Low-grade fibromyxoid sarcoma adjacent to neural structure in a 3-year-old girl: Case report and review of literature

Salih Onur Basat¹, Turgut Kayadibi², İsmail Mithat Akan³, Mehmet Bozkurt¹

ABSTRACT

Low-grade fibromyxoid sarcoma (LGFMS) is a rare adult mesenchymal tumor located mostly in deep soft tissues, which is not common in the pediatric patients. Only 20% of reported cases show patients under the age of 18. A systematic literature review of articles between 1995 and 2013 in MEDLINE and PubMed shows that fewer than 10 fibromyxoid sarcoma have thus far been identified in childhood. In this report authors describe a case of low-grade fibromyxoid sarcoma in the right arm of a 3-year-old girl. The tumor was excised with preservation of nearby neural structures, there was no functional deficit in arm functions and in follow-ups no recurrences have been seen so far. To the best of our knowledge, this is the youngest case of low-grade fibromyxoid sarcoma.

Key words: Low-grade fibromyxoid sarcoma, childhood, axillary nerve, soft tissue tumor

Introduction

Low-grade fibromyxoid sarcoma (LGFMS) is a rare subtype of fibrosing type low-grade fibrosarcoma that was described by Harry L. Evans in 1987¹. LGFMS is seen mostly in young adults and the most common location is in the deep soft tissues of proximal extremities. It is very rare in childhood and seen mostly superficial locations in the children. The youngest cases up to date are two 4-year-old children²-⁴. LGFMS has deceptively benign histological features that are often non-distinctive and blunt nature but paradoxically it is associated with high rates of local recurrence and distant metastases even decades later. It is a slow growing tumor that needs long term clinical follow-up for local recurrences and distant metastases – the lung is the primary site for metastases⁵. Excision with wide surgical margins is the primary treatment option and the tumor is mostly accepted as resistant to usual chemotherapy and radiotherapy treatment modalities. This report describes a 3-year-old child with a deep intramuscular LGFMS, the youngest case reported so far.
Case report

A previously well 3-year-old girl presented with a 3-month history of a mass in the right upper arm’s posterior aspect. The family history was noncontributing. The child’s growth and development had been normal with no past of important disease. On the physical exam, a firm, non-mobile, non-tender mass was palpated at posterior proximal part of right upper arm and no other mass or lymphadenopathy was palpated. Blood analysis values were in normal limits. On ultrasound imaging, the mass was round and measuring 21x18 mm just near the humeral metaphysis. This mass was solid with intralesional nodular areas, with hypoechogenity in periphery and hyperechogenity in the center of lesion. There was arterial blood flow in the periphery of the lesion on doppler imaging. Given that the nature of this lesion was unclear and hypothesized to be a benign desmoid tumor, histopathological examination was recommended. Under general anesthesia an excisional biopsy was performed and the mass was sent to the pathology.

On gross examination the mass was noted to be firm and fibrous, well circumscribed with a thin fibrous pseudocapsule and measuring 2.5x2.3x1.7 cm; the cut surface was shiny white and homogeneous without nodularity or necrosis. (Figure 1a) On microscopy, the tumor had a mixture of heavily collagenized, hypocellular zones and more cellular myxoid nodules. The tumor cells were spindle shaped cells in linear arrangement, with surface was shiny white and homogeneous, B: In second operation, the axillary and radial nerves were preserved, C: Surgical specimen sectioned and sent to 2 independent pathologists.

Figure 1. A: Excisional biopsy material with surface was shiny white and homogeneous, B: In second operation, the axillary and radial nerves were preserved, C: Surgical specimen sectioned and sent to 2 independent pathologists.

Figure 2. A: The tumor cells were spindle shaped organized in and linear arrangement, producing a whorled and swirling growth pattern, B: Immunohistochemically tumor cells stained strongly and diffusely positive for vimentin, CD99 and bcl-2.
producing a whorled and swirling growth pattern. (Figure 2a) Immunohistochemically tumor cells stained strongly and diffusely positive for vimentin, CD99 and bcl-2. Tumor cells did not show immunoreactivity to smooth muscle actin (SMA), S-100 protein, desmin, epithelial membrane antigen (EMA), neurofilament, CD34 and β-catenin. (Figure 2b) In addition, Ki-67 index was less than 2%.

