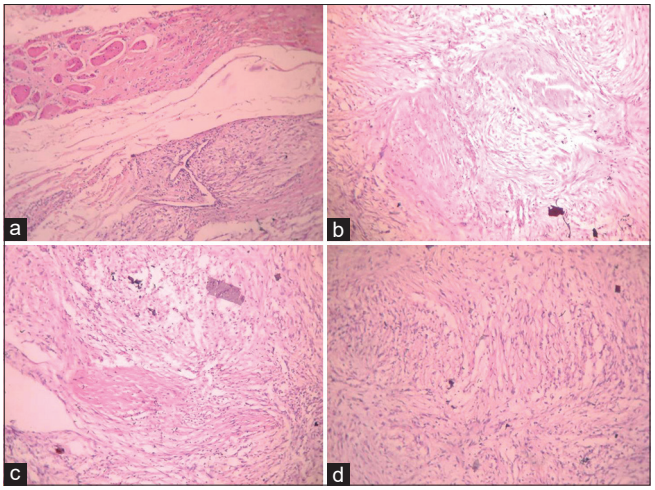

Figure 6: Photomicrograph of excisional biopsy. (a) Atrophic muscle and tumor interface. (b) Cellular areas and myxoid stroma. (c) Collagenous areas and cellular areas. (d) Cellular areas

maxillofacial region (just above the lower border of the mandible) is a very rare reporting of its kind.

Histologically, LGFMS, tumor cells contained oval or short spindle-shaped nuclei without a degree of atypism or pleomorphism. Evans also has mentioned that some recurrent and metastatic tumors contain zones of increased cellularity and mitotic activity. [3,5,7-9]

The differential diagnosis of LGFMS includes myxomas, angiomyxomas myxoid liposarcoma, or those with mixed myxoid and fibrous elements, such as neurofibroma, malignant peripheral sheath tumor, and fibrous histiocytoma which can be differentiated by IHC.[2] Typical malignant fibrous histiocytoma is considerably more cellular and has greater malignant potential than LGFMS that is it shows greater nuclear hyperchromatism, pleomorphism, and mitotic activity.[5] Myxofibrosarcomas are classified into four grades, with grade 1 myxofibrosarcoma considered to resemble LGFMS. However, this tumor can be differentiated from LGFMS by the greater degree of atypia, predominant myxoid component, and absence of metastasis; further myxofibrosarcoma generally occurs in older adults.[4,5]

Immunohistochemically, most cells of this sarcoma are strongly positive for vimentin antibody but are generally

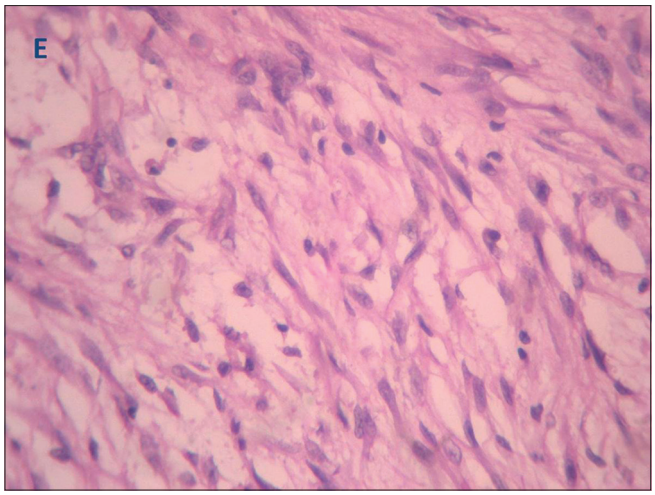

Figure 7: High-power electron microscopy showing spindle cells

negative for α-smooth muscle actin, desmin, S-100 protein, cytokeratin, CD34, and CD56 antibodies.[5,6] In our case, there was diffuse positivity to vimentin, whereas tests for desmin, S-100 protein were negative. In addition to these features, Zamecnik and Micheal described the presence of strong and diffuse bcl-2 reactivity, although we did not investigate bcl-2 reactivity in our case.

A similar entity is characterized by giant rosettes and the presence of areas of hyalinized acellular islands surrounded by oval and spindle cells has been named as "Hyalinizing Spindle Cell Tumor with Giant Rosettes" (HSTGR).[7] Reports have suggested that both entities are the same and reflect the neoplastic process.[7] Both entities shared the same pathologic mechanism: a specific identification of cytogenetics in the form of a balanced t (7;16) (q34;p11) translocation and fusion between FUS and CREB3 L2 genes in both LGFMS and HSTGR were confirmed.[2,7,8] Although we did not attempt to analyze these fusion genes, these methods may be valuable tools in the differential diagnosis.[7]

The HSTGR variant of LGFMS might be mistaken for a neuroblastoma-like schwannoma. This variant of schwannoma usually has areas of more typical appearing schwannoma with characteristic Antoni A and Antoni B areas and thick-walled hyalinized vessels rather than the branching to curvilinear vessels seen in LGFMS. Negativity for S-100 protein in LGFMS militates against the diagnosis of schwannoma.[3]

Accurate diagnosis is paramount to the management of any malignant tumor. In the series reported by Evans in 1993, deep LGFMS was associated with a significant risk of local recurrence (68%) as well as distant metastasis (41%).[3] However, LGFMS may not be as aggressive as originally thought. The original series describing LGFMS was based on a retrospective analysis of cases, the majority of which were also originally diagnosed as benign lesions. In a large series of cases ascertained prospectively, the rates of recurrence and metastasis were 10% and 6%, respectively. Adequate surgical excision of the tumor is necessary because of the frequent recurrence of LGFMS.[3]

The presence of focal areas of high cellularity, nuclear enlargement, increased mitotic activity, and necrosis is not considered of poor prognostic significance for recurrence or metastases.[2] The utilization of external beam radiation therapy (XRT) is recommended as an alternative for re-resection if the latter is difficult. XRT is likely to reduce the chances of local recurrences, but it will not affect overall survival.[2,3,9] Adjuvant radiotherapy or chemotherapy has not been recommended in previous reports; however, these treatment strategies may be used in cases of multiple metastasis or frequent recurrence.[5]

CONCLUSION

LGFMS is a rare type of STS which affects primarily young- and middle-aged adults. It is a form of fibrosarcoma which demonstrates a range of histologic appearances beyond the typical pattern; however, with the exception of dedifferentiation, these differences do not appear to relate to tumor behavior or patient survival. However, it still has a distinctive presentation and histological features, immunohistochemistry results, and specific cytogenetic changes.

LGFMS typically presents in the proximal extremities and trunk and is found rarely in the retroperitoneum, head, or the chest wall. This case we are reporting is perhaps the first reported case in the maxillofacial region. Surgical management is the standard therapy and it has a high recurrence and metastatic potential. Small tumor size may be a favorable prognostic factor. Local radiotherapy needs to be advocated for the prevention of recurrence and metastasis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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