

## **Low-Grade Fibromyxoid Sarcoma: A Rare Distinctive Soft Tissue Tumor**

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**A fifty-three-year-old Bahraini male presented with a painless, slow growing mass in the left gluteal region. A work-up including pelvic MRI, chest CT scan and excisional biopsy was performed. The histology was low-grade fibromyxoid sarcoma (LGFMS). Immunohistochemistry tests confirmed the diagnosis. He received local radiotherapy as an alternative for re-resection.**

**Changes in the liver were suspicious but an ultrasound and MRI of the liver ruled out liver metastases.**

**The patient is maintained under close observation because the tumor has high tendency of local recurrence and possible pulmonary and liver metastases.**

**This specific distinctive entity of soft tissue sarcoma is yet to be reported in Bahrain; it has no clear protocol regarding the best follow-up recommendations.**

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Low-grade fibromyxoid sarcoma (LGFMS) is considered 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<sup>1</sup>. This rare tumor was first reported by Evans in 1987; therefore, it was called Evans tumor<sup>2</sup>. Its exact incidence is not well-known. These tumors usually occur in the proximal extremities and trunk<sup>2,3</sup>. It can be found rarely in the retro-peritoneum, head or the chest wall<sup>4</sup>. The majority of cases occur in sub-fascial deep locations and affects 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<sup>4,5</sup>.

The aim of this presentation is to report a 53-year-old Bahraini male who developed a large mass in the left gluteal region. Imaging findings, surgical approach to the mass, distinctive immunohistochemistry and cytogenetic findings were highlighted.

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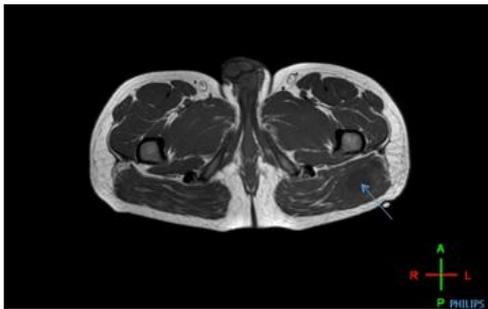
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## THE CASE

A fifty-three-year-old Bahraini male presented with slow growing painless two masses, one on the left gluteal region and the other on the lower left thigh posteriorly. Both masses were noted two years prior to the first visit. The left gluteal mass was painless at the start but began causing discomfort whilst sitting as it increased in size over the course of six months. There was no history of trauma; the mass was deep with no skin changes. His past history was only indicative of hypertension for which he was on regular therapy.

On examination, there was a large 10 cm well-marginated firm mass at the left gluteal region. There was another 3 cm soft mass on the lateral aspect of the lower one third of the left thigh, which appeared to be a lipoma. No enlarged inguinal lymph nodes were noted and also no abdominal organ or mass was palpable. Systemic examination was unremarkable.

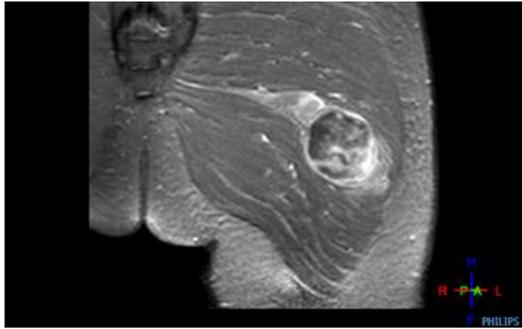
Chest X-ray was normal and pelvic MRI using T1, T2 weighted TSE SPI and post contrast sequences showed 5x3.6x6 cm hypo-intense lesion inside the left gluteus maximus muscle, located posterior to the neck and proximal to femoral shaft, see figure 1. It was multiloculated with 5.7 cm tubular medial extension up to the level of S5 with heterogeneous enhancement after contrast with non-enhancing cystic areas, see figures 2 and 3. No enlarged pelvic or inguinal lymph nodes could be seen. The clinical and radiological impressions were those of possible intramuscular myxoma, myxoid liposarcoma or malignant fibrous histiocytoma.



