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Case Report

Low grade myofibroblastic sarcoma in paraspinal location: A case report with review of literature

Devmalya Banerjee[1], **Shreya Bhattacharya**[2], **Sourav Sarkar**[1], **Nilay Biswas**[3], **Nirmalya Banerjee**[1,*]

[1]*Dept. of Oncopathology, Narayana Superspeciality Hospital, Kolkata, West Bengal, India*
[2]*Dept. of Head & Neck Surgery, Narayana Superspeciality Hospital, Kolkata, West Bengal, India*
[3]*Dept. of Neurosurgery, Narayana Superspeciality Hospital, Kolkata, West Bengal, India*

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ABSTRACT

Myofibroblasts were first detected in the granulation tissue and low-grade myofibroblastic sarcoma has now been recognized as a distinct entity. It has an indolent clinical course and distant metastasis is very rare. Hence surgical excision is the mainstay of the treatment. However, this tumor morphologically mimics other relatively high-grade soft tissue sarcomas, so distinguishing them is of utmost importance. Here we report, a 22-year-old young girl presented with neck pain and a slowly growing mass in the cervical paraspinal region. The patient was managed by wide local excision and the specimen was examined thoroughly. After considering all the features diagnosis of low-grade myofibroblastic sarcoma was offered.

1. Introduction

Low grade myofibroblastic sarcoma (LGMS) is a relatively uncommon sarcoma of low malignant potential.[1] It mostly arises from the subcutaneous, submucosal, deep soft tissue as well as the intraosseous region. Head, neck region is the commonly affected site.[2,3] Myofibroblasts are basically derived from native fibroblasts in response to tissue injury. Therefore myofibroblasts take leading role in wound healing via extracellular matrix synthesis.[4] Low grade sarcoma of myofibroblastic lineage is known for its indolent clinical behavior and wide excision is mostly preferred along with long term follow up. However incidences of local recurrences are more frequent than distant metastasis.[5] LGMS was first enlisted in the WHO bone and soft tissue blue book in the year 2002 and is still referred as a distinguished entity in the fibroblastic/ myofibroblastic tumor category.[6]Here we report a 22 year old young girl who presented with neck pain and a slowly growing mass in the cervical paraspinal region. She was evaluated thoroughly and core biopsy was performed initially. However later on she underwent wide excision procedure. The specimen was examined and the diagnosis of low grade myofibroblastic sarcoma was established after ruling out all the relevant differentials. The diagnostic approach, immunohistochemical features, therapeutic modalities and prognosis have been discussed here.

Table 1: Results of IHC

Positive reaction	Desmin	Calponin	SMA		
Negative reaction	H-caldesmon	MyoD1	STAT6	ERG	MUC4

* *Corresponding author*.
E-mail address: nirmalyapgi@gmail.com (N. Banerjee).

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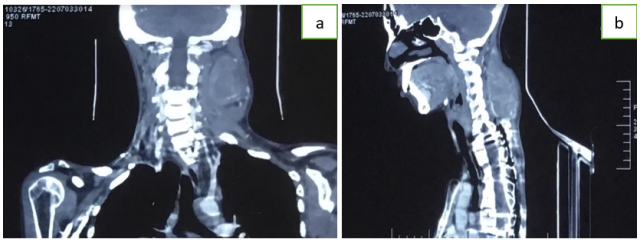

Fig. 1: Radiology images of the tumor; **A:** Coronal view of the tumor; well circumscribed paraspinal tumor mass across the whole length of left paraspinal soft tissue; **B:** Sagittal view of the tumor; Tumor extends from skull base to D1 vertebral level without any bony erosion or intrathoracic extension.

