

Ewing's Sarcoma Of The Distal Phalanx Of Little Finger

J Jose Jerome, M Varghese, B Sankaran, Thirumagal

Citation

J Jose Jerome, M Varghese, B Sankaran, Thirumagal. *Ewing's Sarcoma Of The Distal Phalanx Of Little Finger*. The Internet Journal of Oncology. 2006 Volume 4 Number 1.

Abstract

Ewing's tumor in the hand is extraordinarily rare. Lesions in the hand are prone to metastasize, but have an excellent prognosis with respect to survival rate and morbidity. Primary radiotherapy or amputation with chemotherapy is the therapeutic regimen for distal extremities lesions. The differential diagnosis includes pulp abscess, glomus tumor, osteomyelitis, tuberculosis, benign tumors, and tumorous conditions in the hand. Clinical findings, radiographs and excisional biopsy are essential to confirm the diagnosis. We report a case of Ewing's tumor of distal phalanx of the right fifth digit.

CASE REPORT

A 13-year-old girl came to our outpatient department with complaints of pain and swelling of the right little finger of one-month duration. The swelling was gradually increasing in size. She did not report systemic symptoms such as fever, malaise, weight loss, or night sweats. She did not give any history of trauma. On examination, there was a diffuse tender, warm, cystic swelling in the tip of the finger. The skin over the swelling was red, with induration at the tip of little finger. The lesion was thought to be benign until the biopsy results returned (Fig 1).

Figure 1

Figure 1: Tip of the little finger with a red indurated cystic swelling



Radiographs showed a permeative pattern of destruction of

both distal phalanx and a part of middle phalanx. A soft tissue mass, sclerosis of the tip of the distal phalanx, a lytic lesion in the head of middle phalanx, and osteopenia were also noted (Fig 2). There was no periosteal reaction. A chest radiograph was normal at the time of the first visit.

Figure 2

Figure 2: radiographs show soft tissue permeation, sclerosis and lytic lesions in the distal phalanx and the head of middle phalanx



A fine needle aspiration cytology was inconclusive. Blood stained fluid was aspirated from the cystic swelling. Microscopic examination and culture for aerobic, anaerobic, acid fast and fungal organisms were negative. Complete blood count examination showed a total leukocyte count of 11,200 cells/mm³. Hemoglobin and other parameters were normal. Erythrocyte sedimentation rate was 24mm/hr (Normal <15).

An excisional biopsy with amputation of the distal and middle phalanx was done under local anesthesia. The mixture of well-preserved tumor cells and smaller cells with hyper chromatic pyknotic nuclei, characteristic of Ewing's sarcoma, was designated as cellular dimorphism (Fig 3). Periodic acid Schiff stain was positive indicating abundant glycogen. Reticulum stain was negative.

Figure 3

Figure 3a

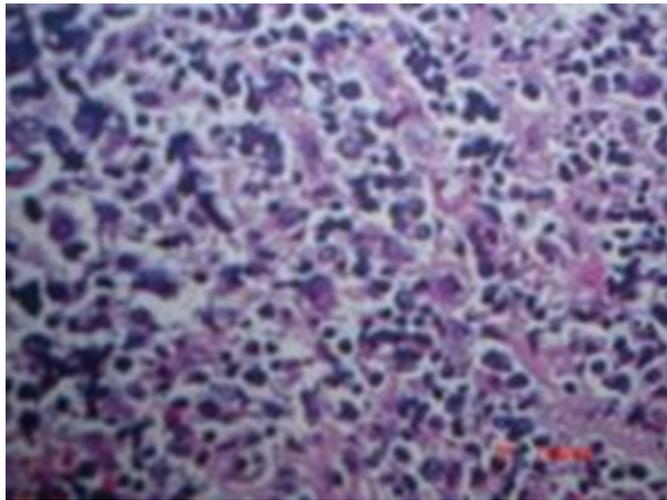


Figure 4

Figure 3b

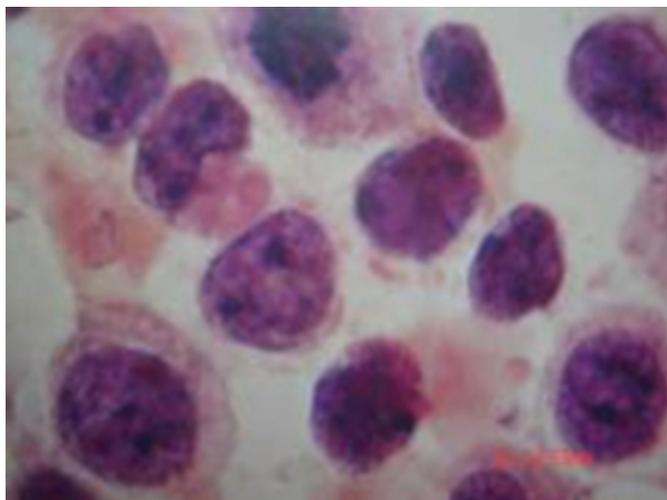


Fig 3a shows cellular characteristics of Ewing's sarcoma. There is a prominent dimorphic light and dark cell pattern associated with hemorrhage and necrosis. Fig 3b shows round nucleus with lightly stippled chromatin and a scant rim of cytoplasm. (H and E stain).

A combination of cyclophosphamide, dactinomycin and vincristine was given to the patient in the postoperative

period^{1,2,3}. The patient was thoroughly investigated for metastases. A computerized tomography of the chest was normal. The local skin condition around the stump was also normal. Radiotherapy was not given to the patient because of its associated complications like radiation induced sarcoma^{6,11,12} and the possibility of epiphyseal damage^{11,12}. The patient is on regular follow up 24 months after the diagnosis with no complaints.