**Figure 1: T1 Weighted Axial Image: Oval Hypointense Lesion at Left Gluteus Maximus Causing Mass Effect on Muscles Fibers**



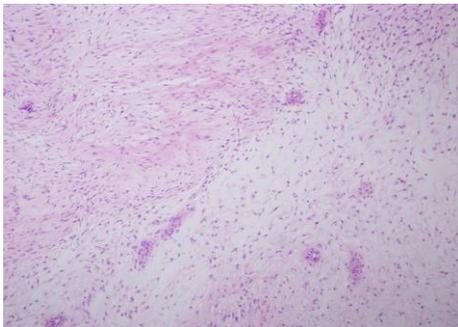
**Figure 2: T2 Weighted Coronal Image: Hyperintense Well-Defined Septated Lesion in the Left Gluteus Maximus Muscle with Medial Extension**



**Figure 3: T1 Weighted Fat Suppression Coronal Image with Contrast Shows Heterogeneous Enhancement 2 of the Mass with Enhancing Extension Medially to Level of the Fifth Sacral Vertebra**

A complete excision of the mass with its extension was done on 23/4/2012. At surgery about 10x12 cm lobulated tumor mass, intramuscular and adherent to muscle fibers. The lump was excised along with the attached muscle bundles of fibers. The operation wound was irrigated with hypertonic saline. On gross examination, these resected specimens consisted of an open oval grayish white cystic mass filled with mucoid gelatinous material measuring 5x4x2.8 cm; another irregular mass measuring 2x1.5x1.2 cm was noted. Multiple sections of these masses were taken for histopathological examination.

Light microscopy revealed well-circumscribed and partly encapsulated pleomorphic lesions of variable cell density with alternating areas of hypo and hyper-cellular areas made of large cells with vacuolated cytoplasm, satellite cells and bland spindle cells with occasional pale eosinophilic cytoplasm and frequent plexiform blood vessels in the myxoid areas with frequent mitotic figures. The picture was of focally infiltrative intermuscular myxoid tumor, see figure 4.



**Figure 4: Hyper and Hypo-Cellular Areas of Bland Spindle Cells in Myxoid Background**

Immunohistochemistry revealed positive vimentin, CD34 was only focally positive; desmin, SMA, S100 were negative. The most likely diagnosis is low-grade fibromyxoid sarcoma (LGFMS). As there was doubt regarding the margins of the excision and the decision was against re-excision; the patient was given 33 sessions of radiotherapy.

Follow-up at the oncology clinic showed that the patient was asymptomatic apart from induration noted at the operative site with evident radiation changes. CT scan of the chest was normal but an upper abdominal ultrasound showed multiple hypo echoic lesions in the

liver. Further work-up by MRI for the liver and the pelvis showed no residual lesion at the operative site and the changes in the liver were suggestive of fatty change with areas of fat sparing around gall bladder bed and focal areas at subscapular locations. These changes were considered to be benign. Ultrasound of the liver performed after one year has shown no changes and no signs of recurrence.

## DISCUSSION

Low-grade fibromyxoid sarcoma (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 (EMA), CD34 and CD31<sup>6</sup>. In our case, these results were negative and in consistence with expected diagnosis of LGFMS.

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)<sup>7</sup>. Reports have suggested that both entities are the same and reflect the neoplastic process<sup>7</sup>. 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 CREB3L2 genes in both LGFMS and HSTGR were confirmed<sup>8,9</sup>. This translocation seems to be specific for the diagnosis of these tumors; these tests are not available locally.

The differential diagnosis of LGFMS include 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 immunohistochemical stains (IHC)<sup>10</sup>.

It is difficult to diagnose LGFMS on fine needle aspiration or needle core biopsy; good excisional biopsy is advised.

LGFMS presents usually as long standing painless soft tissue mass as the case with our patient. The pre-biopsy duration can be up to 5 years in 15% of cases. On rare occasions it may present acutely such as acute respiratory distress and chest pain in case of chest wall LGFMS or as seizure activity in a patient with intracranial LGFMS.

Imaging findings of LGFMS are non-specific but certain CT and MRI findings have been described.

Evans in 1993 and Goodlad et al suggested that LGFMS were paradoxically aggressive tumors<sup>2,3</sup>. In the early series, the local recurrence was noted in 68%, metastases in 41% and death from disease in 18%. All of these cases were initially diagnosed and treated as benign lesions<sup>11</sup>.