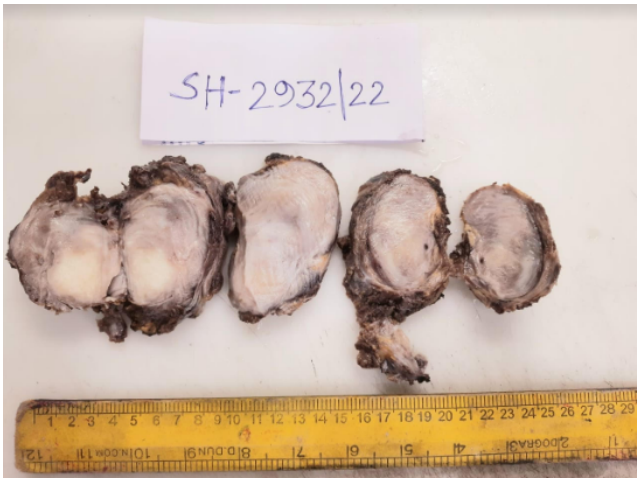

Fig. 2: Gross image of the tumor; **A:** Well circumscribed, firm to hard mass with homogenous, whitish cut surface without any evidence of necrosis and hemorrhage.

2. Case Report

A 22 year old girl came in the Surgical outpatient department with chief complain of slowly progressive, long standing swelling in the back of neck with recent onset pain the neck region. She is a student by profession and she provided past history of scoliosis for which she underwent corrective surgical intervention almost 12 years before. She was examined clinically on that particular day and no evidence of compressive neuropathy has been detected. A multidisciplinary medical team had been created and the members reached to a consensus opinion of taking targeted core biopsy from the lesion. She was referred for magnetic resonance imaging (MRI) scan. However it could not be performed owing to spinal metallic implant which was non-compliant to the MRI scan. Therefore contrast enhanced computed tomography scan was done to evaluate the paraspinal mass. A well circumscribed heterogeneously enhancing soft tissue mass was noted extending from the skull base to the D1 vertebral level, occupying and infiltrating the paraspinal soft tissue. The

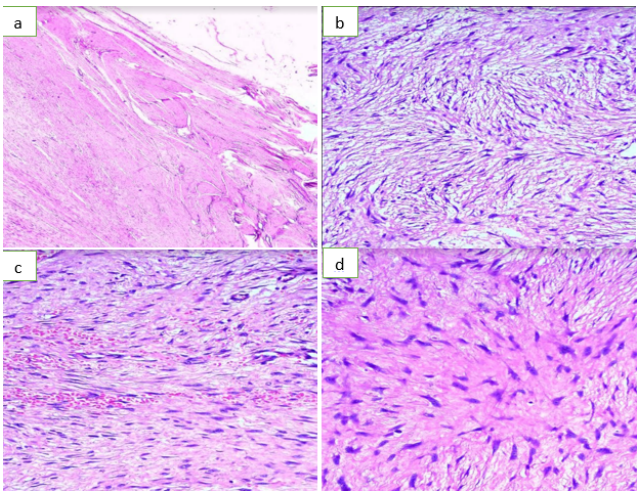

Fig. 3: Photomicrography of the tumor.**A:** Tumor is poorly circumscribed and shows infiltrating margin (H&E, 4X); **B:** Tumor cells are relatively monomorphic, elongated and spindle shaped with loose myxoid stroma in the background (H&E, 10X); **C:** Interstitial space shows foci of microhemorrhage (H&E, 10X); **D:** Mild nuclear pleomorphism is seen with pink, homogenous collagen rich background (H&E, 20X)

