

## A Rare Case Report of Ewing Sarcoma Right Proximal Phalanx of Big Toe

Dr. Anoop S<sup>1</sup>; Dr. Abhishek Shetty<sup>2</sup>; Dr Nithin Chandra Babu<sup>3</sup>

<sup>1</sup> junior Resident, Department Of Orthopaedics Yenepoya Medical College Hospital University Road, Deralakatte Mangalore- 575018 Karnataka, India

<sup>2</sup> asst. Professor, Department Of Orthopaedics Yenepoya Medical College Hospital University Road, Deralakatte Mangalore- 575018 Karnataka, India

<sup>3</sup> junior Resident, Department Of Orthopaedics Yenepoya Medical College Hospital University Road, Deralakatte Mangalore- 575018 Karnataka, India

---

### Abstract

**Introduction:** Ewing's sarcoma localized to foot is extremely rare. Local control is mandatory because of the aggressive nature of the tumor. Therapeutic options for these local tumors include neoadjuvant chemotherapy followed by surgery.

**Case Presentation:** A 16-year-old male patient presented with 8-month history of progressive swelling and intermittent pain of the right great toe. Plain radiography and magnetic resonance imaging revealed an expansile mass that had originated from the proximal phalanx of the great toe and was destructing and surrounding the distal phalanx. Distant metastasis was noted in the T 12 vertebra. The patient underwent a closed biopsy, which confirmed the diagnosis of Ewing's sarcoma. He was treated with neoadjuvant chemotherapy and radiotherapy. Surgical excision of tumor was advised but patient party was not willing for surgical management.

**Conclusion:** The distal phalanx of the foot is an extremely rare site for the development of Ewing's sarcoma. As local control is important to avoid dissemination of the disease, neoadjuvant chemotherapy followed by amputation or disarticulation of the affected digit and subsequently adjuvant chemotherapy may be favorable modality for increasing the patient's duration of survival.

**Keywords:** Ewing's sarcoma, foot, proximal phalanx, disarticulation.

---

### I. Introduction

Ewing's sarcoma (ES) is the second most common primary malignant bone cancer, after osteosarcoma, and is usually diagnosed during the second decade of life [1]. This aggressive sarcoma is composed of small round blue cells and has a high potential for distant metastasis. Although ES can arise in the soft tissue, it most commonly originates from the bone, especially the appendicular skeleton. The most frequently affected bones are the femur (22%), tibia (11%), and fibula (9%). The short bones of the foot or hand are affected in only 3-5% of cases [1]. Primary malignant bone tumors of the foot are extremely rare, and a phalangeal location is considered exceptionally rare [2]. We report a case of ES located at the proximal phalanx of the foot that was treated by chemotherapy and radiotherapy, followed by surgically with disarticulation at the metatarsophalangeal joint.

### II. Case Report

A 16-year-old male patient presented with 8-month history of progressive swelling and intermittent pain of the right great toe (Fig 1). The pain was slightly responsive to analgesics. The patient had an unremarkable medical history, except for a prior history of trauma to the toe, which had occurred immediately before the complaints that lead him to seek medical attention. He was referred to our clinic after the detection of a tumoral lesion in his right foot, which had been observed on radiography. Anteroposterior and lateral radiographs of the left foot showed an aggressive lytic lesion with cortical erosion (Fig 2). The lesion was located at the proximal phalanx of the great toe. Magnetic resonance imaging (MRI) of the left foot revealed an expansile mass that had originated from the proximal phalanx of the great toe, destructing and surrounding the bone (Fig 3). MRI confirmed that the flexor and extensor tendons were affected. Wholebody bone scintigraphy demonstrated increased uptake at the right great toe and T 10 vertebrae (Fig 4). The patient underwent a closed biopsy. Histological examination showed small, round cells (indicating ES) (Fig 5). He was treated with neoadjuvant chemotherapy and radiotherapy. Surgical excision of tumor was advised but patient party was not willing for surgical management.

