Low-grade fibromyxoid sarcoma in a child presenting as a popliteal fossa swelling

Suraj Gandhi, Charu Sharma, Pankaj Dwivedi, Neha Sisodiya Shenoy, Hemanshi Shah

Department of Paediatric Surgery, TNMC & BYL Nair Hospital, Mumbai, Maharashtra, India

Abstract

Popliteal fossa masses are rare in paediatric age group. Even rarer are the malignancies of this area. Low-grade fibromyxoid sarcoma (LGFMS) is a distinctive variant of fibrosarcoma. It is a rare tumor with benign histologic appearance but high metastasizing potential. We describe an 11-year-old child with a popliteal fossa mass, which was excised, and histopathological report revealed LGFMS.

Keywords

Popliteal fossa, children, swelling, low-grade fibromyxoid sarcoma, soft tissue tumor.

Corresponding author

Charu Sharma, Senior Registrar, Department of Paediatric Surgery, TNMC & BYL Nair Hospital, Mumbai, Maharashtra, India; email: drcharusharma18@gmail.com.

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Introduction

Low-grade fibromyxoid sarcoma (LGFMS) is a distinctive variant of fibrosarcoma. It was first reported by Evans in 1987; hence it is also known as Evans’ tumor [1]. It is a rare tumor with benign histologic appearance but high metastasizing potential. LGFMS is also known as spindle cell tumor as
the cells appear spindle shaped with giant rosettes under microscopic examination [2]. It is usually seen in middle aged adults; however, there are also sporadic reports of such tumors in paediatric age group [3]. We describe an 11-year-old boy who presented with a swelling in the right popliteal fossa.

Case report

An 11-year-old boy presented with swelling in the right popliteal fossa since 5 months. A 5 x 5 x 3 cm hard, non-tender, immobile and non-transilluminant swelling was present in the right popliteal fossa (Fig. 1).

An ultrasound of the local part suggested a well-defined oval hypoechoic mass in the right popliteal fossa with internal vascularity. MRI suggested a 7 x 4.6 x 3.3 cm hyperintense soft tissue lesion with homogenous enhancement, which increased during venous phase suggesting neoplastic etiology (Fig. 2).

Intra-operatively, a fleshy solid mass was found deep to the biceps femoris tendon and both the tibial and common peroneal nerve were passing through it (Fig. 3 and Fig. 4). The mass was encasing both the nerves. It was dissected free from both the nerves and excised completely (Fig. 5). Histopathology suggested low-grade spindle cell sarcoma consistent with LGFMS (Fig. 6). A post-operative positron emission tomography (PET scan) showed low-grade activity at right popliteal region consistent with post surgical inflammation. No other activity was detected in this PET scan. The patient was kept on regular follow-up. The patient is asymptomatic and a repeat PET scan after 6 months suggested no activity.

Discussion

LGFMS is a rare, malignant soft tissue tumor, which is usually seen in extremities (arms and legs), chest or back of young adults [4]. It has also been reported in esophagus and abdominal wall [5]. The median age of presentation is 34 years with range being 3 to 78 years [6]. There is slight male preponderance but no racial predisposition [4].

Although at present there are no identified risk factors for LGFMS, the presence of FUS and CREB3L2 (or CREB3L1) gene fusion has been reported to be a characteristic feature of LGFMS, which is seen in about 76 to 96% of cases [4]. Chromosomal translocations have been reported in 65% of cases [4]. Ring chromosome anomaly is seen in 25% of cases [4]. Cytogenetic studies showing two cell lines containing balanced translocation between chromosomes 7 and 16 have
also been reported [2]. Other factors including injury and inflammation have also been reported to be causative factors for LGFMS [5].

LGFMS has usually a prolonged preclinical stage with history of a slow-growing painless, well-circumscribed soft tissue mass [7]. The size of the tumor ranges from 1 to 20 cm with average size at diagnosis reported to be approximately 5 cm [4]. The tumor generally begins in nerve sheath and in layers of connective tissue – under the skin, in fascial planes deep to subcutaneous tissue, between muscles and surrounding organs [5]. Initially, the tumor is well contained and encapsulated and grows very slowly; however, metastasis may occur in later stages [4]. Studies with long-term follow-up have reported recurrence rates of 64%, metastases in 45% and death from this disease in 42% [7]. This tumor usually metastasizes to the lungs, pleura and chest wall [7].

There are no well-established diagnostic criteria for LGFMS [4]. Radiological imaging (CT or MRI) may show multinodular lesion with alternating strongly and weakly enhancing areas [8]. The definitive diagnosis is histopathological.

The tumor is low to moderately cellular with spindle cells containing very scant cytoplasm, uniform elongated nuclei and small inconspicuous nucleoli in a prominent myxoid background with no significant pleomorphism or mitoses [9]. These spindle cells are arranged in interlacing fascicles and bundles with herring bone pattern at places [5]. Forty percent of tumors contain poorly formed but large collagen rosettes [10].

Positive stains include MUC4 (highly sensitive and specific – 100%), CD99 (90%), BCL2 (90%) and EMA, vimentin (non-specific) [11]. The negative stains used are S100, desmin, keratin, CD34, MDM2, smooth muscle actin, h-caldesmon, CD117, nuclear beta-catenin, DOG1 to differentiate LGFMS from other similar tumors [9].

Tumor grading has been recommended so as to improve the management and prognosis and prevent recurrence. Grading is based on nuclear atypia, nuclear overlap, mitotic figures and necrosis [12, 13]. Treatment of spindle cell tumor is complete excision with wide margins [14]. Radiotherapy has minimal role [5]. Long-term follow-up is essential as there is propensity for recurrence [5].

The prognosis is good for small sized and superficially located tumors, which are completely excised [15]. The prognosis is usually not affected by the presence of focal intermediate- to high-grade sarcoma [16]. However, a dedifferentiated recurrence (anaplastic round cell morphology with numerous mitoses) portends short survival [11].
Declaration of interest

The Authors declare that there is no conflict of interest.

References