

EWING SARCOMA OF ULNA TREATED WITH RESECTION AND RECONSTRUCTION WITH FIBULA: A CASE REPORT

S Pandey¹ and S Pokharel²

¹Department of Orthopaedics, Chitwan Medical College, Chitwan Medical College (P) Ltd, Bharatpur-10, Chitwan, Nepal.

²B.P.Koirala Memorial Cancer Hospital, Bharatpur

Correspondence: Dr. Suresh Pandey, Department of Orthopaedics, Chitwan Medical College, Chitwan Medical College (P) Ltd, Bharatpur-10, Chitwan, Nepal.
e-mail: pandeys59@yahoo.com

ABSTRACT

Ewing sarcoma is the second commonest primary bony malignancy in children next to Osteosarcoma. Ewing Sarcoma of ulna extending whole of the diaphysis poses a great challenge for limb salvage surgery. It can be treated with different methods of reconstruction. Wide local excision following neoadjuvant chemotherapy followed by reconstructive procedure is the usual method in limb salvage surgery. Patient needs post operative adjuvant chemotherapy and local radiotherapy to reduce the chance of local recurrence and systemic metastasis. We present a case of Ewing sarcoma of ulna in young lady treated with limb salvage surgery-neoadjuvant chemotherapy followed by wide local excision and reconstruction with non vascularised fibular graft and post operative chemotherapy and local radiotherapy. Graft united well in 4 months with normal range of motion without any local recurrence or systemic metastasis till 1 year of follow up. There was no neurovascular and functional deficit.

Keywords: Ewing sarcoma, ulna, fibular graft

INTRODUCTION

Ewing sarcoma is fourth common primary bony malignancy and second most common in age group 10-20 years after osteosarcoma¹. It affects long bone diaphysis most commonly femur followed by humerus and tibia. Though it starts from metaphysis, it presents as diaphyseal bony malignancy. The current standard treatment schedules for resectable Ewing's sarcoma begin with neoadjuvant chemotherapy, followed by limb salvage procedure and postoperative adjuvant chemotherapy. Although amputation had been the only surgical method for several decades, limb salvage procedures, which include local resection and reconstruction, are currently performed in almost all the cases of Ewing's sarcomas. Limb salvage procedures can be performed without compromising survival rates². If imaging study shows doubtful negative surgical margin obtainable during resection, preoperative Radiotherapy is advised.

Five year survival rate has increased to 60-70% with these modality of treatments. We present a case of Ewing sarcoma of ulna involving whole of diaphysis in a young lady treated with neoadjuvant chemotherapy followed by wide local excision of ulnar diaphysis and reconstruction with non vascularised contralateral fibular graft. Patient received post operative chemotherapy and local external radiotherapy. The graft got incorporated and fracture united in 4 months and

patient is able to do her all previous level activities. There is no residual functional loss and local recurrence or systemic metastasis upto 2 year of follow up.

CASE REPORT

A 20-year-lady presented in our OPD with complain of pain and swelling Left forearm for 4 months. Pain was more at night and it was progressive. Swelling started at midforearm level and it was progressive. No constitutional symptoms were present.

On examination general condition was fair, pallor was present and systemic examination revealed no significant findings. Local examinations showed diffused circumferential ill defined bony hard tender swelling at the mid forearm level arising from the ulnar diaphysis and extending up to proximal and distal metaphysis with locally warm skin and reddish discoloration. Movement at elbow and wrist was terminally restricted and painful. There was no distal neurovascular deficit.

