Myxoid liposarcoma in the extremities: A case report

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Received – 04 June 2018 Initial Review – 04 July 2018 Accepted – 01 August 2018

ABSTRACT

Liposarcoma is among the most common sarcomas in the adult population. On the contrary, their incidence in the pediatric population is rare. The most common subtype affecting extremities is the myxoid liposarcoma. The preferred method of surgery for the limb myxoid liposarcoma is the limb-preserving surgeries. In the current study, we report the case of a patient who was affected by myxoid liposarcoma and who underwent conservative management with the impression of lipoma several months before the definite diagnosis was obtained. After diagnosis, he underwent radical resection in a single procedure and was referred to the oncology ward for further adjunct treatment.

Key words: *Limbs, Myxoid liposarcoma, Sarcoma*

Liposarcoma is among the very rare tumor in the pediatric population. The pediatric liposarcoma differs from those that affect the adult population in histopathological characteristics which might result in non-identical prognosis [1]. Most commonly, the primary presentation simulates those with the benign nature which seems to be true especially in extremities liposarcoma. Similar to other types of sarcoma, primary thorough physical examination accompanying with comprehensive laboratory tests is essential. In the case of respectability, treatment consists of limb-sparing surgery with or without adjunct chemoradiotherapy [2].

In the current study, we report the case of a patient who was affected by myxoid liposarcoma and who underwent conservative management with the impression of lipoma several months before the definite diagnosis was obtained. After diagnosis, he underwent radical resection in a single procedure and was referred to the oncology ward for further adjunct treatment.

CASE REPORT

A 4-years-old boy was referred to our department with a chief complaint of a gradual increase in mass located at the lateral aspect of his left arm. The history was not significant for any trauma or recent vaccination. Slow, obvious increase in size was evident during a 6-month period. Initially, the patient underwent watchful waiting management with the impression of lipoma.

The primary physical examination revealed a 5 cm×6 cm firm mass at the lateral aspect of the deltoid muscle. In a comprehensive examination at the surgical clinic, there was no evidence of mass sensation nor tenderness, and there was no evidence of fixation to the underlying structures. There was no evidence of ulceration or overlying skin discoloration. On palpation, a painless firm mass without any fluctuation with adhesion to overlying tissue was evident. Comprehensive sensory-motor physical examination was negative for pathological finding. A thorough examination of mass includes transillumination, which was done by holding the light behind the mass and then the light was blocked by the mass. Distal pulses measurements including ulnar and radial pulses demonstrated full and symmetric pulses with contralateral limb (Fig. 1). There was no specific point in the physical examination of other parts of the body. The left axillary examination denoted 3–4 discrete lymphadenopathies. There was no evidence of synchronized lymphadenopathy in the systemic evaluation.

Laboratory evaluations regarding complete blood count and erythrocyte sedimentation rate clarified normal findings. Other laboratory tests were normal. Plain X-ray of the left arm did not note bony involvement. T1 weighted magnetic resonance imaging (MRI) delineated a multiloculated cystic mass with fine septa and circumscribed borders. MRI images were not significant for bony or muscular invasion (Fig. 2). While axillary lymphadenopathies measuring 15 mm×18 mm were obvious, the patient was scheduled for the surgery. The highest impression was a malignant tumor.

The patient underwent radical resection of the mass with a longitudinal incision over the maximal fluctuation. Intraoperative description of the mass was significant for a multinodular lipoid texture 4 cm×5 cm mass without vascular involvement. The microscopic assessment of the mass elucidated uniformly round and oval shaped cellular sheets in the myxoid matrix. The proportion of the round cells was approximate 20%.

