JALALABAD MEDICAL JOURNAL

Case Report

Extra Skeletal Ewing's Sarcoma of the Leg: A Case Report

Sumon Mallik[1], Cyrus Shakiba[2], Lata Majumder[3], Nuruddin Ahmed[4], Farid Ahmed[5], Hasan Mohammad Navin[6], ABM Bodrudduza Chowdhury[7]

[1,5]Associate Professor, Department of Orthopaedics, Jalalabad Ragib-Rabeya Medical College, Sylhet.
[2]Professor, Department of Orthopaedics, Jalalabad Ragib-Rabeya Medical College, Sylhet.
[3]Jr Consultant, Department of Orthopaedics, Jalalabad Ragib-Rabeya Medical College Hospital, Sylhet.
[4]Assistant Professor (Retd), Department of Orthopaedics, Sylhet MAG Osmani Medical College, Sylhet.
[6] Assistant Registrar, Department of Orthopaedics, Jalalabad Ragib-Rabeya Medical College Hospital, Sylhet.
[7]Indoor Medical Officer, Department of Orthopaedics, Jalalabad Ragib-Rabeya Medical College Hospital, Sylhet.

ABSTRACT

Among the malignant bone tumours in children and young adults, Ewing's sarcoma (ES) is the second most common. But extra skeletal Ewing's sarcoma (EES) is an extremely rare disease. Hereby we present a case of 23 year old male with extra skeletal Ewing's sarcoma developed in the leg. The diagnosis was done by radiologically and histologically, but the treatment was multidisciplinary as well as early radical excision. It is to mention that, although this was an unusual location, EES should be contemplated in the differential diagnosis of soft tissue tumours in the limbs.

Keywords: *Ewing's sarcoma, Extraskeletal.*

[Jalalabad Med J 2019; 16(1): 35-38]

INTRODUCTION

Ewing's sarcoma (ES) is a rare malignant tumor and represents a family of morphologically similar small round-cell neoplasms[1]. These tumors generally originate in bone tissue, but they can occasionally originate in soft tissue, known as extra skeletal Ewing's sarcoma (EES) relatively rare soft tissue tumor that is histologically indistinguishable from the osseous form. The diagnosis is mainly based on microscopic histology and immunohistochemistry. It mainly affects very young people. Adult patients are very rare and constitute only 5% of all cases[1,2]. Only approximately 16% of all Ewing's sarcomas are extraskeletal[1,3]. This report describes a case of EES of the leg with extensive involvement of soft tissues without erosion of the bone.

Address of Correspondence:
Dr. Sumon Mallik, Associate Professor, Department of Orthopaedics, Jalalabad Ragib-Rabeya Medical College, Sylhet, Mobile: +8801727414114, E-mail: mallikdr.ortho@gmail.com

CASE REPORT

A 23-year-old male presented with a painless palpable mass on the proximal third of right leg for 40 days. His family history was non contributory. He initially noticed a small swelling which eventually increased in size within 40 days with mild paraesthesia along the area supplied by common paroneal nerve. Physical examination revealed a 42×45 mm, mild painful and firm mass in the lateral aspect of upper 3rd of the right leg which had gradually worsened. The skin over the mass was unremarkable. The patient had no history of radiation or trauma. Motor examinations were normal but there was hyposthesia on the outer aspect of leg and dorsum of right foot and there was no inguinal lymphadenopathy. No other acute symptoms were

reported. Haematological and biochemical investigations were normal including white cell count, erythrocyte sedimentation rate and C-reactive protein. Plain radiography was objectified a soft tissue swelling shadow in the upper third of the right leg. Magnetic resonance imaging (MRI) showed a fairly lobulated soft tissue mass measuring 42×45×73 mm (Figure-1 and Figure-2). There was no calcification or ossification objectified. Adjacent bone and periosteum were normal. Excision Biopsy of the lesion was done preserving the common paroneal nerve (Figure-3 a,b,c and Figure-4). The diagnosis was confirmed by histopathological examination as well as immunohistochemistry (positive staining for CD99) as Ewing's sarcoma.

