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Case Report

Rare case of low-grade fibro-myxoid sarcoma presenting as a lateral neck swelling

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ABSTRACT

Low-grade fibromyxoid sarcoma (LGFMS) is an uncommon malignancy. LGFMS is more likely to affect the trunk and extremities in young adults. However, sporadic cases have been reported in many areas of the head and neck region, including the oral cavity, larynx, and oropharynx. LGFMS typically shows areas of collagenized, myxoid stroma with spindle cell appearance in a rolling pattern. Accurate diagnosis is challenging because it can be mistaken for a simple neoplastic entity of round cells. A few cases of LGFMS which have been reported in the head and neck showing that LGFMS is characterized by local recurrence and metastases. Because of the rarity of the head and neck region, there are no specific treatment guidelines or follow-up. Our study presents LGFMS in a 54-year-old patient in the neck region who underwent surgery and was followed up for one year.

Keywords: Low grade fibro-myxoid sarcoma, Neck, Spindle cell neoplasm, Vimentin positive

INTRODUCTION

Low-grade fibro myxoid sarcoma grade 1 (LGFMS) is a unique type of fibrosarcoma with high metastatic potential. In rare cases, LGFMS has a long interval between tumor presentation and metastasis.[1] Although it is a well-defined entity, it is believed that many cases are not recognized as LGFMS, so it is still difficult to accurately diagnose the disease. These tumors usually arise in the adjacent organs and trunk.[2] In some cases, they are found in different locations such as retroperitoneum, head/neck, or chest wall.[2,3] Most LGFMS occurs in a small area. In rare cases, the subcutaneous tissue or skin may be affected. LGFMS is also known as Evans tumor who first described it.[4,5] The other probable of LGFMS includes lesions that show the growth of round cells in a myxoid pattern with or without a granular component.[4,5] The classification of lipomatous tumors has increased, and there is more information about tumors that are similar in appearance and presentation, because of the increased risk of diagnosis. Spindle cell lesions need further pathological differentiation from liposarcoma, other spindle cell neoplasms, and myxoid lesions for clinical diagnostic purposes. Patients all cytology, histology and cytogenetics, along with clinical manifestations, are very important in achieving the correct diagnosis of spindle cell lipoma. We present a case report with characteristics of SCL and a review to further clarify the diagnosis and surgical management of this soft tissue tumor.

CASE REPORT

A 58-year-old Hindu married male patient, resident of Kutch district, Gujarat working as food vendor coming from lower socioeconomic class came with a large right sided lateral neck swelling for 8 years. The swelling was insidious in onset and gradually progressive in nature which increased slowly initially in 5-6 years and then increased progressively over 3 years. The swelling was

painless associated with difficulty in neck movement (Figure 1). Swelling was not associated with pain or difficulty in swallowing or difficulty in breathing and not associated with hot and cold intolerance.

He requested treatment as this gradually enlarging mass was embarrassing on sight and increased in size rapidly over past one year and was causing difficulty with sleep. He had no past history of other soft-tissue tumors and further endocrine workup was within normal limits. He had addiction of tobacco chewing for last 25 years.

On clinical examination, a solitary globular swelling of around 15×10 cm noted with smooth surface over right lateral neck region with well-defined edges and skin over swelling was normal non-erythematous, non-oedematous without abnormal pulsations. Swelling didn't move with deglutition and also protrusion of tongue and respiration even. On palpation after confirming inspection findings, a single solitary swelling of size 16×12×4 centimeters was present over right lateral neck region with smooth surface. Local temperature overlying it was normal without local tenderness. The swelling was extending anteriorly up to lower sternocleidomastoid muscle's posterior border and posteriorly extending the posterior triangle of neck. Swelling was well-defined, without fluctuation and trans-illumination. Swelling was non-compressible and non-reducible in nature. Skin overlying is normal. Consistency was firm and doesn't appear to be fixed to underlying structures. On auscultation, no bruit no murmur and no abnormal sounds heard.

