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Case report

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

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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.

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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

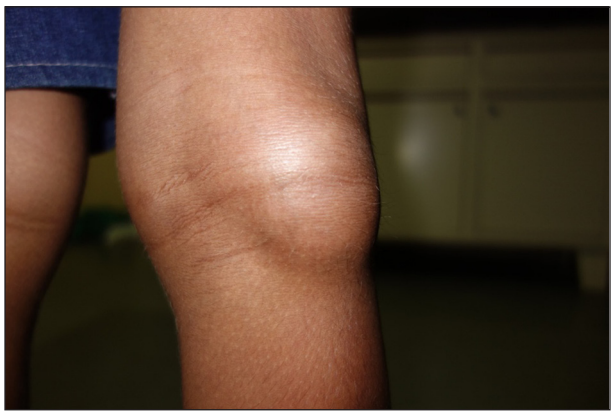

Figure 1. Clinical photograph of the right popliteal fossa swelling.

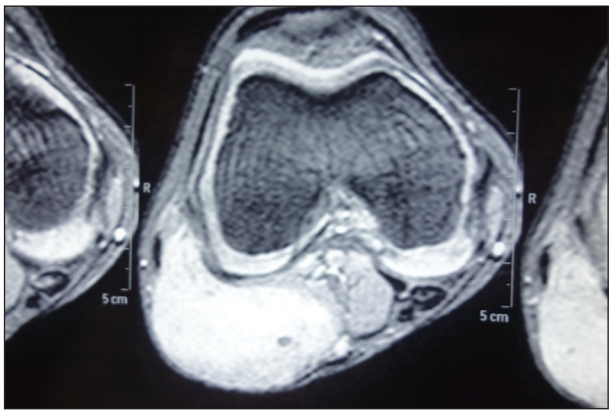

Figure 2. MRI of the popliteal fossa swelling suggesting a 7 x 4.6 x 3.3 cm hyperintense soft tissue lesion with homogenous enhancement, which increased during venous phase suggesting neoplastic etiology.

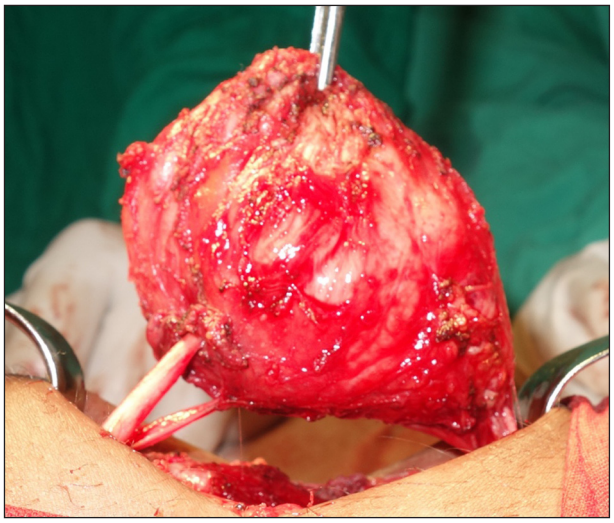

Figure 3. Intraoperative photograph showing the tibial and common peroneal nerves passing through the swelling.

also been reported [2]. Other factors including injury and inflammation have also been reported to be causative factors for LGFMS [5].

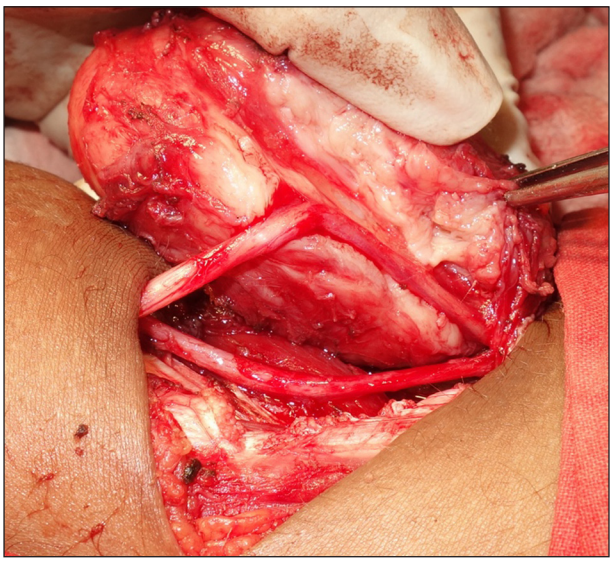

Figure 4. Intraoperative photograph showing the two nerves being dissected off from the swelling.

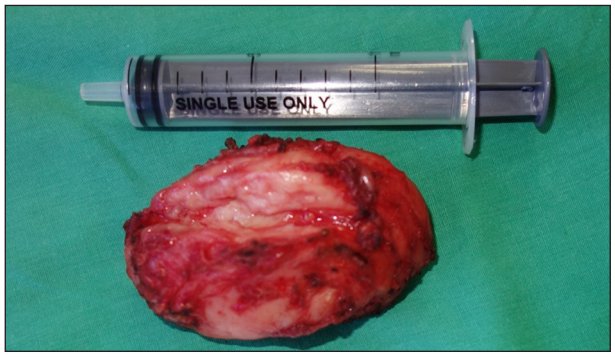

Figure 5. The excised specimen.

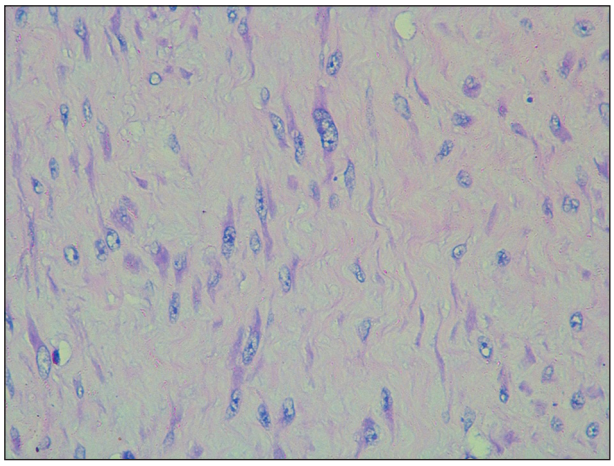

Figure 6. Histopathological image (40x) showing spindle cells containing very scant cytoplasm, uniform elongated nuclei and small inconspicuous nucleoli in a prominent myxoid background.

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.

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