### III. Discussion

ES is a primary malignant bone tumor that usually involves the diaphyseal portion of long bones. ES rarely affects the short bones of the hands and feet; only a few cases of solitary lesions that involve these bones have been reported. ES located in the foot is associated with a substantial delay in diagnosis (median delay of 18 months), as compared with ES located elsewhere [2]. Primary malignant bone tumors of the foot are extremely rare. The lytic lesions of ES initially manifest symptomatically as pain, swelling, and sometimes fever (especially in metastatic disease) [3]. Consequently, a large proportion of patients with ES is initially diagnosed with tendinitis or osteomyelitis and is treated accordingly [1, 4]. Therefore, early imaging studies should be prompted by pain without defined trauma that is sufficient to explain the symptoms, pain lasting longer than a month, pain continuing at night, and pain with any other unusual features [1]. In the reported case, the patient's history was unremarkable except for a minor suspicious trauma, and his pain was partially responsive to analgesics. Our patient had an 8-month delay between symptom onset and his referral to our hospital. Clinicians should remember that patients with sarcoma might relate the onset of symptoms to minor trauma occurring around the same time [4]. Clinicians usually perform clinical imaging when the patient does not improve after receiving antibiotics and/or analgesics. When initial radiological imaging in two planes reveals tumor-related osteolysis and periosteal reactions, a diagnosis of primary malignant tumor is suggested [5]. However, MRI provides the most precise definition of the local extent of disease as well as the relation of the lesion to adjacent blood vessels and nerves [5]. Additionally, MRI is widely used to assess responses to neoadjuvant chemotherapy. Indeed, it was used for this purpose in the present case. Radiologic imaging alone is not sufficient for the diagnosis of ES; the diagnosis should be confirmed histopathologically. Tumor specimens are most optimally obtained through incision open biopsy performed by an experienced orthopaedic surgeon [5]. There are no specific laboratory tests for ES. However, elevated ESR, moderate anaemia, leukocytosis, or elevated levels of LDH may be detected in cases of ES [1].

Modern multimodal therapeutic regimens for ES include a combination of chemotherapy, surgery, and radiotherapy [1, 3, and 6]. Radiotherapy and surgery are the main forms of treatment that are used to achieve local control of ES. Therefore, the choice between treatment modalities for the control of the primary lesion should be made on an individual basis [3]. Retrospective analyses by several groups have provided the impression that surgery is preferable when local control is possible [7]. Our patient underwent neoadjuvant chemotherapy and radiotherapy. Surgical excision of tumor was advised but patient party was not willing for surgical management.

The use of surgery for these lesions remains controversial. Many authors have stated that radical surgical treatment is associated with lower recurrence rate and longer duration of survival [2, 8]. Although amputation was the only available surgical method for several decades, limb salvage procedures (such as wide resection and biological reconstruction) are currently performed in most reported cases of ES [5]. Furthermore, it has been shown that limb salvage procedures can be performed without compromising survival rates [7, 9]. The current standard treatment schedules for resectable ES begin with neoadjuvant chemotherapy followed by a limb salvage procedure and postoperative adjuvant chemotherapy. In the case reported here, after the appropriate neoadjuvant chemotherapy and radiotherapy we planned for amputation of 1<sup>st</sup> metatarsophalangeal joint, but they denied the procedure.

