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Low-Grade Fibromyxoid Sarcoma in Breast: A Rare Case Report

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Abstrak

Sarkoma fibromyxoid tingkat rendah (LGFMS) adalah tumor jaringan lunak langka yang terdiri dari sel-sel gelendong hambar dalam stroma fibrosa hingga myxoid yang bervariasi, meskipun penampilan histologisnya jinak, menunjukkan risiko metastasis yang signifikan. 1% atau kurang dari semua kanker dengan asal rongga adalah LGFMS. Jarang, mereka dapat ditemukan di tempat-tempat aneh termasuk retroperitoneum, kepala, atau dinding dada. Berikut ini kami sajikan sebuah kasus, Seorang pasien wanita berusia 61 tahun datang dengan massa yang tumbuh lambat tanpa rasa sakit dan secara klinis ganas di payudara kiri. Sudah hadir selama 4 tahun. Biopsi dilakukan, ditemukan stroma tersusun atas sel-sel berbentuk gelendong, nukleus vesikuler tersusun melintang. ada sedikit jaringan myxoid dan ada fokus jaringan hialin dengan sedikit kalsifikasi. 1-2 sel mitosis dan banyak infiltrasi limfosit plasma terlihat. LGFMS secara histologis ditandai sebagai neoplasma jaringan lunak yang tampak hambar, tetapi tumor ditemukan terkait dengan perilaku agresif dan tingkat kekambuhan lokal yang tinggi atau metastasis jauh. Entitas serupa dicirikan oleh mawar raksasa dan adanya area pulau aselular terhialinisasi yang dikelilingi oleh sel oval dan gelendong telah dinamai sebagai "Tumor Sel Spindel Hyalinisasi dengan Mawar Raksasa" (HSTGR). Kehadiran area fokal seluleritas tinggi, pembesaran inti, peningkatan aktivitas mitosis, dan nekrosis tidak dianggap sebagai prognostik yang buruk untuk kekambuhan atau metastasis. Manajemen bedah adalah terapi standar dan memiliki potensi kekambuhan dan metastasis yang tinggi. Ukuran tumor yang kecil mungkin merupakan faktor prognostik yang menguntungkan. Radioterapi lokal perlu dianjurkan untuk pencegahan kekambuhan dan metastasis.

Kata kunci: *Sarkoma Fibromyxoid Tingkat Rendah, Spindled Cells*

Abstract

Low grade fibromyxoid sarcoma (LGFMS) is a rare soft tissue tumour composed of bland spindled cells in a variably fibrous to myxoid stroma, despite the benign histologic appearance, exhibits a significant risk for metastasis. 1% or less of all cancers with cavity origins are LGFMS. Rarely, they may be discovered in odd places including the retroperitoneum, head, or chest wall. Here we present a case, A 61-year-old female patient presented with a slow-growing painless mass and clinically malignant on the left breast. It has been present for 4 years. Biopsy was performed, found The stroma is composed of spindle-shaped cells, vesicular nuclei arranged crosswise. there is little myxoid tissue and there are foci of hyaline tissue with little calcification. 1-2 mitotic cells and numerous plasma lymphocyte infiltrates are seen. LGFMS was histologically 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. 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). 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. 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.
Keywords: Low Grade Fibromyxoid Sarcoma, Spindled Cells

INTRODUCTION

Low-grade fibromyxoid sarcoma (LGFMS) is a rare soft tissue tumour composed of bland spindled cells in a variably fibrous to myxoid stroma, despite the benign histologic appearance, exhibits a significant risk for metastasis. These tumors generally occur in young to middle-aged adults, sometimes in children, but rarely in the high-aged adults. 1% or less of all cancers with cavity origins are LGFMS.(1,2)

Lately, studies on genetic analysis have unraveled a characteristic underlying recurrent identified a hyalinizing spindle cell tumor with giant rosettes as a variant of LGFMS. LGFMS typically presents as deep, intramuscular, soft-tissue masses of the proximal extremities and trunk. Rarely, they may be discovered in odd places including the retroperitoneum, head, or chest wall.(1)

RESULT AND DISCUSSION

LGFMS patient were identified using a validated clinical database in RSUP Prof. Dr. R.D. Kandou. A retrospective review of our histopathological database was carried out. We collects basic patient data, specific data on tumor characteristics and treatment, and data on followup. Presented a patient with a slow-growing painless mass and clinically malignant on the left breast. It has been present for 4 years before the first visit. The tumor mass was hard in consistency, immobile, was growth out the tissue, approximately 8 x 10 cm in size, and located at the left of the left nipple in the chest region. There was no history of trauma. There are enlarged local lymph nodes in unilateral were noted and also hepatomegaly was palpable.

