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Ewing's Sarcoma in a Rural Medical College Hospital in West Bengal- A Retrospective Study

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Abstract
Objective: *To outline the epidemiological profile and prognostic factors for Ewing's sarcoma in a rural medical college in hilly area in West Bengal*
Material and Methods: *The medical records of 27 patients with Ewing's sarcoma who were treated at North Bengal Medical College, Darjeeling in West Bengal in India from January 2010 to December 2017, were retrospectively evaluated.*
Results: *Out of the the total 27 patients, commonest age group was 8-16 years. Male to female ratio was 1.25:1. Most of the patients presented with pain, swelling and alteration in gait. Involvement of appendicular skeleton particularly femur was commonest. Many patients presented with metastases. Lung was commonest site of metastases. Patients who presented at younger age, had involvement of peripheral skeleton, less bulky disease and underwent surgery along with chemotherapy and Radiotherapy did better with treatment.*
Conclusion: *The prognosis in cases of Ewing's sarcoma was mainly influenced by the presence of metastases at the time of diagnosis, involvement of peripheral (appendicular) skeleton, disease bulk and age.*
Keywords: *Ewings Sarcoma; Metastases; Prognosis.*

Introduction

Ewing's sarcoma is a malignant disease (named after James Ewing who described it in 1921) affects mainly adolescents and pediatric age group. It is the third commonest primary malignant bone tumor, after multiple myeloma and osteosarcoma. Ewing's sarcoma is 2nd most common childhood primary bone tumor and comprising of 3% of paediatric cancers.[1-2] The incidence is 3 cases per 1 million US population/yr. The affected age is mostly in 5 – 25 yrs and incidence in males is more than females (M:F=1.5-2:1). It is rare in Asian/ African children.

The management involves early diagnosis substantiated by exact immunohistology and then treatment by surgery, chemotherapy and radiotherapy. However in spite of extensive

treatment by surgery, radiotherapy and chemotherapy sometimes there are failures and disease progression[3]. Moreover, in a peripheral rural hospital set up often it is difficult to proceed with extensive chemotherapy and state of the art radiation and surgery. Though Ewing's Sarcoma is mainly bone tumor, it may be extra-osseous. It is rare and more virulent in nature.[4]

ESFT (Ewing's Sarcoma family of tumors) is a general term used to describe the tumors[5] which share the same histochemical staining profile and a unique nonrandom translocation and include:

- Ewing's Sarcoma of bones
- Extra osseous ES (ES of soft tissue)
- Peripheral PNET (primitive neuro ectodermal Tumor of bone & soft tissue) /differentiated ES

This was a retrospective review of patients of Ewing's Sarcoma who attended Radiotherapy department of North Bengal Medical College, Darjeeling in West Bengal from January2010 to December, 2017 to evaluate the outcome of treatment and identify the prognostic factors .

Material and Methods

Study period- Jan 2010- December 2017; Radiotherapy department, North Bengal Medical College (NBMC)

Study design- retrospective

Total no of patients- 27 histological proven Ewing's sarcoma

- **Inclusion criteria**

1. Biopsy proven Ewings sarcoma/PNET
2. Treatment naïve

- **Exclusion criteria**

1. Biopsy inconclusive- like use of terms poorly differentiated, small round cell, malignant tumor
2. Any previous malignancy – metachronous / synchronous.
1. Patients not fit for any cancer treatment

All the medical files were reviewed to seek data on sex, age, staging, clinical signs and symptoms, treatment procedures, location of the lesions, and treatment results.

Results

The series included 27 patients. The age range was 2yrs – 30 years. The male: female ratio was 1.25:1 (Table1) Majority of the cases were in the age group of 8-16yrs age (Table 2).

Table 1- Distribution according to sex

Male	15	55.56%
Female	12	44.44%

Table 2- Distribution according to age group

Age group(Years)	No.	Percentage
Less than 8	4	14.82
8-16	14	51.85
16-24	8	29.63
More than 24	1	3.70

The presenting features consisted of mainly pain, swelling and alteration in gait. Out of 27 cases 1 patient was of extraosseous variety. However of the osseous variety majority were in the appendicular skeletons (18 patients). Commonest site of involvement was femur (30%) followed by humerus (12%).Axial skeleton was involved in 8 patients.(Table3)

Table 3- Distribution according to site

site	No.	percentage
Axial skeleton	8	29.63
Appendicular skeleton	18	66.67
Extraosseous	1	3.7

10 patients presented with metastases from beginning. 6 more patients developed metastases after or during treatment. So, total metastatic disease was 59.26% Lung metastases was commonest (43.75%) (Table 4). Patients with bulky disease particularly of axial skeleton progressed mostly.

Table 4- Distribution of site of metastases

Site of metastases	Total metastatic patients (16)	Percentage
Lung only	7	43.75
Bone only	3	18.75
Liver only	2	12.5
Others and multiple sites	4	25

13 patients progressed during treatment and surgery could not be undertaken. Commonest surgery undertaken was removal of the tumor. 24 patients were administered radiotherapy in

combination with other modalities. All patients received chemotherapy.

After treatment, 9 (33.33%) patients had complete remission; 7 patients (25.93%) had partial remission, 11patients (40.74%) had progressive disease. (Figure 5). Lungs were the most common site of metastasis (Figure 6). At the end of period of analysis only 9 patients survived (33.33%).All these 9 patients were age below 16 years, and disease confined to appendicular skeletons and surgery was possible along with Radiation or chemotherapy.

Discussion

This study showed that patients with younger age and peripheral skeleton involvement did better than old age and pelvic bone and other axial skeleton involvement. Similar data was seen in other study[3]. Metastases was the most important factor which affected survival like other study[6]. The overall percentage of patients developing metastatic disease was 59.26% in our study. Lung was commonest site of metastases (43.75%). Our result was similar to IESS2 study[7]. Exta-0sseous variety was seen only in one patient (3.7%) with relatively older age. They showed poor response to treatment. It was similar to other study [4]. Bulky tumors had poor outcome irrespective of site. Simlar result was seen in Cooperative Ewing's Sarcoma Study.[8] In our study all patients irrespective of site of metastases had poor outcome. This was in contrast to the study conducted in Europe by Cotterrill et al[9] where it was shown that presence of distant metastasis like in lungs do better than other sites of metastasis. Overall, treatment result was poor than other study. In this study, none of the metastatic disease survived more than 5 years. Only 9 patients (33.33%) who survived more than 5yrs were non-metastatic. But in study by Sari N et al[10], for non-metastatic disease, the rates of 5-year overall survival (OS) was 51% and for metastatic disease it was 27%. This may be due to lack of proper infrastructural set-up in this rural medical college.

Conflict of Interest- Nil

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