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Case Series

Ewing sarcoma: a tumor with an uncanny predilection for uncommon sites

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ABSTRACT

Ewing sarcoma is one of the most common bone tumors diagnosed in pediatric age group second only to osteosarcoma. The tumor is known to involve most commonly pelvic bones and long bones while extra-skeletal Ewing sarcoma accounts for 15%. Ewing sarcoma of other sites like scapula, clavicle, hands, and feet are quite rare, accounting for only 3-5%. To determine the epidemiological profile of Ewing sarcoma of unusual sites treated in our department including age at diagnosis, site of origin and mode of management along with survival data. Details of Ewing sarcoma patients reported our department in last 9 years were collected and data was analysed. We report a total of 20 cases of Ewing sarcoma in last 9 years with 4 (20%) extra skeletal Ewing sarcoma arising from soft tissues of extremities and 30% from rare sites (3 scapula, 1 clavicle, 1 phalanx, 1 calcaneum). All patients were operated after 4-7 cycles of chemotherapy with 66% limb salvage rate. With an average necrosis of 40%, with a median follow up of 36 months, the overall survival of the group was found to be 77.7%. Ewing sarcoma is one of the rare tumors of bone and soft tissue with the predilection for unusual sites, with reasonable survival outcomes in localized disease. Though scapula, clavicle, hands and feet are rare sites for Ewing sarcoma, they accounted for 30% in our department, possibly because of the referral patterns.

Keywords: Ewing sarcoma, Ewing sarcoma of uncommon sites, Limb salvage in Ewing sarcoma

INTRODUCTION

Ewing sarcoma is second most common primary bone tumor of paediatric age group with the incidence of 1 in a million population. The axial skeleton accounts for 54% cases while 43% are seen in appendicular skeleton. Among these, clavicle is affected in nearly 1% of cases, while scapula and hand bones are affected in 4% and 1% respectively.[1] Extra-skeletal Ewing is another subset (15%) where the disease predilection is more central and these tend to occur more in females of older age group.[2] The chromosomal translocation t (11;22) (q24; q12) is present in 90% to 95% of these tumors, which involves the chimeric fusion gene EWS/FLI-1.[3] The common presentation is usually swelling in the shaft of the bone with pain, while the disease of axial skeleton may present with radiculopathy. Ewing sarcoma of clavicle scapula, calcaneum and phalanges are quite uncommon and not much of literature is available on prognosis and management of primaries in these sites. Here we report our experience in management of these tumors.

CASES REPORT

We report a total of 20 cases of Ewing sarcoma in last 9 years with mean age of 13.9 years. Age at diagnosis ranged from 3 to 27 years. 9 of the cases were females while rest 11 were males. Among 20, 4 (20%) were extra skeletal Ewing sarcoma arising from soft tissues of extremities.

Among the 20, 10 patients presented with the tumors arising in the upper limb and 10 in the lower limb. In 50%

of the patients the tumor involved the long bones and in another 50% it originated from unusual sites (4 extra skeletal, 3 scapula, 1 clavicle, 1 phalanx, 1 calcaneum).

The 12 cases were treated with limb salvage while 6 patients ended with amputation either due to non-functional limb or due to progressive disease. The 2 scapular tumors were treated with total scapulectomy with humeral suspension, calcaneal primary with calcanectomy and allograft reconstruction, while patient with clavicular Ewing's underwent total claviculectomy. 3 Patients with tumor of soft tissue origin under went wide excision of the lesion while one was treated with amputation. The 11 patients had residual disease while 7 did not. 5 patients in residual group showed average necrosis of 40%. All patients were operated after 4-7 cycles of chemotherapy while one patient with proximal humeral Ewing sarcoma received preoperative radiation too (Table 1 and 2).

On a median follow up of 36 months, with the available data, the overall survival of the group was found to be 77.7%

Table 1: Pre-operative data.

Variables	N
Total number of cases	20
Limb salvage	12
Amputation	6
On treatment	2

Table 2: Pre-operative treatment.

