Low-Grade Fibromyxoid Sarcoma of Anterior Abdominal Wall

Karam Singh · Sumeet Singh · Narinder Pal ·
Sunil K. Sampley · Kapil Chhabra

Received: 25 June 2009 / Accepted: 14 July 2009 / Published online: 1 May 2012
© Association of Surgeons of India 2012

Abstract Low grade fibromyxoid sarcoma (LGFMS) is a rare soft tissue tumor, having metastatic potential and high local recurrence rate despite its low grade histologic findings. This tumor of deep and subcutaneous soft tissues occurs in 3rd and 5th decade of life. Diagnosis of LGFMS remains problematic because of its bland looking histologic features that can be potentially confused with other benign or low grade soft tissue tumors. We report here a rare case report of low grade fibromyxoid sarcoma of anterior abdominal wall.

Keywords Low grade fibromyxoid sarcoma · Immunohistochemistry · Anterior abdominal wall · Epigastrium

Introduction

Low grade fibromyxoid sarcoma (other names: Evan's tumor, hyalinising spindle cell tumor with giant rosettes) is a rare, low grade malignant tissue neoplasm with a potential for local recurrence as well as distant metastases [1]. It is characterized by bland histologic features and a paradoxically aggressive clinical course. This tumor is commonly located in the lower extremities. Other sites include neck, axilla, chest wall, shoulder, inguinal region and rarely mediastinum, retro peritoneum, mesentery and pelvis [2]. We report a rare case of low grade fibromyxoid sarcoma of anterior abdominal wall.

Case Report

A 30 year old married female presented to surgery OPD with chief complaint of swelling epigastric region for last 6 months (Fig. 1). Swelling was associated with mild pain and occasional watery discharge. There was no history of fever, loss of weight, loss of appetite, jaundice, cough, urinary or bowel complaints. Past history of surgery for swelling at same site 1 yr back was present. After 6 months the swelling recurred at the same site (Fig. 2).

Histopathological report of the swelling excised 1 year back showed desmoid tumor and neurofibroma at a laboratory outside. On examination a horizontal scar was present in the epigastric region with swelling in the midline of the scar. Swelling was about 10 cm × 8 cm in size, well circumscribed, hard in consistency, with smooth surface having hemorrhagic patches and not attached to the underlying structures. Color of the swelling was dusky red with no active discharge from the swelling.

All routine investigations were within normal limits. Ultrasonography of abdomen, chest x-ray and liver function tests were also within normal limits.

Wide excision of the swelling was done and specimen was submitted for histopathological examination. The histopathology report suggested possibilities of desmoid tumor and neurofibroma and advised immunohistochemistry. The specimen blocks and slides were sent to a higher institute for histopathology and immunohistochemistry. Histopathological examination showed partially circumscribed tumor in the dermis infiltrating into subcutaneous fat and soft tissue at deeper aspect. The tumor predominantly displayed areas of variable cellularity- loose, predominantly spindle cells with elongated nuclei, mild nuclear pleomorphism and increased mitotic activity (Fig. 3). Cartilage formation was also seen. On immunohistochemistry the tumor cells were

K. Singh (✉) · S. Singh · N. Pal · S. K. Sampley · K. Chhabra
Department of General surgery, Govt. Medical College Patiala,
Patiala 147003 Punjab, India
e-mail: dr_karam52@yahoo.co.in

Fig. 1 Swelling present in the epigastric region over the old operative scar

vimentin positive and S-100 negative. These features were suggestive of fibromyxoid sarcoma, low grade.

After two weeks patient underwent split skin grafting of wound by plastic surgeon. The patient was subsequently referred to radiotherapy department for further management.

Discussion

Soft tissue sarcoma is the collective term for malignancies arising in muscles, fat, vessels, peripheral nervous system and fibrous tissue [1]. Histopathological examinations have revealed a large number of distinct entities. Low-grade fibromyxoid sarcoma is a distinctive, indolent soft-tissue sarcoma with aggressive clinical course characterized by multiple local recurrences over several years [3].