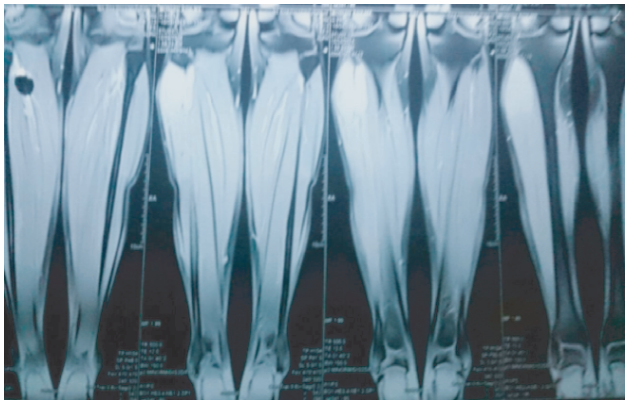


Figure-1: *MRI image showing a heterogeneous and hyper intense tumoral process occupying the upper third of the right leg.*

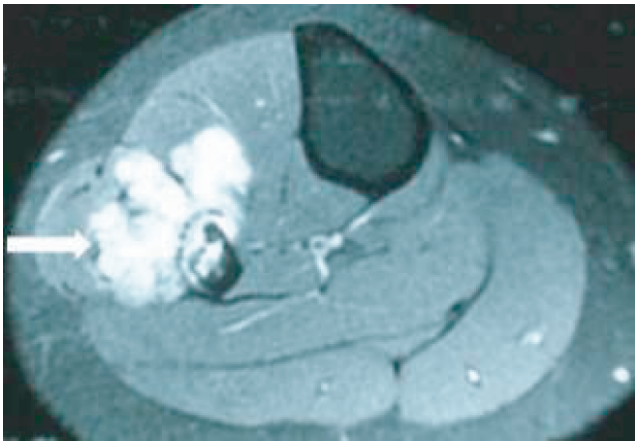


Figure-2: *Axial MRI image showing extra skeletal tumour at the anterior and lateral part of the right leg.*

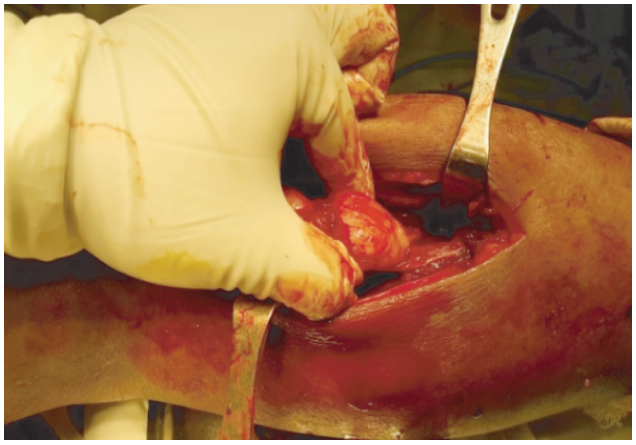


Figure-3(a): *Intra-operative aspect of extra skeletal Ewing's sarcoma of the right leg.*

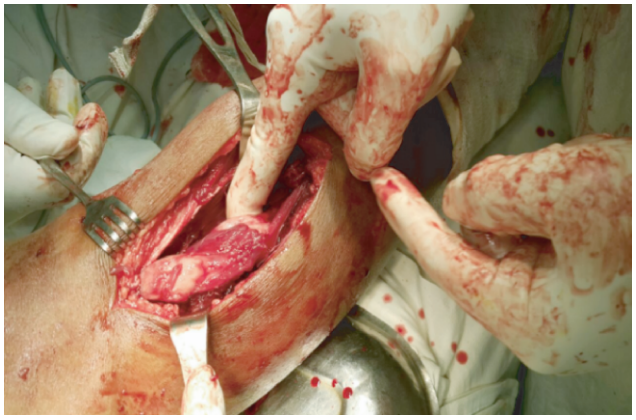


Figure-3(b): *Intra-operative aspect of extra skeletal Ewing's sarcoma of the right leg.*