### References

- [1]. Bernstein M, Kovar H, Paulussen M, Randall RL, Schuck A, Teot LA, et al. Ewing's sarcoma family of tumors: current management. *Oncologist*. 2006; 11: 503-519.
- [2]. Brotzmann M, Hefti F, Baumhoer D, Krieg AH. Do malignant bone tumors of the foot have a different biological behavior than sarcomas at other skeletal sites? *Sarcoma*. 2013; 2013: 767960.
- [3]. Rammal H, Ghanem I, Torbey PH, Dagher F, Kharrat K. Multifocal Ewing sarcoma of the foot. *J Pediatr Hematol Oncol*. 2008; 30: 298-300.
- [4]. Widhe B, Widhe T. Initial symptoms and clinical features in osteosarcoma and Ewing sarcoma. *J Bone Joint Surg Am*. 2000; 82: 667-674.
- [5]. Iwamoto Y. Diagnosis and treatment of Ewing's sarcoma. *Jpn J Clin Oncol*. 2007; 37: 79-89.
- [6]. Adkins CD, Kitaoka HB, Seidl RK, Pritchard DJ. Ewing's sarcoma of the foot. *Clin Orthop Relat Res*. 1997; 343: 173-82.
- [7]. Schuck A, Ahrens S, Paulussen M, Kuhlen M, Konemann S, Rube C, et al. Local therapy in localized Ewing tumors: results of 1058 patients treated in the CESS 81, CESS 86, and EICISS 92 trials. *Int J Radiat Oncol Biol Phys*. 2003; 55: 168-177.
- [8]. Casadei R, Magnani M, Biagini R, Mercuri M. Prognostic factors in Ewing's sarcoma of the foot. *Clin Orthop Relat Res*. 2004; 420: 230-238.
- [9]. Bacci G, Ferrari S, Bertoni F, Rimondini S, Longhi A, Bacchini P, et al. Prognostic factors in nonmetastatic Ewing's sarcoma of bone treated with adjuvant chemotherapy: analysis of 359 patients at the Istituto Ortopedico Rizzoli. *J Clin Oncol*. 2000; 18: 4-11.

### Figure legends

**Figure 1.** Clinical pictures of right big toe

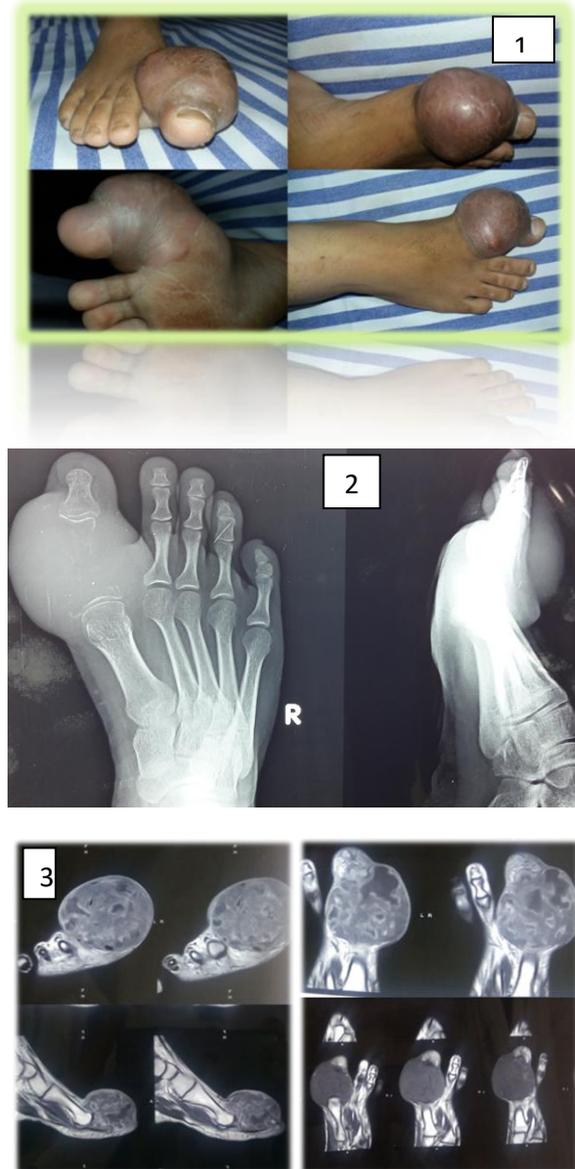
**Figure 2.** Anteroposterior and lateral radiographs showing a lytic and destructive lesion in the proximal phalanx of the great toe bone.

**Figure 3.** T1 and T2 sequence, coronal magnetic resonance imaging (MRI) sections of the proximal phalanx and soft tissue components of the tumor. Destruction of the interphalangeal fat plane can be observed on the lateral side of the tumor. T1 sequences of the axial MRI section of the proximal phalanx, showing the soft tissue component of the tumor.

**Figure 4.** Whole-body bone scintigraphy showing focally increased activity in the right great toe and T 12 vertebrae.

**Figure 5.** Histopathological report

**Figures**



*A Rare Case Report Of Ewing Sarcoma Right Proximal Phalanx Of Big Toe*