On FNAC, myxoid spindle cell neoplasm of right neck swelling was seen. CECT scan done suggestive of a large exophytic lobulated heterogeneously enhancing soft tissue density in postero-lateral aspect of neck on right side with characteristic extension to surrounding structures consistent with spindle cell neoplasm without evidence of lymphadenopathy (Figure 2).

Differential diagnosis on clinico-radiological correlation was lipoma, ossifying fibroma, non-ossifying fibroma, nodular fasciitis, inflammatory myo-fibroblastic tumor, angiosarcoma, leiomyosarcoma, kaposi sarcoma, giant cell fibroma and low-grade fibro-myxoid sarcoma.

Surgery was performed and the removed mass was sent for histopathological analysis. The surgical specimen post operation was sent to the pathology laboratory of our hospital. The gross size of the tumor is 16 x 12 x 4 cm and weighs 1.8 kg and it is partially covered by the regional muscular system. The surface of tumor belly is smooth and shiny and light brown in color. In the cut section, a small area is found that is full of hemorrhagic and seromucous fluid. The cut surface of the lateral solid area revealed a whitish-yellow tissue with a gelatinous focal consistency (Figure 3).

Microscopic evaluation of the mass revealed a LGFMS tumor with features of a moderate, spherical tumor composed of round cells with mild atypia and features of fibroblasts embedded in the collagenous stroma. Tumor cells showed eosinophilic cytoplasm with vesicular nuclei and small nuclei, arranged in a plexiform or annular pattern. Extracellular mucin is labeled. Mitosis was not observed (Figure 4).

On IHC evaluation, tumor cells showed to be positive for MUC4, Vimentin (marked diffuse positivity) and BCL2 (scant positivity) while the tumor was negative for CD99 (Figure 5).

The characteristic presence of slow growing, low-grade fibro-myxoid tumor along with the strong and positivity for MUC4 IHC markers confirmed the diagnosis. The patient was followed up to one year with no signs or symptoms of local or distal metastasis.

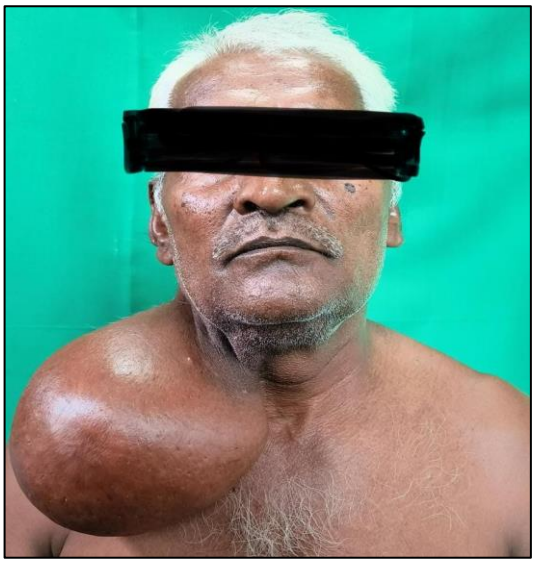

Figure 1: Clinical photograph of patient taken with consent showing the mass in right lateral neck.

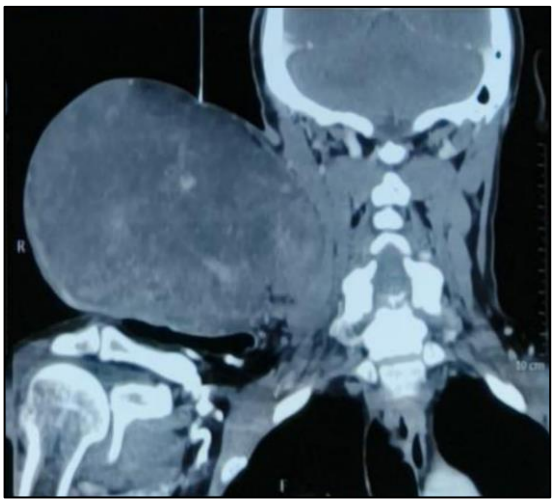

Figure 2: CECT image of the tumor showing large exophytic growth over postero-lateral aspect of right side of neck.

