Low grade fibromyxoid sarcoma of the mesentery, an under recognised entity: A case report

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ABSTRACT

Low grade fibromyxoid sarcoma (LGFM5) is an indolent, rare soft tissue tumor having potential of late metastasis, but high local recurrence rate despite its low grade histologic findings. The morphologic diagnosis may be challenging, owing to its typically low cellularity, abundant collagen, and relatively bland cytology. We present here a rare case report of a mesenteric mass in an elderly male. The mass was excised. On microscopy, the tumor was composed of sweeping fascicles of spindle cells, which were positive for vimentin and negative for CD117, CD34, smooth muscle antigen and S-100. Herein we report a rare, under recognized case of LGFM5 arising in the mesentery.

KEY WORDS: Low grade fibromyxoid sarcoma, mesentery

INTRODUCTION

Low grade fibromyxoid sarcomas (LGFM5) (Evans’s tumor, hyalinizing spindle cell tumor with giant rosettes) are uncommon low grade malignant deep soft-tissue neoplasms initially described by Evans in 1987, with a deceptive benign histological appearance [1]. With the incidence being 0.18/million, these tumors represent 0.6% of all soft-tissue sarcomas [2]. These fibroblastic tumors typically occur in the lower extremities, particularly the thigh but can occur sporadically in other deep soft-tissues [3]. The other sites include the neck, chest wall, axilla, mediastinum, inguinal region, buttock, and even the brain [4]. Abdominal LGFM5 are extremely rare. They have been reported to arise from anterior abdominal wall, small bowel mesentery, colon, falciform ligament, and retroperitoneum [4-6]. These tumors most commonly occur in young or middle-aged adults. However, occurrence over a wide age range has been noted [3]. Although these are indolent tumors, the potential for late metastasis suggests a need for prolonged follow-up [7,8].

CASE REPORT

A 62-year-old male presented with dull aching abdominal pain since 15 days. There was no history of fever, nausea, vomiting or change in the bowel and bladder habits. On examination, abdominal tenderness was present, a mass was palpable at the hypogastric region which was hard and measured about 6 cm × 8 cm. On investigation, the hemoglobin was 13.7 g/dl and total white blood cells count was 9.2 × 10³/μL. Liver function tests were normal.

Computed tomography scan showed a circumscribed, lobulated homogenously enhancing abdomino-pelvic mass measuring about 11.6 cm × 9.5 cm × 8 cm. Clinically, differential diagnosis of mesenteric desmoid tumor/ileo-cecal gastrointestinal stromal tumor was considered. Hence ileo-colectomy with resection of the tumor was performed and the specimen sent for histopathological examination. Post-operative period was uneventful and sutures were removed on the 11th post-operative day. He was stable on discharge and is lost to follow-up.

HISTOPATHOLOGICAL FINDINGS

The ileo-colectomy specimen with the tumor mass weighed 652 g. Grossly, a well-circumscribed mesenteric mass was identified, which measured 11.5 cm × 9 cm × 5 cm. The cut section revealed homogenous grey-white areas with specks of hemorrhage [Figure 1].

On microscopy, the tumor was composed of ill-defined sweeping fascicles of spindle to stellate cells, which were positive for vimentin and negative for CD117, CD34, smooth muscle antigen and S-100. Herein we report a rare, under recognized case of LGFM5 arising in the mesentery.
stroma [Figure 4], the latter exhibiting focal keloidal texture. Interspersed branching thin walled, stag horn vessels were also noted [Figures 5 and 6]. On immunohistochemistry (IHC), the tumor cells were positive for vimentin [Figure 7] and negative for CD117, CD34, smooth muscle actin and S-100. These findings were consistent with LGFMS.

**DISCUSSION**

Evan in 1987 described LGFMS as deceptively benign-appearing, but metastasizing mesenchymal tumors characterized by contrasting fibrous and myxoid areas, bland regular fibroblastic spindle cells, low to moderate cellularity, and a swirling, whorled growth pattern [1,7]. Subsequently, additional examples of these tumors were described and found to have occurred most often in young adults and had a substantial rate of local recurrence and metastasis, predominantly to the lungs [7].

LGFMS demonstrates a wide range of histologic appearances [7]. The bland fibroblasts are arranged in a whorled or linear pattern and alternating hypo cellular areas with a myxoid stroma. The cells are small with round to ovoid nuclei, indistinct nucleoli with poorly defined, pale eosinophilic cytoplasm. Mitotic figures are

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**Figure 1:** Gross photograph shows cut section of the tumor with grey white myxoid areas

**Figure 2:** Ill-defined sweeping fascicles of spindle to stellate cells oval nuclei (H and E, ×40)

**Figure 3:** Ill-defined sweeping fascicles of spindle to stellate cells oval nuclei (H and E, ×40)

**Figure 4:** Stellate cells in a myxoid stroma (H and E, ×40)

**Figure 5:** The collagenized stroma can also exhibit keloidal texture. Interspersed thin walled vessels are seen (H and E, ×40)
sparse. Nuclear anaplasia and necrosis are generally not evident, although a dedifferentiated variant with sheets of anaplastic cells has also been described [3]. Various other histological patterns include storiform, fascicular-herringbone pattern, formation of rosettes, areas of hypercellularity and foci of rounded epithelioid cells. The less commonly noted features include focal osseous metaplasia, scattered multinucleated cells and cystic change [7,8].

