Asian Journal of Medical Research Case Report

Disseminated Ewings Sarcoma With Bilateral Proptosis– A Case Report

AshaLatha D[1], Lokesh V[1], Ajay GV[1], K.P.Jaganath[1], Anand John[1], Shruthi K[1]
[1]Department of Radiation Oncology,Kidwai Memorial Institute of Oncology, Bangalore Karnataka State, India

Abstract

Ewing's tumor, primarily a bony lesion, has classically been studied under the heading of small, round cell tumors.A 13year old boy presented with complaints of painful swelling over the left thigh since 4 months, loss of vision in both eyes since 3 months.Tumor was organized into cell nests, separated by thin trabeculations

Key Words: Ewings tumor,pain,eyes

INTRODUCTION

Ewing's tumor, primarily a bony lesion, has classically been studied under the heading of small, round cell tumors.[1] The mean age of occurrence is between the first and the second decades in 80% of cases.[2] Ewing's sarcoma is a malignant tumor which very rarely involves cranial cavity, of its cranial sites, temporal location assumes low priority and further intraorbital extension is even rarer. We report a young boy who presented with complaints of intolerable painful proptosis of the left eye, bilateral complete blindness and left temporal fossa swelling along with swelling over the left thigh. Patient was treated with palliative radiotherapy for proptosis of the left eye. Patient later expired due to advanced disease.

CASE REPORT

A 13year old boy presented with complaints of painful swelling over the left thigh since 4 months, loss of vision in both eyes since 3 months, protrusion of both the eyes more in the left eye associated with severe pain and generalized weakness since 15 days. General physical examination revealed an emaciated, bedridden and poor general condition of the patient. Clinical examination revealed a huge mass measuring 20*10 cm, hard, non- tender, painful and restricted mobility over the left thigh (Fig 2). On local examination of face-bilateral proptosis of eyes was noted. In left eye-multiple lobulated and hard bosselated mass was felt, protruding out of orbit extending laterally to temporal region just above the pinna of the left ear (Fig 1). In right eye- a hard bosselated mass was felt. X-ray of the left femur revealed- sub acute osteomyelitis involving proximal and middle 3rd of the shaft of the left femur. USG guided biopsy of left thigh mass revealed a neoplasm composed of fairly uniform round to oval cells having few mitotic figures with relatively clear and indistinct cytoplasm .Tumor was organized into cell nests, separated by thin trabeculations .On staining, these cells were positive to PAS and vimentin but negative to leukocyte common antigen, desmin and synaptophysin(Fig 3). Bone scan showed increased osteoblastic activity in head and shaft of the left femur, T5, L5 vertebra, skull and multiple bilateral ribs. CT thorax showed pulmonary metastasis. Bone marrow aspiration showed no evidence of metastasis. Based on these findings, a diagnosis of disseminated Ewing's sarcoma with bilateral proptosis was made.

Patient received 1600cGy in 4 fractions at 4cms depth to his left eye following which proptosis decreased in the next 2 days, and the patient's condition improved symptomatically. One week later patient had dyspneoa and severe pain over his left thigh, investigation with chest x ray revealed bilateral pleural effusions and bilateral multiple in homogenous opacities (metastasis), for which he was given symptomatic and supportive care .Patient expired due to advanced disease after 2weeks. Good palliation due to local radiotherapy was obtained in terms of pain relief & decreased proptosis to the left orbit.

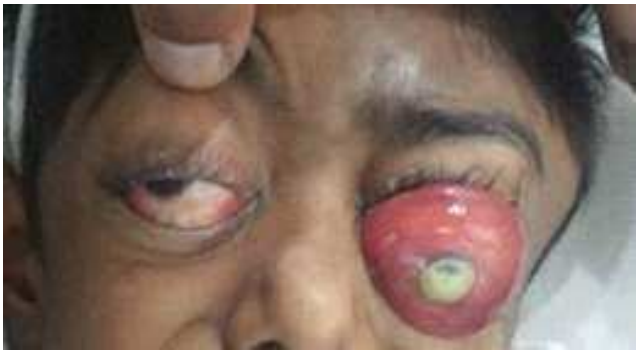

Figure 1: Bilateral proptosis of eyes. In left eye-multiple lobulated and hard bosselated mass, protruding out of orbit.

