

Ewing's Sarcoma of Scapula in a 12 Year Old Child- A Rare Case Report

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Abstract: Ewing's sarcoma is the second most common primary malignant bone tumour of childhood and adolescents after osteosarcoma. Ewing's sarcoma is a highly malignant small round cell tumour that arises from embryologic neural crest cells. Most frequently the sites involved are femur and tibia in lower extremity and in upper limb humerus is involved commonly. In upper limb involvement of scapula is rare. We hereby report a case of 12 year old girl with ewing sarcoma of the scapula. The treatment involves surgery (previously amputation was preferred surgery but now a day's limb sparing surgery is preferred), followed by chemotherapy to kill the malignant cells and to take care of micrometastasis. Ewing sarcoma is the second most common highly malignant primary bone tumour of childhood and adolescents after osteosarcoma, that arise from medullary cavity and invade the Haversian system of bone.

Keywords: Ewing's sarcoma, scapula, CD99

INTRODUCTION

Ewing sarcoma is a highly malignant small round cell tumour and was first described by James Ewing in 1921 as a "diffuse endothelioma of bone" [2]; previously it was believed that this tumour arises from mesenchymal stem cells [3] and these cells have the capability to convert into many tissue types of the body but now it is believed that these cells arise from embryologic neural crest cells [4]. Ewing sarcoma comprises 6-8% of the primary malignant bone tumours with most common age group of presentation is 5-25 years with males more commonly affected than females[1]. It usually involve the long bones of extremities, most common are femur, tibia in lower limb and in upper limb mainly humerus. Like any other sarcoma it mainly metastasizes by the haematogenous route involving bones, lung, and bone marrow.

CASE REPORT

A 12 year old girl presented in the orthopaedic outpatient department of KGMU with history of pain and rapidly growing swelling over back for last two months. The pain was moderate in intensity present mainly over (R) shoulder girdle region. Pain was none radiating in nature and pain was not very much relieved by medications. The General and systemic examinations were within normal limits.

On local clinical examination the swelling was about 8x5 cm in size arising from the scapula. The swelling had ill-defined margins and firm to hard in consistency. The skin overlying swelling was smooth

and shiny with few visible dilated veins. There was no distal neurovascular deficit.

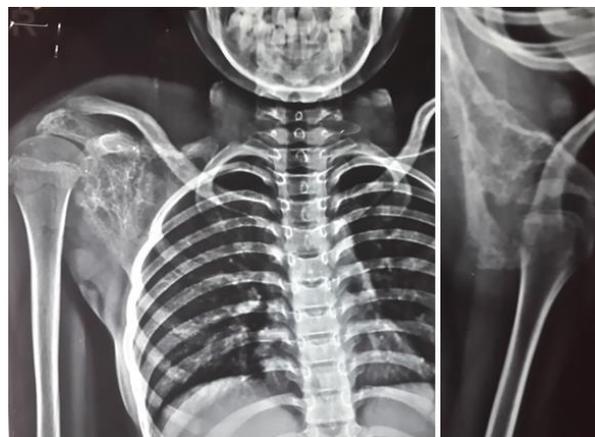


Fig-1: plain radiograph showing gross destruction of scapula with involvement of articular margins

Complete hemogram of the patient shows Hb-9.8mg/dl, TLC -14000 AND DLC (N68 L26 E3 &M3). Other blood tests including urea, creatinine, SGOT SGPT and serum calcium and serum alkaline phosphatase were normal. Plain x ray of the (R) shoulder shows osteolytic lesion involving scapular blade, acromion and spinous process with destruction of the articular margins also. After confirmation with biopsy the patient was managed by wide excision, chemotherapy and radiotherapy. The patient was discharged well but she did not returned for further follow up.

Histopathological examination report showed monomorphic small malignant cells disposed in sheets. Individual cells are monomorphic, round to oval nuclei with hyperchromatic chromatin, inconspicuous to conspicuous nucleoli and scant amount of eosinophilic cytoplasm.

Immunohistochemistry of the cells shows CD99 positive tumour cells.