Some tumors have a rich capillary vascular network in myxoid areas, as seen in myxoid liposarcoma. However, the vascular channels seen in LGFMS are usually curvilinear, thicker and coarser than those seen in myxoid liposarcoma and myxofibrosarcoma [9].

At the ultra-structural level the neoplastic spindle cells show characteristics of fibroblasts consistent with the fibroblastic nature of LGFMS [6].

The microscopic differential diagnosis of LGFMS includes a number of entities characterized by spindle cell proliferations [3,10]. These include the following:

### Differential diagnosis

<table>
<thead>
<tr>
<th>Entities</th>
<th>Architectural and IHC features</th>
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<tbody>
<tr>
<td>Gastrointestinal stromal tumor</td>
<td>• Predominantly spindled or epithelioid, seldom show broad zones of collagenization, vaguely neural-appearing cells with fibrillar cytoplasm</td>
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<tr>
<td>• CD117, CD34: Positive</td>
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<tr>
<td>Intra-abdominal fibromatosis</td>
<td>• Long, sweeping fascicles of uniform myofibroblastic cells arranged around thin walled, dilated blood vessels</td>
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<td>Desmoid fibromatosis</td>
<td>• The cellularity is uniform from field to field in contrast to the variable cellularity of LGFMS</td>
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<td>• Cells have vesicular nucleus</td>
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<tr>
<td>• Contain more collagen</td>
<td></td>
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<td>• β-catenin protein: Aberrant nuclear accumulation</td>
<td></td>
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<tr>
<td>Inflammatory myofibroblastic tumors</td>
<td>• Prominent stromal collagenization reminiscent of areas within LGFMS, contain cellular spindled zones composed of plump myofibroblasts and “ganglion-like” myofibroblasts</td>
</tr>
<tr>
<td>• Prominent chronic inflammatory cell infiltrate</td>
<td></td>
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<tr>
<td>• Smooth muscle actin and anaplastic lymphoma kinase (ALK-1): Strong positivity</td>
<td></td>
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<tr>
<td>Myxoid neurofibroma</td>
<td>• Encapsulated</td>
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<tr>
<td>• Tumor cells have slender and wavy nucleus</td>
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<tr>
<td>• S-100: Positive</td>
<td></td>
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<tr>
<td>Fasciitis with myxomatous degeneration</td>
<td>• Superficial location, Similar histology</td>
</tr>
<tr>
<td>• Smooth muscle actin, muscle specific actin: Positive</td>
<td></td>
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<tr>
<td>Myxoid variant of malignant fibrous histiocytoma</td>
<td>• Cellular areas, more pleomorphic cells</td>
</tr>
<tr>
<td>Myxoid malignant peripheral nerve sheath tumor</td>
<td>• Alternating hypocellular and hypercellular areas, focal peripheral palisading</td>
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<td>• Only focally shows myxoid change</td>
<td></td>
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<tr>
<td>Synovial sarcoma with myxoid component</td>
<td>• Some tumors are S-100 positive</td>
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<td>• Cytokeratin and epithelial membrane antigen: Positive</td>
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IHC: Immunohistochemistry, LGFMS: Low grade fibromyxoid sarcoma

It is important to distinguish LGFMS and low grade myxofibrosarcoma. The latter is considered as a low grade myxoid variant of malignant fibrous histiocytoma, because of the significantly lower metastatic potential of low grade myxofibrosarcoma compared with LGFMS [9,10]. Low grade myxofibrosarcoma typically has more uniform myxoid stroma, with less swirling of tumor cells and more cellular atypia [3,11].

Despite the varied histological features, these appearances are not related to patient survival although a small tumor size may be a favorable prognostic factor [7,11].

On IHC, these tumor cells show positive staining with vimentin, but no immune-reactivity with antibodies to muscle specific actin, S-100 protein, CD34, CD31, keratin, desmin or epithelial membrane antigen [3,4,12]. Mucin 4 has been recently
described to be a highly sensitive and quite specific IHC marker for LGFMS [13,14].

A chromosomal translocation (t (7;16) (q34;p11)) which results in a novel fusion gene (Fused in Sarcoma/cyclic adenosine monophosphate responsive element binding protein 3-like 2) is well characterized and constitutes an excellent tool in the differential diagnosis of LGFMS as it accounts for more than 95% of the cases. The demonstration of a ring chromosome may be associated with an increased risk of tumor progression [4,8].

In LGFMS, surgery is the only treatment resulting in disease-free periods. LGFMS is not very chemo- or radiosensitive due to the low grade of malignancy with low mitotic rate. Atypical metastatic potential, with atypical sites of metastases poses a problem in deciding the optimal duration of follow-up in these patients [2].

CONCLUSION

LGFMS are rare soft tissue tumors characterized by a deceptively bland appearance and potential for late metastases. At present, surgery is the only treatment that results in disease-free periods. Diagnosis of LGFMS remains problematic because of its bland histologic features that can be potentially confused with other benign or low grade soft-tissue tumors. Careful consideration of the morphological and IHC features of these tumors would render a positive diagnosis.

REFERENCES


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