Original Article

# An Epidemiologic Study of Ewing Sarcoma Family at SHAFA Hospital in Khozestan Province-IRAN, a Referral Children Cancer Treatment Center

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## ABSTRACT

### Background

Ewing Sarcoma is the second most frequent primary bone cancer, following Osteosarcoma in children. These tumors consist of small, round, or oval cells, which are believed to derive from parasympathetic autonomic nervous system. The common clinical presentations are pain, local tenderness, fever, palpable mass, and pathologic fractures.

### Methods and Materials

This study describes 47 children affected by Ewing Sarcoma registered since 1991 to 2007. All data were extracted from hospital admission notes and outpatient clinical records.

### Results

Twenty seven patients were male and 20 were female. It is slightly more common in boys. Pain, Local tenderness, fever, and Pathologic fractures were common presentation. Majority of patients were between 10-15 years old.

### Conclusion

Local tenderness, pain, and limping are important symptoms in children that should be check-up for immediate diagnosis and also proper management.

### Key words

Sarcoma, Ewing, bone cancer, children

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## Introduction

Ewing sarcoma is the second most frequent primary bone cancer, following osteosarcoma. Eighty percent of affected patients are younger than 20 years old. Ewing sarcoma is an uncommon primary sarcoma of osseous origin that usually arises in children and adolescents (1). It is an undifferentiated bone sarcoma. It represents approximately 1% of all cancers reported in children but approximately 30% of all bone tumors in them (2, 3). The frequency of Ewing sarcoma in the population younger than 20 years old is approximately 2.9 per million (1). It is slightly more common in males (1- 4). Cases usually diagnosed before 20 years old and a few number before 30 or less after (1, 4). It is a small round cell tumor (5). Because of a poorly

differentiated or undifferentiated histology, it may be confused with other undifferentiated round cell tumors of childhood (3, 5, 6). Ewing Sarcoma family tumors (ESFTs) are consist of a group of round, small, and undifferentiated cells, which mostly (85%) associated with reciprocal chromosomal translocation between chromosome 11 and 22, t(11;22)(q24;q12) (1, 2, 4, 5, 7). Classic Ewing sarcoma, atypical Ewing sarcoma, and peripheral primitive neuroectodermal tumors (PNETs) are derived from postganglionic parasympathetic primordial cells, which located through the parasympathetic autonomic nervous system (2, 3, 5, 7). ESFTs usually occur in bones (1), and opposite to Osteosarcoma, flat bones of axial skeleton are more commonly affected (1-5). Obvious Metastasis to lungs, skeletal system, and bone marrow or combinations is seen in 25% of patients at diagnosis (1). Common clinical findings are local tenderness and/or mass (1, 3, 5). The prognosis of those with metastatic or recurrent disease has improved very little over the past three decades (8).

## Methods and Materials

This study describes all pathologically diagnosed children with Ewing sarcoma since 1991 to 2007. These patients were referred to Shafa hospital, a referral center of children cancer management in southwest of Iran. During these 16 years, Ewing sarcoma family tumors were diagnosed in 47 patients using histophatology and immunohistochemistry methods. Information was obtained from the patients hospital records, and they classified in 4 age groups. Clinical features at the time of diagnosis, primary tumor site, and metastatic site(s) were compared between the age groups using MINITAB 14 software.

## Results

Of the 47 referred patients, 27 were male (57%) and 20 were female (43%). Twenty three patients (49%) were between 10-15 years old. Thirty seven patients had classic Ewing sarcoma (79%) and 10 had PNETs (21%).

Table1. Age distribution

| Age (year) | 0-<5 | 5-<10 | 10-<15 | >15 | Sum |
|---|---|---|---|---|---|
| No | 8 | 15 | 23 | 1 | 47 |
| Percent | 17% | 32% | 49% | 2% | 100 |

Patients mostly aged between 10 to 15 years old.

The primary tumors of 18 patients (38%) were found in lower limbs. Tibia- Fibula region was involved in 11 patients (23.5%) and femoral bone involved in 7 patients (14.5%). Chest wall involved in 9 patients (19%), which were in ribs (9%), scapula (9%), and clavicle (2%).
Pelvic bones were primary tumor site in 8 patients (17%). four patients (9%) were presented with spinal cord compression symptoms. Humerus bone involvement was seen in one patient (2%). Skull bones were affected in 4 patients (9%). However, some unusual primary tumor sites were found; retroperitoneal (2 cases), ovarian (one case), and cervical soft tissue mass (one case). Clinical finding were local tenderness (68%), palpable mass (62%), fever (13%), limping (11%), pathologic fractures (9%), dyspnea (9%), weight loss (7%), and abdominal mass (7%). Nine patients (19%) suffer from metastasis at the time of diagnosis. The most common metastatic sites were lungs (13%), skeletal system (9%), and bone marrow (2%).

## Discussion

ESFT can arise in almost every age group, but more than half of the patients are adolescents, the median age being 15 years (1). Opposite to osteosacoma, flat bones of axial skeleton and long bones were the most commonly affected sites. ESFT, unlike osteosarcoma, tends to arise from the diaphyseal rather than the metaphyseal part (1, 3). The most common sites of primary ESFT are the pelvic bones, the long bones of the lower extremities, and the bones of the chest wall (1, 2, 5). Primary metastases in lungs, bones, bone marrows, or combinations of there are detectable in about 25% of patients (3, 9). ESFTs are composed of small to large, round to oval irregular cells (7). Symptoms and signs of the tumor depend on the tumor size and location (7-9). Present study showed that 98% of the patients were younger than 15 years old, and Male/Female ratio was 1.3 to 1. Local mass with or without pain were found 68% and 62% respectively. Lower limbs (38%) and chest wall (19%) were the most common primary tumor sites in this study. Primary pelvic bone tumors were found in 17% of patients, which was lower than previous reports (1, 2, 6, 8, 9). Scapula (9%) and skull bones (9%) were primary site, which was higher than previous reports (1, 4-7, 9). Lungs were the most common site of metastatic involvement at the time of the diagnosis in present study.

In conclusion, Ewing sarcoma is a highly malignant tumor with a great propensity for metastatic spread before diagnosis. Special attention needs for local tenderness, local pain, and limping in children, because early diagnosis and treatment of ESFTs improve general outcomes

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