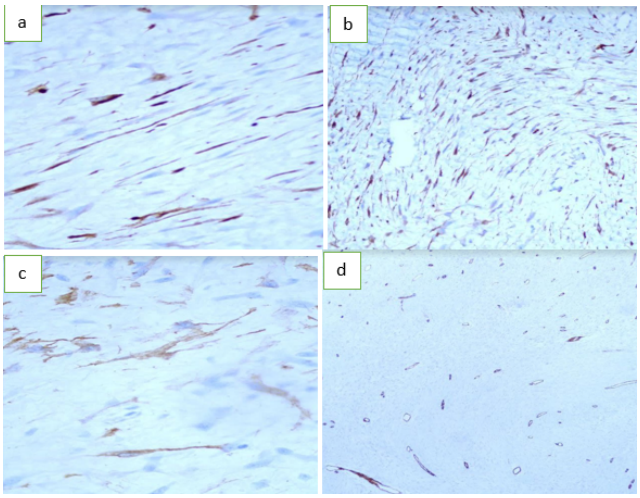

Fig. 4: Photomicrography of the immunohistochemistry. **A:** Tumor cells show cytoplasmic reactivity to desmin (H&E, 40X); **B:** Tumor cells show cytoplasmic reactivity to calponin (H&E, 10X); **C:** Tumor cells show variable cytoplasmic reactivity to smooth muscle actin (H&E, 40X); **D:** None of the tumor cell is immunoreactive to CD34 immunostain (H&E, 4X)

mass approximately measures 7.5x5 cm in the craniocaudal and transverse direction. The tumor was predominantly nourished by the vertebral artery. There was no vascular encasement, bony erosion or intrathoracic extension noted (Figure 1A,B). She was motivated for wide local excision which was the only treatment option available to the surgical team.

After the surgical excision the entire specimen was sent to the department of oncopathology where it was sliced and examined macroscopically. It was 7.5x5 cm firm to hard mass with ill-defined margin. The cut surface appeared homogenous, whitish and areas of trabeculation had been identified. No areas of necrosis and hemorrhage was identified (Figure 2). Histomorphological examinations showed cellular spindle cell tumor with infiltrating margin. Tumor cells are elongated, spindle shaped having wavy nuclei with variable nuclear atypia. Background contains myxoid material along with branched capillary network. Focally collagenous background was identified (Figure 3A-D). Mitotic activity was imperceptible (1-2/10 high power field). On immunohistochemistry, these cells were focal but distinct positivity for the desmin (Figure 4A) and diffuse cytoplasmic reactivity for calponin (Figure 4B) was also identified. The immunostain for smooth muscle actin (SMA) (Figure 4C) showed typical tram track appearance. There was no reactivity for H-caldesmon, Myo-D1, ERG, STAT-6, MUC4 and CD34 (Figure 4D). Beta-catenin immunostain showed very focal and faint cytoplasmic reactivity (Table 1). The proliferative index was low. Therefore in view of histomorphological and immunohistochemical features, diagnosis of the low grade sarcoma of myofibroblastic origin was preferred in this case. The patient was discharged on the seventh post-operative day and was kept in close follow up. She is doing well until now with no obvious physical morbidity.