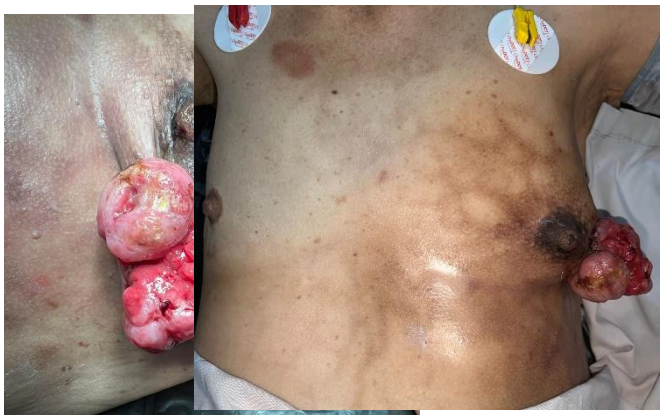
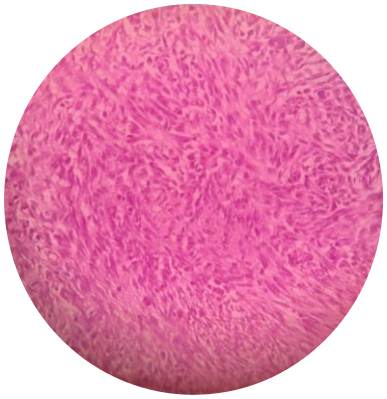


Figure 1 and 2 : Preoperative profile

Routine blood investigations were normal. Chest X-ray was normal. We performed an excisional biopsy to confirm the diagnosis, A complete excision of the mass with its extension was done and at surgery. Found the stroma cells contained composed of spindle-shaped cells, vesicular nuclei arranged

crosswise. there is little myxoid tissue and there are focus of hyaline tissue with little calcification. 1-2 mitotic cells and numerous plasma lymphocyte infiltrates are seen. So the diagnosis was given as LGFMS.



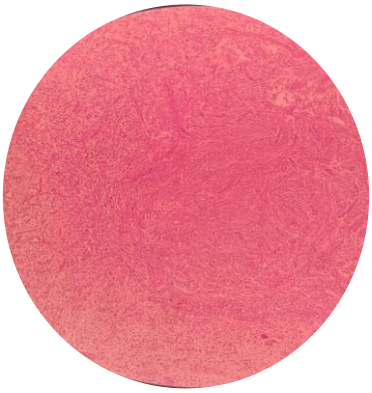


Figure 3 & 4, Excision biopsy shows that the final histopathologic findings show a proliferation of spindle cells

LGFMS is a rare tumor type first described by Evans in 1987 as a distinctive masswith bland histological features and a paradoxically aggressive behavior.(Wang et al., 2017) LGFMS typically presents in the proximal extremities and trunk and is found rarely in the retroperitoneum, head, or the chest wall. LGFMS is a variant of fibrosarcoma with distinctive histopathological features.(Naik et al., 2021; Tay et al., 2018) 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. 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.

Deep LGFMS was linked to a high probability of both distant metastasis (41%) and local recurrence (68%) in the series reported by Evans in 1993.(Almaskati et al., 2013; Naik et al., 2021) 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%. Tumor recurrence following surgery occurs in 18%-29% of patients, most often within the first 2 years after surgery.(Grimaldi et al., 2018) 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. 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.(Almaskati et al., 2013) Respectively. Adequate surgical excision of the tumor is the best treatment option because of the frequent recurrence of LGFMS.(Grimaldi et al., 2018).

CONCLUSION

Surgical management is the standard therapy and it has a high recurrence and metastatic potential. Complete surgical excision is currently the best treatment option. Small tumor size may be a favorable prognostic factor. Local radiotherapy needs to be advocated for the prevention of recurrence and metastasis.

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