In recent series, local recurrence was noted in 54%, metastases in 6% and death from disease in 2%<sup>8</sup>.

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<sup>8</sup>. 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<sup>2,3,12</sup>.

Because of the high risk of late metastases, clinical follow-up and chest imaging are recommended for an extended period of time that is not yet well-defined.

## CONCLUSION

**Low-grade fibromyxoid sarcoma is a rare type of soft tissue sarcoma and it is a form of fibrosarcoma affecting young and middle-aged adults. It has a distinctive presentation and histological features, immunohistochemistry results and specific cytogenetic changes. Surgical management is the standard therapy and it has a high recurrence and metastatic potential. Local radiotherapy may be an alternative for re-resection if this is found to be technically difficult.**

**This case could be considered one of the early cases to be reported in Bahrain. Specific cytogenetic tests may be needed if there is doubt about the diagnosis and these tests are not yet available in Bahrain.**

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## REFERENCES

1. Arnaoutoglou C, Lykissas MG, Gelalis ID, et al. Low-grade Fibromyxoid Sarcoma: A Case Report and Review of Literature. *J Orthop Surg Res* 2010; 5: 49.
2. Evans H. Low Grade Fibromyxoid Sarcoma: A Report of 12 Cases. *Am J Surg Pathol* 1993; 17(6): 595-600.
3. Goodlad JR, Mentzel T, Fletcher CD. Low-grade Fibromyxoid Sarcoma: Clinicopathological Analysis of Eleven New Cases in Support of a Distinct Entity. *Histopathology* 1995; 26(3): 229-37.
4. Dei Tos AP. The Conceptual Evolution of Low-grade Fibromyxoid Sarcoma. Available at: <http://www.uscap.org/site~/iap2006/slides18-2v.htm>. Accessed on 12.2.2013.
5. Steiner MA, Giles HW, Daley WP. Massive Low-grade Fibromyxoid Sarcoma Presenting as Acute Respiratory Distress in a 12-year-old Girl. *Pediatr Radiol* 2009; 39(4): 396-9.
6. Sanford School of Medicine. Low-grade Fibromyxoid Sarcoma. Available at: [http://surgpathcriteria.stanford.edu/softfib/low\\_grade\\_fibromyxoid\\_sarcoma/printable.html](http://surgpathcriteria.stanford.edu/softfib/low_grade_fibromyxoid_sarcoma/printable.html). Accessed on 18.2.2013.

7. Folpe AL, Lane KL, Paull G, et al. Low-grade Fibromyxoid Sarcoma and Hyalinizing Spindle Cell Tumor with Giant Rosettes: A Clinicopathologic Study of 73 Cases. *Am J Surg Pathol* 2000; 24(10): 1353-60.
8. Wu X, Petrovic V, Torode IP, et al. Low Grade Fibromyxoid Sarcoma: Problems in the Diagnosis and Management of a Malignant Tumour with Bland Histological Appearance. *Pathology* 2009; 41(2): 155-60.
9. Möller E, Hornick JL, Magnusson L, et al. FUS-CREB3L2/L1-Positive Sarcomas Show a Specific Gene Expression Profile with Upregulation of CD24 and FOXL1. *Clin Cancer Res* 2011; 17(9): 2646-56.
10. Jagtap SV, Nikumbh DB, Chougule PG, et al. Low-grade Fibromyxoid Sarcoma Presented with Local Recurrence and an Extensive Foot Deformity. *JCDR* 2012; 6(6): 1070-2. Available at: [http://www.jcdr.net/article\\_fulltext.asp?issn=0973-709x&year=2012&month=August&issue=6&id=2337](http://www.jcdr.net/article_fulltext.asp?issn=0973-709x&year=2012&month=August&issue=6&id=2337). Accessed on 15/2.2013.
11. IARC. WHO Classification of Tumours. Low-grade Fibromyxoid Sarcoma. Available at: <http://www.pubcan.org/icdotopo.php?id=5238>. Accessed on 15.8.2012.
12. Shidham VB, Ayala GE, Lahaniatis JE, et al. Low-grade Fibromyxoid Sarcoma: Clinicopathologic Case Report with Review of Literature. *Am J Clin Oncol* 1999; 22(2): 150-5.