A Rare Case of Primary Ewings Sarcoma of the Nasal Bone

M.S. Vidyasagar, Professor & Head of Unit-II, **Suresh Rao**, Assistant Professor,
Donald. J. Fernandes, Additional Professor, **Ramanujam. A.S.**, Assistant Professor,
Dept. of Radiotherapy and Oncology,
Kasturba Medical College & Hospital, Manipal-576119, Karnataka, India.

Primary Ewing's sarcoma of the nasal bone has not been previously described. This case presented as a mass in the left ala of the nose in a five year old female child. The clinical, radiological, microscopic features are described and a review of literature is presented. The case was treated with neoadjuvant chemotherapy and local electron beam radiation therapy. The child was free of disease when she reported for follow up in July 1997. Although wide excision is part of the treatment approach in Ewing's sarcoma, in sites where surgery is not suitable local radiotherapy and chemotherapy adequately controls primary disease.

INTRODUCTION

Ewing's sarcoma accounts for approximately 10% of all primary bone tumours. Primary Ewing's sarcoma in the cranial and facial bone is rare.

Ewing's sarcoma most commonly occurs in the long bones and pelvis. The diaphysis is often involved when it occurs in the long bones. There is a male preponderance of approximately 1.6 : 1¹. Primary Ewing's sarcoma of the head and neck is rare condition, accounting for 2-3%¹. The majority occur in the mandible² or the maxilla³. Cases originating from skull⁴ and facial bone⁵ has been described but this is believed to be the first case arising from the nasal bone. The case is of interest because of the young age of the child and the difficulty encountered in planning the radiation treatment.

CASE REPORT

A girl aged five years and six months presented in January 1996 with a three months history of a

painless swelling in the left ala of the nose. There was no history of nasal block or discharge. The child was active and attended school. She was seen by a family doctor who performed a biopsy of the swelling. It was reported as lymphoma and the case was referred to the Shirdi Sai Baba Cancer Hospital and Research Center, Manipal, India.

Examination showed a firm mass in the right nasal region with a linear scar. The swelling was non-tender. The nasal septum was central. There were no other abnormal neurological or ophthalmological findings and the regional nodes were not enlarged.

Urine microscopy was normal. Haemoglobin concentration was 11 gms percent. The liver and renal function tests were within normal limits.

A bone marrow aspiration and bilateral iliac crest biopsy showed no evidence of infiltration. Ultrasound abdominal scan showed mild hepatomegaly with normal echotexture. A chest radiograph was normal and skull radiograph showed normal maxillary sinus and central nasal septum. The frontal sinus was not pneumatized. Bone scan with 99 Tc MDP showed abnormal uptake in nasal bone with rest of the skeleton being normal. CT scan revealed a soft tissue mass in the nasal region with erosion of the nasal bone (Figures 1 and 2).

The biopsy specimen of the tumour showed uniform, small round cells with abundant PAS positive material. The nuclei were round with spicled chromatin particles. The cytoplasm was sparse and showed ill defined margins. The neoplastic cells were displayed in sheets and also

Fig. 1 & 2

CT scan (Pre-chemotherapy) shows mass lesion with soft tissue component and nasal bone erosion

around blood vessels. Many mitoses and large areas of necrosis were present. Abundant endothelial lined vessels and muscle fibers were entrapped in the tumours. Individual or groups of cells were not surrounded by reticulin fibers and collagen fibers were present in the connective tissue stroma of the tumour. The histopathological features were interpreted as Ewing's sarcoma.

In view of the site of the lesion and the soft tissue component three courses of neoadjuvant

chemotherapy using VACA regime (Vincristine, Actinomycin, Cyclophosphamide, Adriamycin) were given. This produced complete clinical regression of the soft tissue mass and more than ninety percent regression on CT scan (figure 3).

Fig. 3

CT scan (Post-chemotherapy) shows excellent regression of the lesion in same section -0025

Local radiotherapy was planned using 6 Mev electron beam from a Mevatron KDs 2 with a margin of one centimeter around the lesion as seen on pre-chemotherapy CT scan. A total dose of 34 Gy in 180 cgy per fraction was given over three and a half weeks. This was followed by adjuvant chemotherapy using VACA three weekly for three courses and VAC (Vincristine, Actinomycin, Cyclophosphamide) three weekly for one year.