Plan x-ray of the Rt. Forearm with elbow and wrist showed destruction of the ulnar diaphysis with cortical and endosteal irregularity of the margin, sun ray appearance and codmans' triangle with evidence of soft tissue swelling. Hb was 9.9,

total count 13,200 and ESR 60 mm in 1st hr. Random blood sugar, urea creatinine, liver function test, serum calcium and Alkaline phosphatase were within normal limit. Chest x-ray and CT scan of the chest and skeletal survey didn't show any evidence of metastasis. Incisional biopsy showed features suggestive of Ewing Sarcoma. Patient was treated with neo-adjuvant chemotherapy consisting of cisplatin and adriamycin 3 weekly for 3cycles followed by wide excision of the tumor mass through normal tissue margin and reconstruction of gap with 15 cm long contralateral non-vascularized fibular diaphysis and fixed with reconstruction plate. Immobilization was done with above elbow pop slab for 6 weeks, Range of motion and grip exercise was started after 6 weeks. Post operative adjuvant chemotherapy was given after 3 weeks of operations for 3 cycles. Local external radiotherapy was given 6 weeks after the surgery as the margin was doubtful. Patient was followed for two years and fracture showed union in distal site at four months

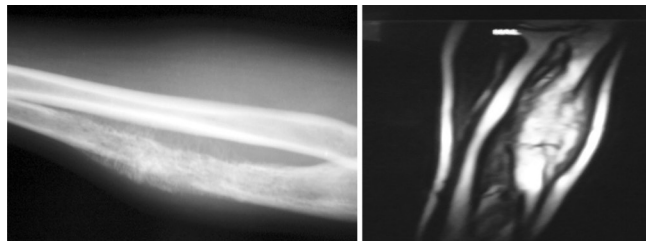


Fig I. X ray and MRI showing diaphyseal Ewing Sarcoma of Ulna

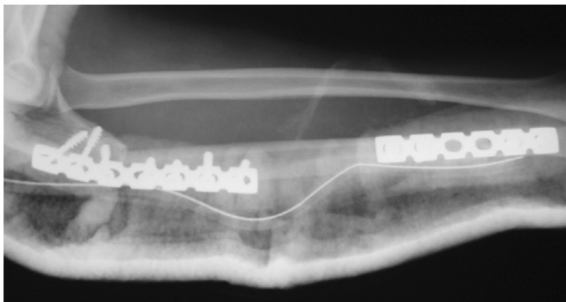


Fig II. X-ray postoperative after resection and reconstruction with fibula



Fig III. Photograph showing range of motion at 4 month follow up

and union at proximal site at one year of follow up. There is no evidence of local recurrence or systemic metastatic till the last follow up. She is asymptomatic and is able to do her previous level activities without any functional impairment.

DISCUSSION

Ewing sarcoma involving long bone is common and most common primary malignancy in less than 10 years of age. The most unfavorable prognostic factor in Ewing's sarcoma is the presence of distant metastasis at the time of diagnosis. Even with aggressive treatment, patients with metastases have only an approximately 20% chance of long-term survival. Involvement of the ulnar diaphysis with this tumor poses challenge to surgeon for limb salvage surgery. If expertise and resources are limited excision through normal plane after neo-adjuvant chemotherapy and reconstruction with non-vascularised diaphysis of the fibula is viable option. Other options for reconstructions are vascularized fibular graft which demands much more operative time, microsurgery expertise and vascular instrument sets.

Chemotherapy most commonly used to treat Ewing sarcoma regardless of their identification at initial staging includes doxorubicin (DXR), cyclophosphamide (CPA), vincristine (VCR), actinomycin-D (ACT), ifosfamide (IFM), and etoposide (VP16)³. As Ewing's sarcomas are sensitive to both chemotherapy and irradiation, even questionable candidates for limb salvage may be eligible after neoadjuvant chemotherapy with or without irradiation. If the surgical margins are found to be inadequate after surgery, postoperative radiotherapy is added. When surgical margins are certain to be inadequate at preoperative imaging, amputation is the only surgical option available.

CONCLUSION

Ewing Sarcoma of ulnar diaphysis can be treated with neoadjuvant chemotherapy followed by limb salvage surgery with resection and reconstruction with nonvascularised fibular graft and postoperative chemotherapy and radiotherapy with satisfactory result.

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