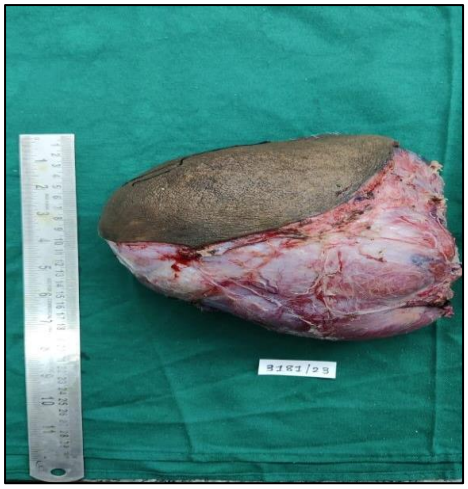

Figure 3: Tumor surgical resection done showing 1.8 kg weight and measuring specimen 16×12×4 centimetres.

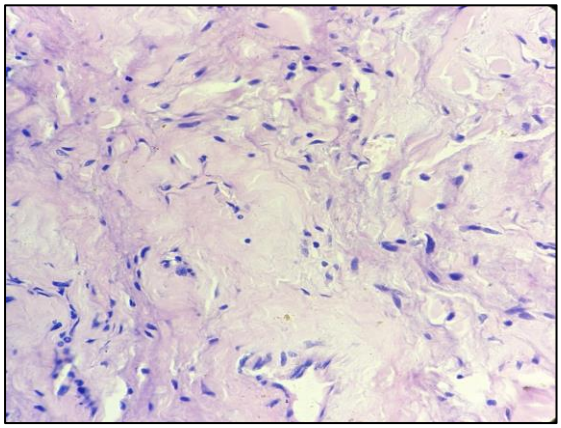

Figure 4: Microphotograph showing moderately cellular, low-grade spindle cell neoplasm with mild nuclear atypia in fibroblasts with collagenous stroma (H&E, X 100).

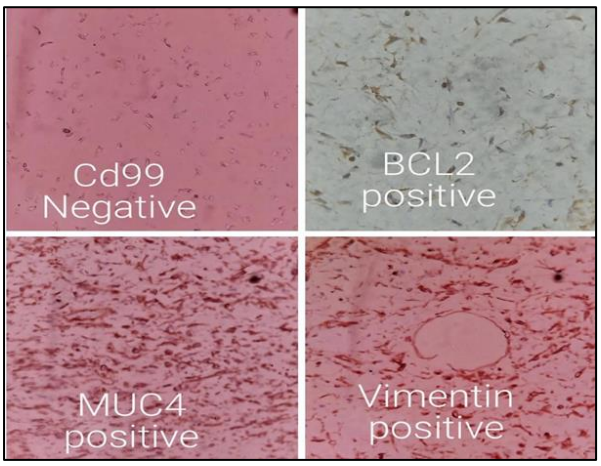

Figure 5: IHC images of LGFMS showing MUC4, BCL2, Vimentin positive (IHC-DAB, x100).