DISCUSSION

Ewing sarcoma is the second most common malignant bone tumour of childhood and adolescence. The tumour consist of embryonic neural crest cells which looks small, round uniform cells with no microscopic matrix production and stains blue with H&E stain, ewing sarcoma of bone belongs to a family of tumours in which most of the tumours have a common gene translocation .The family is entitled as ewing's sarcoma family of tumours(ESFT) [4]. The translocation occurs between chromosme 11 and 22 and is referred to as t(11;22).The FLI1 gene from chromosome 11 is translocated to chromosome 22 ewing sarcoma gene(EWS) on chromosome 22 forma a complex gene called as EWS/FLI, encodes a altered gene product that lead to cancerous transformation of cells. On immunohistochemistry these cells were positive for CD99 and MIC2 [5], and negative for CD45.

Ewing sarcoma is a tumour of the childhood and adolescence with most of the cases occur between 5-15 years. It is very rare for the tumour to occur before 5 years or after 30 years of age. Males are more commonly involved and the tumour is rapidly growing and metastasis occurs early in the course of disease.

Most frequently the sites involved are femur and tibia in lower extremity and in upper limb humerus is involved commonly. In upper limb involvement of scapula is rare. The patient usually presents with pain and a rapidly growing swelling followed by systemic symptoms like weight loss and fever in some cases [6,7].

Plain radiographs show a permeative lytic lesion with wide destruction and narrow zone of transition.MRI shows significant destruction with soft tissue infiltration and usually suggestive of tumour .So, the diagnosis is confirmed by open biopsy.

The treatment involves surgery (previously amputation was preferred surgery but now a days limb sparing surgery is preferred), followed by chemotherapy to kill the malignant cells and to take care of micrometastasis. The chemotherapeutic combination preferred these days is of Ifosfamide and Etoposide. Radiation may or may not be used in

combination to these two modalities. The management should be aggressive because almost all patients with presentation of local disease have asymptomatic metastasis. With the combination of surgery (partial or total scapulectomy) and chemotherapy the five year survival rate ranges 57% to 77 % [8].

CONCLUSION

In summary the management of the Ewing's sarcoma includes neoadjuvant therapy to decrease tumour mass followed by wide excision and then followed by chemotherapy and radiation is for margin positive tumours after biopsy or for palliation from pain. The prognosis depends upon Stage of the disease, tumour size and anatomic location [9].

REFERENCES

1. Campbell's operative orthopaedics 12 th edition.
2. Kaler AK, Sheriff S; Primary ewing's sarcoma of the paranasal sinuses and orbit. *Innovative Journal of Medical and health Science*, 2013; 3(1).
3. Lin PP, Wang Y, Lozano G; Mesenchymal stem cells and the origin of Ewing's sarcoma. *Sarcoma*, 2011, 2010.
4. Tao HT, Hu Y, Wang JL, Cheng Y, Zhang X, Wang H, Zhang SJ; Extraskeletal Ewing sarcomas in late adolescence and adults: a study of 37 patients. *Asian Pacific Journal of Cancer Prevention*, 2013; 14(5): 2967-2971.
5. Shahid M, Varshney M, Maheshwari V, Mubeen A, Siddiqui MA, Julfiqar J, Gaur K; Ewing's sarcoma of scapula: a rare entity. *BMJ case reports*, 2011.
6. Askri A, Farhat LB, Ghariani B, Rabeh A, Dali N, Said W, Hendaoui L; Extraskeletal Ewing sarcoma of the abdominal wall. *Cancer Imaging*, 2008; 8(1): 156.
7. Asif N, Khan AQ, Siddiqui YS, Mustafa H; Metastasis from scapular Ewing's sarcoma presenting as sutural diastasis: An unusual presentation. *International journal of shoulder surgery*, 2010; 4(1): 18.
8. Randall L, Calvert G, Spraker H, Lessnick S; Ewing's Sarcoma Family of Tumors (ESFT). *ESUN*. Retrieved, 2009; 04-15.
9. Picci P, Böhling T, Bacci G, Ferrari S, Sangiorgi L, Mercuri M, Benassi MS; Chemotherapy-induced tumor necrosis as a prognostic factor in localized Ewing's sarcoma of the extremities. *Journal of Clinical Oncology*, 1997; 15(4): 1553-1559.