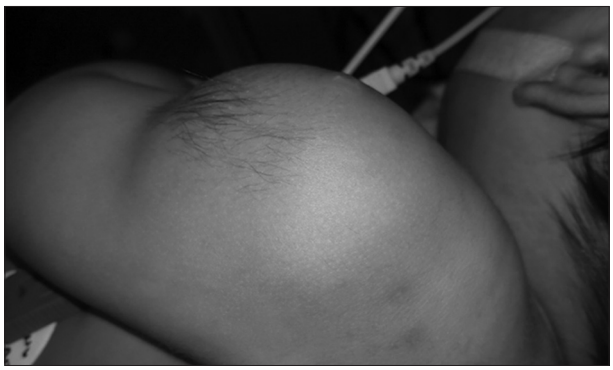

Figure 1: A 4-year old boy with a chief complaint of left arm mass

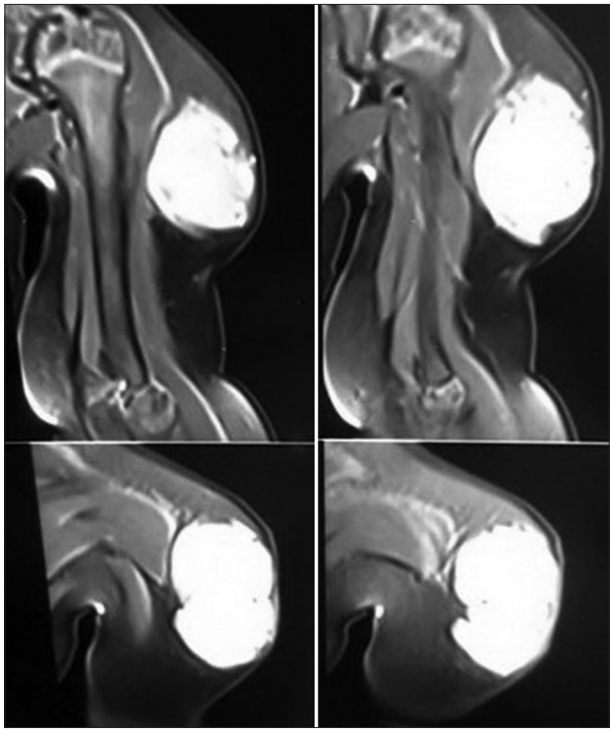

Figure 2: T1 weighted magnetic resonance imaging delineated a multiloculated cystic mass

The hyperchromatin nucleus with the capillary network was significant. The immunohistochemistry assessment was positive for S100, Vimentin, CD34, and CD31, while Cytokeratin and Desmin markers were negative.

Ultimately, the pathological evaluation suggested myxoid liposarcoma. His 1-month early post-operative period before adjuvant therapy was uneventful. The patient follow-up was undertaken by a scheduled method of biennial chest X-ray and the whole body computed tomography scan during the 1st year and annually for the following 3 years. The consequent treatment was administered by the oncology team.

DISCUSSION

Liposarcoma constitutes 20% of sarcomas and is the most common sarcoma during adulthood [1]. Four subtypes of liposarcoma have been described including well-differentiated, dedifferentiated, myxoid round cell, and pleomorphic [2]. The main index of prognosis in liposarcoma is the degree of differentiation, which determines the tumor grade. The anatomical distribution of liposarcoma follows its histologic subtype. Accordingly, the myxoid round cell and pleomorphic subtypes are more common at extremities while the well and dedifferentiated subtypes should be suspected in retroperitoneal involvement.

The extremities are most commonly affected at the thigh, while leg and ankle are among the rare possible locations for liposarcoma. Well-differentiated liposarcoma might invade locally; however, distant metastasis is among their rare presentation. The estimated overall survival for this subtype is approximately 90%, and mortality might be seen due to their local invasion to adjacent retroperitoneal structures [3-6]. On the contrary, those patients affected by dedifferentiated liposarcoma, most commonly present with lung metastasis. Their 5-year survival might be 75% [7]. A pleomorphic subtype accounts for the minority of patients with liposarcoma. It follows a very aggressive, high grade and metastatic course, with survival rate ranging from 30% to 50% [8]. Myxoid liposarcoma accounts for more than 40% of all liposarcomas [9] and commonly affects thigh with a cystic feature in MRI images [10].

A direct correlation among the percentage of a myxoid cell and distant metastasis has been documented. Accordingly, the rate of distant metastasis significantly increases in those myxoid liposarcomas with more than 10% round cells [11].

Translocation of 12; 16 (q13, p11); as a genetic error has been well documented in the myxoid liposarcoma. The most possible mechanism for the mentioned error is fusion among the transcription factor of cyclophosphamide-Halotestin-Oncovin-prednisolone with sarcoma-associated fusion proteins which be diagnosed by the reverse transcriptase-polymerase chain reaction method [12].

Treatment of extremities liposarcoma includes radical resection; adjuvant chemotherapy might be applied based on the histologic subtype [13,14]. Radiotherapy as an adjunct modality of treatment might improve the outcomes, especially in the myxoid subtype. As mentioned, myxoid liposarcoma as an uncommon malignant entity in the pediatric population most commonly affects lower extremities. However, our patient was affected by the upper limb liposarcoma.

Moreover, the radical surgery was delayed due to missed diagnosis, and occasionally, the delayed diagnosis might result in retroperitoneal or neurologic metastasis which precludes the patients from curative management. In one of the studies, Estourgie *et al.* in 2002 mentioned the devastating outcomes of "metastatic patterns of extremity myxoid liposarcoma" [15].

CONCLUSION

Myxoid liposarcoma is an uncommon malignant entity in the pediatric population most commonly affects the upper and lower extremities. Due to the lack of pathognomonic features in radiologic or clinical assessment to differentiate liposarcoma from lipoma, we should keep in mind this diagnosis while proceeding with surgical management.

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Funding: None; Conflict of Interest: None Stated.

How to cite this article: Atqiaee K, Lotfollahzadeh S, Mirshemirani A. Myxoid liposarcoma in the extremities: A case report. Indian J Case Reports. 2018;4(4):310-312.

Doi: 10.32677/IJCR.2018.v04.i04.019