DISCUSSION

Because of its rarity, little has been published about the treatment and outcomes of LGFMS. This creates a significant problem with a lack of evidence to guide treatment in pre/metastatic conditions. LGFMS was first described by Evans in 1987 with a benign history and a pattern of recurrence and metastases.[5] In Evans' first report, 7 of 12 LGFMS patients developed malignancy showing distant metastases, with a follow-up of 5.5 to 50 years.[5]

However, in 54 LGFMS patients studied by Folpe et al, only 5 (9%) had local recurrence and 3 (6%) had distant metastases, because most patients were treated with invasive or short-term surgery following period, averaging of 24 months.[6] LGFMS mostly seen in young and middle-aged adults. It usually involves the deep soft tissues of the lower extremity like thigh leg, buttocks, followed by the chest wall, shoulder, neck and other areas. The most common site for metastasis was seen to be lung. Tumors shows mixture of myxoid and fibrous morphology include neurofibroma, fibromatosis, perineurioma, squamous cell carcinoma and fibrous histiocytoma. Other entities at risk include desmoid tumor, desmoplastic fibrosarcoma, and low-grade liposarcoma. It is histologically characterized by alternating myxoid and fibrous areas, also shows bland fusiform cells with a whorled growth pattern.[7] Other variations include giant rosettes, areas of hypercellularity, rounded epithelioid foci cells, less commonly observed are focal osseous metaplasia and significant nuclear pleomorphism.[8]

In the past, myxoid areas are characterized by tissue areas, small round-shaped cells and a circular growth pattern. Focal bone metaplasia and nuclear pleomorphism are rare. In addition, features of focal sclerosing epithelioid fibrosarcoma have been reported in LGFMS However, correlation of histological changes and the biological behaviour of the tumor has not been reported.[9] In our study, focal cells with less stroma and higher atypia were observed in the newly resected tumor, which may be related to the growth rate and aggressive behaviour.

LGFMS consists of hypercellular and hypocellular areas of tumor cells in a background of collagenous, myxo-collagenous or myxoid stroma. There is transition between the collagen and the myxoid region. The lesional cells are smooth, have small nuclei which are angulated, have scanty cytoplasm and have very little nuclear atypia, and are arranged in circular growth patterns. Mitotic figures are negligible or absent. Curvilinear blood vessels, seen are long, sinusoidal vessels with collapsed lumens, are seen with perivascular sclerosis, especially in myxoid areas. The histopathological features of LGFMS are usually classic. In addition, several cases showed other morphological changes described in the literature, including a loose storiform pattern, pericellular spaces

with more rounded cells and increased activity. mitotic, multinucleated giant cells, and bone metaplasia. Immunohistochemical staining shows vimentin protein to be positive and negative with various antibodies such as desmin, keratin, S100 protein, epithelial membrane antigen, CD34 and CD31.In the wall of small vessels in the stomach and very fine in the fibrous tissue layer muscle specific action is also found.

Most LGFMS seem to be well maintained but no caps. Local infiltration may occur, and resection is usually incomplete.[10] Therefore, the preferred treatment for LGFMS is wide resection of tumor with margins and radical surgery.[2,11] LGFMS is also reported a case in larynx and was surgical resected via transoral route Immunohistochemical analysis plays a crucial role in confirming the diagnosis of LGFMS, where most tumor cells were strongly positive for vimentin.[12] In this case, the immunohistochemical profile supported the diagnosis, ensuring accurate identification of this rare tumor in the laryngopharynx. All cases diagnosed for LGFMS were done further workup with genetic studies as it showed variation and translocation at specific genome but in our study though patient advised for further genetic makeup and PETCT for evaluating locoregional spread post-surgical excision patient being from lower socioeconomic site and inadequate funding patient denied for the same and agreed for regular follow-up for the same.

CONCLUSION

LGFMS is a type of low-grade sarcoma that rarely occurs in the head and neck. Because of its rarity, the pathologist may not consider LGFMS in the differential diagnosis of head and neck neoplasms. Understanding LGFMS and its counterparts should be considered in the differential diagnosis of head and neck tumors, with MUC4 immunohistochemical staining as an IHC marker to confirm the diagnosis. This case report shows and enriches the literature with information on the imaging diagnosis and surgical treatment of this rare tumor. Because there is no specific protocol for follow-up examinations, and early detection of metastasis, it is important to inform patients about the long- term prognosis of metastatic disease.

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