On the basis of clinical, histological and immunohistochemical features, a LGFMS diagnosis was confirmed and the histopathological report revealed that there were no clear zones at surgical excision margins. A wider excision was planned for positive surgical margins. On magnetic resonance imaging, there was a triangularly shaped hyperintense lesion in the surgical area, which was extending to the neurovascular bundle through intermuscular septum on T2 weighted images. After contrast admission, the image of the lesion was enhanced. (Figure 3) Under general anesthesia, in the posterior side of the right upper arm, the incision was made through 1 cm security margin around the old operation scar. The posterior cuff muscles were excised till they were macroscopically tumor free. The tumor was traced deep through intramuscular septum and finally reached the radial and axillary nerves, which were preserved (Figure 1b) The surgical specimen was sectioned, sent to 2 independent pathologists and pathology reports concordantly confirmed the original LGFMS diagnosis and that the re-excision was free of LGFMS. The patient received no chemotherapy or radiotherapy. The patient is now being followed-up with serial radiology and physical examination. During the postoperative first-two years, the patient had no evidence of recurrence or metastasis, and all the arm functions were within normal limits.

**Discussion**

LGFMS is a rare type of fibrosarcoma seen mostly in young adults with no known true incidence. The male to female ratio is 1:1. Synonyms of this tumor include hyalinising spindle cell tumor with giant rosettes and fibrosarcoma - fibromyxoid type. LGFMS is mainly an adult disease and only 20% of the reported cases involve patients under the age of 18[6]. A systematic literature review of articles between 1995 and 2014 in MEDLINE and PubMed shows that fewer than 10 fibromyxoid sarcomas have thus far been reported in childhood[7-8]. In 1987, Evans described two cases, both women in their 20’s with tumors in the scapular region and chest wall soft tissue with lung metastases. Then, 24 years later, the author published long-term re-
sults with 33 cases with ages ranging from 6 to 52 and 3 of them were children. The masses located mostly intramuscularly in the thigh, shoulder and inguinal region, 3 of them were subcutaneous, one had diffuse intra-abdominal presentation. Local recurrence occurred in 21 cases from intervals 0 (incomplete excision) to 15 years, with a median of 31/2 years. Metastases developed in 15 cases after intervals from 0 to 45 years with a median of 5 years. Metastases were mostly in lungs, pleura and chest wall. Fourteen of 33 patients died of tumor after 3 to 42 years (median of 15 years) [9].

The youngest reported cases in the literature, published in 1996 by Canpolat et al and in 2005 by Rando et al, were of two 4-year-old boys. The first case had a tumor in the distal thigh region presenting at 11 years of age with lung nodules [2-3]. After extensive chemotherapy and excision of the tumor and lung metastases, the patient developed recurrent disease. The second case presented with paraparesis and was found to have the tumor located in the paravertebral region. After radical resection of intravertebral region, he was treated with chemotherapy and radiotherapy.

The recommended treatment of LGFMS includes the radical surgery, widely en bloc surgical resection. The role of chemotherapy and radiotherapy remains still unclear. Considering the risks of the high rate of local recurrence and late metastasis, long term close follow up is required [9].

**Conclusion**

The 3-year-old patient was re-operated with wider excision of the involved area and preservation of posterior arm compartment including axillary and radial nerves. There was no functional deficit. The pathology reports confirmed there was no local recurrence or residual tumor. After two years since the operation there is no evidence of local disease or distant metastases, but a long term follow up is needed as a median of 5 years is needed for development of distant metastases. LGFMS is a very rare tumor, difficult to diagnose without an excisional biopsy, but it must be kept in mind in differential diagnosis of unusual mesenchymal tumors [10]. LGFMS is a benign low-grade sarcoma, with malignant behavior, local recurrences and distant metastases.

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