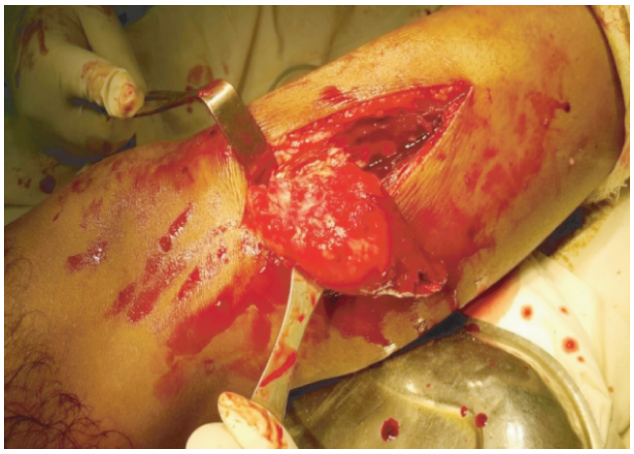
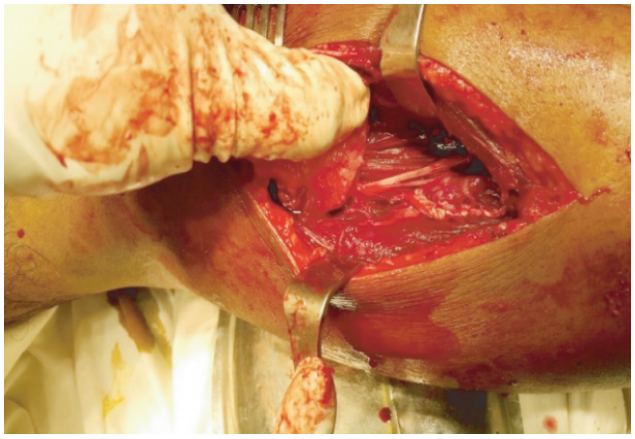


Figure-3(c): *Intra-operative aspect of extra skeletal Ewing's sarcoma of the right leg.*



Fiureg-4: *Intra-operative exposure of preservation of common paroneal nerve after excision of the tumor.*

DISCUSSION

Ewing's sarcoma (ES) is a malignant tumour of bone which is composed of small round tumour cells commonly extend into extraosseous soft tissues at the time of diagnosis[4]. It accounts for 6-8% of all primary malignant bone tumours and predominantly affects children, adolescents and young adults[5]. A substantial proportion of Ewing's sarcoma arise from extraskeletal sites known as extraskeletal Ewing's sarcoma (EES)[6]. The EES develops preferentially on the trunk and axial soft tissues in 50-60% of cases and secondarily in the limbs in 20-25% of cases[7]. Pain and swelling are the leading symptoms associated with general symptoms (fever, asthenia and weight loss). MRI also gives useful information regarding tumor extension, anatomical location, tumor size, and for appropriate planning of surgical treatment. It mainly allows an accurate assessment of the chemotherapy response[8]. Biopsy is essential for definitive diagnosis. The most commonly used is open biopsy technique[7]. Microscopically, the tumor had a small, round, and blue cells with abundant glycogen and an absence of cytoplasmic filaments[1]. It is rather easy to confuse ESS with embrionary neuroblastoma, lymphoma or rhabdomyosarcoma. This is due to the fact that EES shares the histopathologic and immunohistochemistry findings with Ewing's sarcoma. Confirmation of the diagnosis should be based on positive staining for CD99 during immunohistochemistry[7].