DISCUSSION

Ewing's sarcoma, first described by James Ewing in 1921, comprises 10% of all bony tumours[3]. It commonly affects pediatric population with 75% of patients under 20 years of age. The peak incidence is between 5 - 13 years.[4]Males are more frequently affected, with male-to-female ratio being about

Address for correspondence*
Dr AshaLatha D,
Resident, Department of Radiation Oncology,
Kidwai Memorial Institute of Oncology,
Hosur road, Bangalore 560029 Karnataka State, India
E mail: drashalatha85@gmail.com

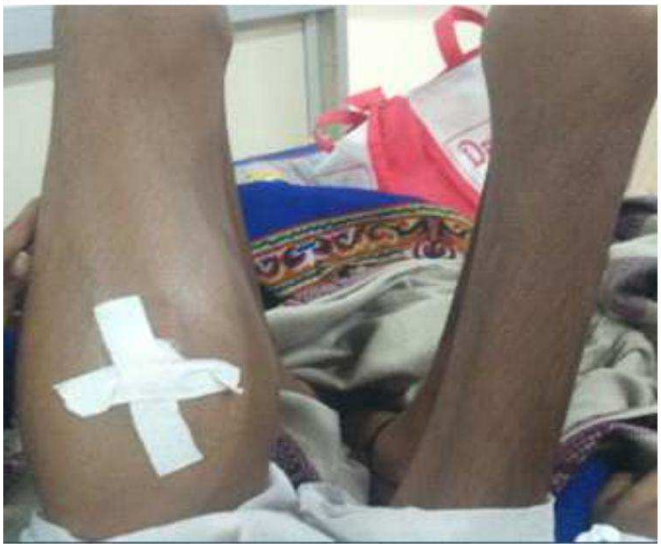

Figure 2 : Huge mass measuring 20*10 cm, hard, non-tender, painful and restricted mobility over the left thigh

1.6:1.4.[4,5] Long bones of lower extremity and flat pelvic bones are favored sites of origin, although the tumor can arise in other bones and, on occasion, in soft tissues also.[6] Primary Ewing's sarcoma of the calvarium is very rare, and is found in less than 1% of the cases.

Frontal and parietal convexities are the common sites of occurrence.[4,6,7] Multicentricity have been reported in one or two cases, affecting temporal bone with significant intracranial and intra-orbital extension (as seen in our case).

However, significant unilateral extra-cranial, intracranial and intra-orbital extension has been reported.[8]

Headache and features of raised intracranial pressure are most common presenting complaints, followed by localized scalp swelling, hemiparesis, diplopia and seizures.[9] Primary involvement of orbit is rare, more often it occurs in setting of disseminated disease.[10] Bony involvement and bony healing are better demonstrated on CT scans. Radioisotope bone scan is essential to rule out lesions elsewhere.

Though radical surgery has been advocated, a less aggressive approach can be considered as these tumors are highly sensitive to chemoradiation.[6,9,10] The differential diagnosis of intracranial small round cell tumors includes primitive neuroectodermal tumors, rhabdomyosarcomas, metastatic neuroblastomas and lymphomas.[4,7] The characteristic histological findings of Ewing's sarcoma includes round tumor cells with round vesicular nuclei and prominent nucleolus compactly

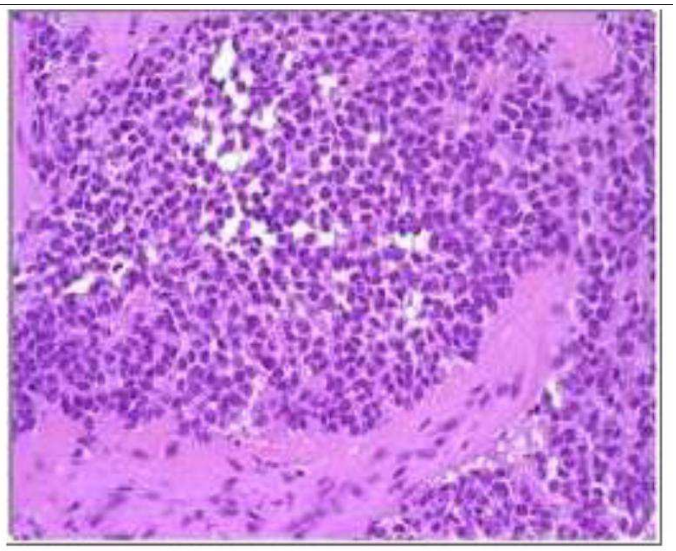

Figure 3 : Figure 3: Histological features of Ewing's sarcoma includes round tumor cells with round vesicular nuclei and prominent nucleolus compactly arranged in sheets, in frequent mitoses, scanty cytoplasm and absence of rosettes.