The child came for follow up on July 1997 and was found to be free of disease. Regular bone scan and chest X-ray every three months is planned to detect early local recurrence or distant metastasis.

DISCUSSION

In 1921, James Ewing's reported the first case of Ewing's sarcoma in a 14 year old girl with a lytic lesion in the ulna which responded dramatically to radiation. Significant progress has been made

in the treatment of Ewing's sarcoma with effective chemotherapy which has increased the 5 year survival of 5-10% twenty years ago to the current 5 year survival in excess of 70%^{6,7}. Extraskelatal Ewing's sarcoma is indistinguishable both clinically and pathologically from the tumour arising from the bone⁸. The case reported here has a large proportion of its bulk involving the soft tissue but appears to arise from the nasal bone.

The histogenesis of Ewing's sarcoma is controversial although Roessner et al. feel that it arises from immature reticulum, myogenous, endothelial and undifferentiated mesenchymal cells⁹. The histological differential diagnosis of Ewing's sarcoma has been improved by immunohistological methods. In most cases they can be distinguished from lymphoma (leucocyte common antigen, B and T cell marker) and embryonal rhabdomyosarcoma (muscle specific action, desmin) without problems.

Most of the primary tumours occur in the long bones (47%), pelvis (29%), ribs (12%). The skull is primarily involved in fewer than 4% cases¹⁰. Pain and swelling are the most common symptoms. Approximately one fifth of the patients have fever, which may lead to the mistaken diagnosis of osteomyelitis. The case presented here had only a swelling which was not associated with any other symptoms. Involvement of facial bones is characterized by clinical and radiological features distinct from those commonly observed in other sites³. Hence a delay in the diagnosis is likely in such cases.

The major prognostic factors in Ewing's sarcoma are the site, volume of the tumour¹¹, presence of metastasis at diagnosis² and the tumour response to chemotherapy.

The disease evaluation is performed using plain radiograph of bone, MRI or CT scan. MRI is more sensitive than the CT scan in assessing soft tissue involvement and the bone marrow spread. The early recurrence at the primary site can be detected by performing a bone scan. Biopsy

related problems occur more frequently when biopsy is done at a referring institution than the treating center. The case here also was reported as lymphoma from the referring hospital and a review of the tissue and slide at our center was reported as Ewing's sarcoma. The diagnosis of Ewing's sarcoma was traditionally made by excluding other round cell tumours that occur in bone and by demonstrating the presence of glycogen in the tumour cells using periodic acid Schiff (PAS) stain. Recent advances in the cytogenetic, immunocytochemistry and electron microscopy have allowed pathologists to diagnose Ewing's sarcoma more accurately.

The control of local disease has become more important with increased long term survival rates following the advent of modern chemotherapy. The general philosophy is to give intense neoadjuvant chemotherapy (Vincristine, Actinomycin, Cyclophosphamide, Adriamycin) with VACA regime to decrease the size of the primary tumour and to control distant micrometastasis. This is followed by wide en-bloc excision or radiation therapy if excision is not possible because of the location of the tumour or lack of microsurgical facilities¹². The case reported here had more soft tissue component and because of the location neoadjuvant chemotherapy and local irradiation with 6 Mev electrons was planned.

Radiation therapy is an important modality in treating Ewing's sarcoma locally. Refinement in radiotherapy technique and availability of electron beam with suitable energies has enabled Radiation Oncologist to deliver adequate dose to the target volume with minimal dose to the surrounding normal tissues. Two cases of primary Ewing's sarcoma of the maxillary antrum treated with radiation therapy and systemic chemotherapy was reported recently³. This is more so in particular sites which are not suitable for radical surgery. In patients with good response of the primary lesion to neoadjuvant chemotherapy the dose of radiation for local control can be reduced

leading to minimal late complication.

The small number of Ewing's sarcoma originating in facial region and the variety of treatment used preclude any meaningful evaluation of definitive

therapy. This case shows the successful treatment of Ewing's sarcoma in an unusual location with neoadjuvant chemotherapy and electron beam radiotherapy.

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