Low-grade fibromyxoid sarcoma was first described by Evans in 1987 [4]. Patients of any age may be affected, and

Fig. 2 Lateral view of the swelling over the previous scar

Fig. 3 Histopathology of low grade fibromyxoid sarcoma showing predominantly spindle cells with elongated nuclei, mild nuclear pleomorphism and increased mitotic activity (H&E,100X)

the male: female ratio is 1:1. This is a neoplasm of the deep and subcutaneous soft tissue and is commonly located in the lower extremities. Other sites include neck, axilla, chest wall, shoulder, buttock and the inguinal region. The tumor may be rarely located in the mediastinum, retro peritoneum, mesentery and the pelvis.

Diagnosis of low-grade fibromyxoid sarcoma is problematic due to its deceptive bland histologic features [5]. The tumor is grossly well circumscribed and size ranges from 1–20 cm. Histologically, the tumors are composed of alternating fibrous and myxoid areas with various cellularity and with swirling and whorled growth patterns [6]. At the ultrastructural level the neoplastic spindle cells have characteristics of fibroblasts consistent with the fibroblastic nature of low-grade fibromyxoid sarcoma [7]. Immunohistochemically, the tumor cells are diffusely and strongly positive for vimentin and desmin. Immunohistochemical features of low-grade fibromyxoid sarcoma can help to exclude many cutaneous and deep soft tissue tumors from the differential diagnosis [4].

Recent cytogenetic and molecular analyses have shown that most low-grade fibromyxoid sarcomas have a characteristic chromosomal abnormality, t(7;16)(q33;p11), resulting in the FUSCREB3L2 fusion gene [8]. However, such assays have only rarely been used to analyze formalin-fixed, paraffin-embedded tumor samples.

Conclusion

Careful consideration of the morphological and immunohistochemical features of these tumors permits a positive diagnosis of low grade fibromyxoid sarcoma and allows its distinction from a number of other benign and malignant soft tissue neoplasms.

References

1. Evans HL (1987) Low grade fibromyxoid sarcoma, A report of 2 metastasizing neoplasms having a deceptively benign appearance. *Am J Clin Pathol* 88(5):615–19
2. Goodlad JR, Mentzel T, Fletcher CD (1995) Low grade fibromyxoid sarcoma: clinicopathological analysis of eleven new cases in support of a distinct entity. *Histopathology* 26(3):229–37
3. Shidham VB, Ayala GE, Lahaniatis JE, Garcia FU (1999) Low-grade fibromyxoid sarcoma: clinicopathologic case report with review of the literature. *Am J Clin Oncol* 22(2):150–5
4. Nichols GE, Cooper PH (1994) Low-grade fibromyxoid sarcoma: case report and immunohistochemical study. *J Cutan Pathol* 21(4):356–62
5. Goodlad JR, Mentzel T, Fletcher CD (2000) Low grade fibromyxoid sarcoma: clinicopathological analysis of eleven new cases in support of a distinct entity. *Histopathology* 36(1):92–3
6. Zámecnik M, Michal M (2000) Low-grade fibromyxoid sarcoma: a report of eight cases with histologic, immunohistochemical, and ultrastructural study. *Ann Diagn Pathol* 4(4):207–17
7. Lindberg GM, Maitra A, Gokaslan ST, Saboorian MH, Albores-Saavedra J (1999) Low grade fibromyxoid sarcoma: fineneedle aspiration cytology with histologic, cytogenetic, immunohistochemical, and ultrastructural correlation. *Cancer* 87(2):75–82
8. Mertens F, Fletcher CD, Antonescu CR, Coindre JM, Colechia M, Domanski HA, Downs-Kelly E, Fisher C, Goldblum JR, Guillou L, Reid R, Rosai J, Sciort R, Mandahl N, Panagopoulos I (2005) Clinicopathologic and molecular genetic characterization of low-grade fibromyxoid sarcoma, and cloning of a novel FUS/CREB3L1 fusion gene. *Lab Invest* 85(3):408–15