arranged in sheets, in frequent mitoses, scanty cytoplasm and absence of rosettes (Fig 3). Lack of expression of leukocyte common antigen, desmin and synaptophysin excludes possibility of lymphoma, rhabdomyosarcomas and neuroblastoma respectively. Periodic acid Schiff and vimentin positivity further concluded the diagnosis of Ewing's sarcoma.

The 5-year survival rates have dramatically improved from 15 to 60% since introduction of combination multi-drug chemotherapy and radiotherapy.[6,7,12,13] Only palliative care was given in our case as the general condition was very poor and due to the advanced disease.

Alvarez-Berdecia et al described prognostic factors in primary cranial Ewing's sarcoma.[8] Good prognosis was associated with female sex, long duration of symptoms, absence of systemic symptoms, peripheral localization of tumor, absence of metastases, initial LDH levels less than170 IU/L, leukocyte count less than 2000/dl, tumor size less than 8 cm and small cell histology.

CONCLUSION

In view of poor general condition, no active treatment was contemplated, but palliative radiotherapy was given for severe painful proptosis of the left eye. Good palliation due to local radiotherapy was obtained in terms of pain relief & decreased proptosis of the left orbit.

REFERENCES

1. Esiashvili N, Goodman M, Marcus RB., Jr Changes in incidence and survival of Ewing sarcoma patients over the past 3 decades: Surveillance Epidemiology and End Results data.J Pediatric Hematological Oncol. 2008; 30:425–30.
2. Arndt CA, Crist WM. Common musculoskeletal tumors of childhood and adolescence. N England Journal of Med.1999; 29:341–52.
3. Rosai J: Bone and joints. In: Rosai J, Ackerman LV (eds),*Surgical Pathology*, 9th Ed. St. Louis, Mosby 2009, pp 2137-2236.
4. Steinbok P, Flodmark O, Norman MG, Chan KW, Fryer CJ: Primary Ewing's sarcoma of the base of the skull.Neurosurgery 1986, 19: 104-107.
5. Li WK, Lane JM, Rosen G, Marcove RC, Caparros B, HuvosA, Groshen S: Pelvic Ewing's sarcoma. Advancesintreatment. Bone Joint Surg Am 1983, 65: 738-747.
6. Alvarez-Berdecia A, Schut L, Bruce DA: Localized primaryintracranial Ewing's sarcoma of the orbital roof. J Neurosurg1979, 50: 811-813.
7. Fitzer PM, Steffey WR: Brain and bone scans in primary Ewing's sarcoma of the petrous bone. J Neurosurg 1976, 44:608-612.
8. Kuzeyli K, Aktürk F, Reis A, Cakir E, Baykal S, Pekínce A, UsulH, Söylev AE: Primary Ewing's sarcoma of the temporal bonewith intracranial, extra cranial and intra-orbital extension.Neurosurg Rev 1997, 20: 132-134.
9. Desai KI, Nadkarni TD, Goel A, Muzumdar DP, Naresh KN,Nair CN: Primary Ewing's sarcoma of the cranium. Neurosurgery2000, 46: 62-69.
10. Woodruff G, Thorner P, Skarf B: Primary Ewing's sarcoma ofthe orbit presenting with visual loss. Br J Ophthalmol 1988,72: 786-792.
11. Watanabe H, Tsubokawa T, Katayama Y, Koyama S, NakamuraS: Primary Ewing's sarcoma of the temporal bone. SurgNeurol 1992, 37: 54-58.
12. Pilepich MV, Vietti TJ, Nesbit ME, Tefft M, Kissane J, BurgertEO, Pritchard D: Radiotherapy and combination chemotherapyin advanced Ewing's sarcoma-Intergroup study. Cancer 1981,47: 1930-1936.
13. Rosen G, Wollner N, Tan C, Wu SJ, Hazdu SI, Cham W, D'Angio GJ, Murphy ML: Disease free survival in children with Ewing's sarcoma treated with radiation therapy and adjuvant four-drug sequential chemotherapy. Cancer 1974, 33: 384-393.