3. Discussion

Myofibroblastic tumor cells are contractible, spindle to fusiform shaped specialized fibroblasts having contractile elements within their cytosol. It was first identified by the Mentzel et al.[7] who also reported 18 cases of LGMS in the year of 1998.[7,8] They are primarily associated with the inflammatory and fibrosing conditions. In adults, they exist around the testicular seminiferous tubules.[9,10] According to the Gabbani et al.[11] transforming growth factor-beta, fibronectin and mechanical tension play in consortium for transition of myofibroblast from fibroblast.[11] The myofilaments are distributed predominantly in the subplasmalemmal layer which is in contrast to the smooth muscle cell where the myofilaments are present throughout the cytosol.[9] Earlier in the literature, LGMS was described under various nomenclature, including myofibrosarcomas, sarcomas of myofibroblasts, myofibroblast-rich fibrosarcomas, leiomyosarcoma myofibroblastic variant and spindle-cell sarcomas showing myofibroblastic differentiation.[12] Mentzel et al.[7] first coined the term LGMS and after that the World Health Organization reclassified this tumor as a distinct entity.[13] According to the published literature most common site is head neck region followed by the lower extremity.[3] According to the Mentzel et al.[7] adult males are most commonly affected and they also mentioned head neck is the commonest site. However according to the Meng GZ et al.[5] femur and trunk are also commonly affected.[5] According to the Wang L et al.[8] bones and soft tissue of the extremities are two most common sites.[8] There is no such documented evidence of LGMS in the cervical paraspinal region in the English published literature. LGMS is a slowly progressive tumor and mostly present as painless mass except those arise from bone which causes painful osteolytic lesion.[8] In the index case it was also a slowly growing paraspinal mass with maximum dimension of 8.5 cm. Wang L et al.[5] conducted a retrospective study and they included 14 cases.[5] According to them the average maximum diameter is 7.8 cm which is almost similar to that of the index case. Mentzel et al.[7] conducted one of the largest study highlighting the clinicopathological features of LGMS and they included 18 cases of the same. They described this tumor as diffusely infiltrative, spindle shaped tumor mainly arranged in fascicles. Background showed collagenous matrix with areas of hyalinization. Almost all the cases (16/18) showed mild nuclear atypia with mean mitotic activity is 2/10 high power field.[7] After extensive morphological evaluation we had noted that this tumor is cellular and comprises of relatively monomorphic spindle cells with myxoid matrix and dilated blood vessels in the background. Fascicular to storiform growth pattern also has been detected. Therefore possibility of deep (paraspinal fibromatosis) fibromatosis also was considered. According to the morphological classification there are five patterns of fibromatosis seen and they are staghorn vessel type, nodular fasciitis like, hypocellular type, myxoid type and hypocellular/hyalinized type.[14] However reactivity to desmin and calponin immunostains are extremely rare in deep fibromatosis.[14,15] Similarly the distinguishing features between LGMS and nodular fasciitis depend on the immunoreactivity to desmin, as calponin can highlights the lesional myofibroblastic cells in both LGMS and nodular fasciitis.[16–18] LGMS should be distinguished from the low grade fibromyxoid sarcoma which is having higher propensity of metastatic potential, in spite of its benign histological appearance.[19] The low grade fibromyxoid sarcoma typically shows alternating fibrous and myxoid stroma. Mucin 4 (MUC4) which is considered as highly sensitive and specific immunostaining marker for low grade fibromyxoid sarcoma was also immunonegative in our case.[19] Even leiomyosarcoma sometimes mimic LGMS on light microscopy. However LGMS typically lacks cigar

shaped nuclei rather they are having elongated nuclei with tapering nuclear ends. The h-caldesmon which is one of the specific marker for smooth muscle differentiation is hardly detected on the LGMS. In the index case there was no reactivity to h-caldesmon rather cytoplasmic reactivity to calponin was noted and characteristic elongated nuclei with pointed nuclear ends was also conspicuous.[20] Hence this case was reported as low grade myofibroblastic sarcoma and after the surgical procedure, patient is doing reasonably well and has resumed her daily activities.

4. Conclusion

LGMS is therefore a low grade sarcoma of myofibroblastic differentiation and wide excision with sufficient margin contributes to favorable prognosis. According to the published literature metastasis and local recurrence are rare. Radiotherapy and chemotherapy have no established role in the therapy and prognosis. In the index case no recurrence have been noticed so far but further observation is required.

5. Authors' Contributions

All authors have directly participated in planning, execution of the study and preparation of the manuscript. All authors proofread and approved the final version for publication.

6. Ethics Statement

The authors retain informed consent signed by the patient's next of kin authorizing the data publication.

7. Source of Funding

None.

8. Conflict of Interest

None.

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Author biography

Devmalya Banerjee, Consultant and Lead Oncopathologist https://orcid.org/0000-0001-9822-3664

Shreya Bhattacharya, Consultant, Head & Neck Surgery https://orcid.org/0000-0002-7085-2044

Sourav Sarkar, Consultant, Oncopathologist https://orcid.org/0000-0001-6952-0133

Nilay Biswas, Consultant, Neurosurgery https://orcid.org/0000-0001-6765-0348

Nirmalya Banerjee, Consultant, Oncopathologist. https://orcid.org/0000-0001-6557-2956

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