Case Report

Ewing’s sarcoma of fifth metacarpal with lung metastasis in a child: an unusual presentation

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ABSTRACT

Ewing’s sarcoma is the second most common bone tumour second only to osteosarcoma which represents a family of malignancies of neuroendothelial origin, which are highly aggressive and poorly differentiated. The most frequent sites involved are shaft of long bones such as femur, tibia and flat bones such as pelvis and vertebra. Ewing’s sarcoma of small bones of hands is very rare and distant metastasis with hands as the primary is even rarer. Here we describe the course of a 11-year-old male who presented with ewings sarcoma of fifth metacarpal who presented with lung metastasis without local recurrence of the disease after primary surgery and later succumbed to his illness.

Keywords: Ewing’s sarcoma, Metacarpal, Tumour

INTRODUCTION

Ewing's sarcoma (ES) is a malignant nonosteogenic primary tumour of the bone which was first described in 1921 by James Ewing as a tumour arising from undifferentiated osseous mesenchymal cells.¹ ES involves diaphysis of the long bones and the flat bones of the pelvis in young patients more commonly while it is relatively uncommon in the small bones of hands and feet with 1% incidence.²

CASER REPORT

A 11-year-old male was admitted to our hospital with pain and swelling of ulnar border and adjoining areas of right hand which was diagnosed after histopathological examination as a case of ES of fifth metacarpal bone of right hand. MRI right hand, CT Chest, bone scan was done to rule out distant metastasis. At the time of presentation, it was a localized disease and wide local excision of the mass was done without local reconstruction (Figure 1).

Figure 1: Post surgery X-ray hand showing excised fifth metacarpal bone along with tumour mass.
The surgical margins were negative. Patient was advised four drug chemotherapy VACD regime (vincristine, actinomycin, cyclophosphamide and doxorubicin) after surgery which he abandoned after 2 cycles.

He returned to us 9 months later with fever, cough, dyspnea and tachypnea. Blood investigations revealed low haemoglobin, leucocytosis and raised erythrocyte sedimentation rate (ESR). Chest X-ray showed massive right sided pleural effusion with mediastinal shift to left side (Figure 2).

**Figure 2: X-ray chest showing massive pleural effusion.**

Aspiration of pleural fluid was done which was sent for examination. Radiographs and MRI scan of right hand revealed no significant signs indicating presence of local recurrence of disease but CT scan of the chest revealed large homogenous mass suggesting lung metastasis (Figure 3).

**Figure 3: CT chest showing lung metastasis.**

Tc 99 whole-body bone scan was done but no other site of metastasis was found. Palliative chemotherapy (VACD regime) and supportive treatment was started. After 10 months of follow-up, he succumbed to his illness.

**DISCUSSION**

ES of the hand is very rare and metacarpal bones are most commonly affected.\(^1\)\(^2\) ES in the hand has a generally better prognosis compared to other sites.\(^3\) Survival rate is highest in lesions involving distal bones of the extremities.\(^4\)

ES can be easily confused with acute infection of bone presenting with pain, swelling, localized tenderness with increased total leukocyte count and raised ESR.\(^5\) ES can also present with atypical radiological appearance in hand involvement mimicking enchondroma, osteomyelitis, spina ventosa, bone infarction and sickle cell disease.\(^6\) Reinus et al reported in their study that the most common feature in cases of ES involving small joints of hand was a permeative bone lesion with poorly defined margins and an associated soft tissue mass which was also presented in our case.\(^7\)

Treatment of these tumors include local control of disease and adjuvant chemotherapy. Local control of disease can be achieved by surgery, radiation or both. With the advent of modern chemotherapy, the long term, 5-year survival rate has improved to approximately 70%.\(^8\)\(^9\) The prognosis of the disease depends upon many factors like gender, presence of anemia, fever etc. but location of tumour and presence of known metastasis at the time of presentation is the most important factor.\(^7\)

Kinsella reported use of combination therapy of chemotherapy and radiotherapy to control the primary lesion in the hand but the long-term result remains uncertain.\(^10\) The treatment of ES is achieved through a Common European protocol called Euro Ewing 99.\(^11\) Baccari et al reported 2 cases of ES involving phalanx of hand of which one case succumbed due to lung metastasis while remission was achieved in other case.\(^3\) Mahan et al reported a rare case of ES in the hand of an adult woman at 22 months after definitive treatment, remained free of disease.\(^12\)

The diagnosis should be considered in any lesion suspicious for malignancy. Currently, the most favourable treatment of ES of the small bones of extremities is surgery followed by chemotherapy. Neoadjuvant chemotherapy plays an important role to control local disease, skip metastasis and response of tumour to drugs.

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**REFERENCES**