DISCUSSION

A review of Ewing's sarcoma in the hands and feet reported in large series published since 1950 reveals 48 cases, with an incidence of 3.5%^{5,6,7,8,9}. Hand lesions are extraordinarily rare^{13,14,15,16}. Pain and swelling of the affected fingers are the most frequent complaints at diagnosis. The lesion is thought to be benign until the time of biopsy⁴.

Classically systemic symptoms such as fever, malaise, weight loss, or night sweats are present, and the condition is usually mistaken for a local infection. The duration of local symptoms before diagnosis ranges from 2 to 12 months. The clinical presentation is not distinctive.

Roentgenographically, as typical seen with Ewing's sarcoma, permeation^{17,18} is the most common findings. Sclerosis, soft mass, cystic change was surprisingly common while periosteal reaction was remarkably uncommon, compared with other reports of Ewing's sarcoma in more general populations^{4,17,18}. The general population include IESS protocols No 7299 (373 patients) and No. 7450 (61 patients). The latter (periosteal reaction was remarkably uncommon) may be due to the fact that periosteal reactions from other causes is usually less marked in bones of the hands and feet than in many other bones of the body.(S.K Shirley e al 1985)

An Intergroup Ewing's Sarcoma Study showed that prognostic factors with respect to histopathology were topographic pattern and mitotic rate. Our patient had a predominant diffuse pattern, which correlates with a better survival rate³. Two-cell dimorphism was seen in most of the patients. The cell type was 'large clear' and 'very large' corresponding to the atypical Ewing's sarcoma of Nascimento and Dahlin. Glycogen presence remained in the gray zone, while mitotic counts were generally <1 per 10 high powers (40X objective fields (HPF)). Reticulin was patchy and found only around blood vessels. Necrosis was also seen in most of the patients. Additional diagnoses other than Ewing's sarcoma included infective etiology, poorly differentiated chondrosarcoma, chondroblastic sarcoma,

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mesenchymal chondrosarcoma, pseudo-Ewing's sarcoma, and lipoblastic type⁴.

Patients with hand lesions have survived for more than 41 months. This is median value the follow up of survivors ranged from 36 to 85 months after diagnosis. (S.K.Shirely et al 1985)The current Intergroup Ewing's Sarcoma Study data showed a 68% over all three-year survival rates in patients with distal extremity lesions^{2,3}. Patients who had metastatic disease in the lung at the time of initial diagnosis were, however noted to clear over a period of course of 1,800-rad radiotherapy. This was over the lungs which was usually noted to clear over few cycles (usually 19 days course) of 1,800-rad radiotherapy. Refractory cardiac failure attributed to drug cardiotoxicity, diffuse parenchymal consolidation consistent with shock lung and pulmonary infarct were the complications noted. No microscopic tumor was identified⁴. The lungs did not showed any diffuse parenchymal consolidation (in CT chest) or pulmonary infarct. Our patient had no microscopic tumor.

Figure 5

Table 1: Differential Diagnosis

Felon	Penetrating injury, redness, cellulites, abscess, osteitis
TB dactylitis	Short tubular bones, spina vertosa, subperiosteal new bone formation.
Enchondromas	Most common destructive primary bone tumors. Proximal metaphysis of the proximal phalanx, eccentric and expansile. Pathological fracture .
Aneurysmal bone cysts	Eccentric ballooning lesions (metaphyseal, not epiphyseal) ,pain and limitation of motion.
Glomus tumors	<1 cm in diameter, deep red or purple discoloration, beneath the fingernail (25%-65%). Pain, cold sensitivity and point tenderness
Osteoid osteoma	Pain, usually worse at night, drastic pain relief by aspirin. Carpal bones with a small oval or round sclerotic nidus surrounded by first an area of less dense bone, like a halo, and then by an area of sclerotic.

Other small cell malignant tumors (e.g. rhabdomyosarcoma) can have a similar histological appearance and can be PAS positive. Rhabdomyosarcomas are relatively common in the hand¹⁰.

Proximal phalanx involvement in Ewing's sarcoma, and fatal cardiac metastasizes⁸ were reported in the literature. The distal phalngeal involvement and absent metastasis is rarely reported in the literature.

Ewing's sarcoma of the hand has excellent prognosis. Although we are dealing with a rare presentation of Ewing's sarcoma of distal phalanx, this feature together with accessibility to amputation raises the question of radiation v surgery for primary control of this lesion. Lesion in any of this location may, however, metastasize, and the importance of adjuvant chemotherapy cannot be overemphasized.

We feel it is important to include Ewing's sarcoma in the differential diagnosis of unusual finger swelling without distinctive systemic symptoms. Histopathology confirms the diagnosis. Chemotherapy and/or radiotherapy remain the mainstay of treatment. Fusion gene detection is needed in such a rare case of Ewing's sarcoma, which is the future promising diagnostic and confirmative investigating tool.

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Author Information

J. Terrence Jose Jerome, M.B.B.S.

Post graduate in orthopedics, Department of Orthopedics, St Stephen's Hospital

Mathew Varghese, M.S. (ortho)

Head, professor, Department of Orthopedics, St Stephen's Hospital

Balu Sankaran, FRCS(C), FAMS

Professor Emeritus, Orthopedics, St Stephen's Hospital

Thirumagal, MBBS, MD