Clinically and radiologically, it is difficult to distinguish this disease from other soft tissue malignancy, so diagnosis of EES is based on histopathology[7]. Extraosseous Ewing's sarcoma is a curable disease. The disease-free survival rate has been significantly increased by managing these tumors with aggressive surgical resection in combination with multi agent chemotherapy, with or without radiotherapy[1]. For localized ES, the treatment of choice is chemotherapy; 3-6 cycles followed by local treatment then a 6 to 10 cycles through 6 weeks intervals. The most active drugs are doxorubicin, CY clophosphamide, ifosfamide, vincristine, dactinomycine and etoposide. The most protocols are based on the combination of 4 to 6 of these drugs[6].

Generally, the ES is a very aggressive tumour with a high risk of metastasis (40% are localized in the lungs, 30% in bone) and frequent recurrences (30%)[7]. Rhythm of surveillance follow up is every 2-3 months during the first three years; once every 6 months to 5 years and at least once a year beyond 5 years[6]. In a retrospective study of 24 patients with EES, a global 5 years survival rate of 61% was seen[3]. Another study estimated a 5 years disease free survival rate of 60-70% for localized disease when treated with chemotherapy based on multiple anti-neoplastic drugs and surgical exision[7].

As this type of case reports were rare, some studies reported such case. They diagnosed the case on the basis of MRI and immunohistochemistry which supported our case[9,10].

CONCLUSION

Extra skeletal Ewing's sarcoma is an extremely rare disease. Despite It rarity, it is always necessary to consider EES in the differential diagnosis of soft tissue sarcomas for early diagnosis and therapeutic management, because its identification in localized stage is often associated with a good prognosis.

REFERENCES

1. Hwang SK, Kim DK, Park SI, Kim YH, Kim HR. Primary Ewing's Sarcoma of the Lung. Korean J Thorac Cardiovasc Surg 2014; 47(1): 47-50.

2. Shimada H, Newton WA, Soule EH, Qualman SJ, Aoyama C, Maurer HM. Pathologic features of extraosseous Ewing's sarcoma: a report from the Intergroup Rhabdomyosarcoma Study. Human pathology 1988; 19(4): 442-53.

3. Raney RB, Asmar L, Newton WA Jr, Bagwell C, Breneman JC, Crist W, et al.; Ewing's sarcoma of soft tissues in childhood: a report from the Intergroup Rhabdomyosarcoma Study, 1972 to 1991. J Clin Oncol 1997; 15(2): 574-82.

4. Kashima TG, Gamage NG, Dirksen U, Gibbons CL, Ostlere SJ, Athanasou NA. Localized Ewing

sarcoma of the tibia. Clin Sarcoma Res 2013; 3(1): 2.

5. En-Nafaa I, El Ounani F, Latib R, Jroundi L, Boujida N, Chami I. Ewing sarcoma of the anterior mediastinum: a rare tumor localization. J Radiol 2010; 91(12 Pt 1):1287-1288.
6. Granowetter L, Womer R, Devidas M, Krailo M, Wang C, Bernstein M, et al. Dose-intensified Compared with Standard Chemotherapy for Nonmetastatic Ewing Sarcoma Family of Tumors: A Children's Oncology Group Study. J Clin Oncol. 2009; 27(15): 2536-41.
7. Halefoglu AM. Extraskeletal Ewing's Sarcoma Presenting as a Posterior Mediastinal Mass. Arch Bronconeumol 2013; 49(2):82-4.
8. Geens L, Robays JV, Geert V, Speeten KV. An Unusual Location of Extraosseous Ewing's Sarcoma. Case Rep Oncol 2013; 6(2): 293-302.
9. Boufettal M, Azouz M, Rouhi AH, Mahfoud M, Bardouni AE, Berrada MS, et al. Extraosseous Ewing's sarcoma of the leg: A case report. Sch Acad J Biosci 2014; 2(8): 510-2.
10. Lai MLD, Sheu RS, Chen CY, Liu GC. Extraosseous Ewing's Sarcoma: A case report with MR manifestation. Chin J Radiol 2000; 25(4): 251-4.