Subsite	N	Treatment
Extra skeletal	4	Wide excision/ amputation
Scapula	3	Total scapulectomy with humeral suspension
Clavicle	1	total claviculectomy
Phalanx	1	-
Calcaneum	1	calcanectomy and allograft reconstruction

DISCUSSION

The Ewing sarcoma most commonly arises from long bones of extremities and pelvis while scapula, bones of hands and feet are affected considerably less often. According to Cotterill et al who conducted an analysis of prognostic factors in 975 patients from the European Intergroup co-operative Ewing's sarcoma study group, patients with axial tumors (including scapula and clavicle) do worse than those with extremity tumors. Other prognostic markers include the presence or absence of metastasis, tumor volume, response to therapy measured in terms of percentage of necrosis.[1]

The Ewings of bones of the hands and feet account to 3-5% of all tumors, among which the calcaneum is the most common.[4-6] Adkins reported 38 years' experience of Ewing sarcoma of foot where among 16 cases 3 were of calcaneum.[7] As major reconstructions are often difficult after calcanectomy many authors prefer to go for amputations to provide good functional results. For malignant tumors of calcaneum though partial and total calcanectomy without any reconstruction have been tried as an alternative to transtibial amputation, total calcenectomy with reconstruction with homologous allograft is the most common procedure reported in literature. Other options include below knee amputation, calcanectomy with autograft reconstruction, extracorporeal irradiation and reimplantation of the bone and finally custom-made prosthesis reconstruction.[8] In our institute the patient was treated with total calcanectomy and allograft reconstruction. The patient was mobilising without any difficulty by 8weeks after the procedure.

Scapula is another rare subsite affected by Ewing sarcoma. Mallik et al in their cohort study on Ewing sarcoma reported 29 cases in scapula in 20 years.[9] Like in any other site neoadjuvant chemotherapy followed by surgery is most common mode of management, a nonsurgical candidate can be offered radiotherapy following chemotherapy. Survival is marginally better with surgery in comparison with radiation, but positive margin and less than 100% necrosis are associated with worse survival.[9] The surgical options include partial or total scapulectomy depending on the extent of the disease.[10] Enneking classification of bone tumors divides scapula in to two zones S1 and S2. Tumors confined to S1 can be treated with partial scapulaectomy without reconstruction while those of S2 require glenoid reconstruction. Tumors involving both the zones will require total scapulectomy and reconstruction options include humeral suspension (flail shoulder), total endoprosthesis and bone allograft. Humeral suspension was the most commonly used reconstructive option till 1990s where the proximal humerus is simply stabilized with heavy nonabsorbable sutures or wires to the clavicle. Limited shoulder abduction and loss of shoulder contour were the shortcomings reported with this procedure.[11] With the introduction of endoprosthetics the functional results have significantly improved along with cosmetic appearance.[12] Both the patients in our institute were treated with total scapulectomy and humeral suspension. Though there was some limitation of abduction at shoulders, patients retained complete function of elbow and wrist.

Literature is again limited when Ewing sarcoma of clavicle is considered. There are few case series reported with limited number of cases. Though surgery and radiotherapy can both be considered for local control of the disease, total or partial claviculectomy are most reported in literature. Clavicle is considered to be an expendable bone like rib and fibula and the resection without reconstruction in well tolerated with good functional outcome though allograft and vascularised fibular graft have been tried in past.[13] Our patient showed good range of motion with no cosmetic disfigurement and received adjuvant radiation as soft tissue margin was close.

Ewing sarcoma of the phalanx are extremely rare with isolated case reports in the literature. Among these, proximal phalanx seems to be the most affected bone with circumferential soft tissue involvement.[14] Almost all cases in the past were treated with neoadjuvant chemotherapy followed by ray amputation of the affected finger.[15] The patient in the present series in undergoing neoadjuvant chemotherapy and is planned for ray amputation of the proximal phalanx of middle finger.

The prognosis of Ewings sarcoma is greatly affected by presence or absence of metastasis at the time of presentation. The 5-year disease free survival in nonmetastatic setting is 70-75% while that of patients with metastasis is 33%.[1] In our study the overall survival at 5 year was found to be 77.7%. The dramatic increase in the survival in the recent years is owing to the new multidrug chemotherapy schedules rather than any progress in surgical field. We could not appreciate much difference in survival of patients with rarer site affliction compared to those of long bones and pelvis, probably due to the fewer number of cases in the study, though literature does not through much light in this scenario. Retrospective multicentre analysis are needed to understand the behaviour of the tumor in these rare subsites.

CONCLUSION

Ewing sarcoma is one of the rare tumors of bone and soft tissue with the predilection for unusual sites, with reasonable survival outcomes in localized disease. Though scapula, clavicle, hands and feet are rare sites for Ewing sarcoma, they accounted for 30% in our department, possibly because of the referral patterns.

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