

Primary Extraosseous Ewing Sarcoma of Abdominal Wall: A Rare Entity

Channabasappa Kori^{1*}, Neelesh Jain², D Prashant Kumar³, Jeewanram Vishnoi⁴, Sameer Gupta⁵, Vijay Kumar⁶

¹⁻⁴Senior resident, Department of Surgical Oncology, King George's Medical University, Lucknow, UP, India

⁵Assistant Professor, Department of Surgical Oncology, King George's Medical University, Lucknow, UP, India

⁶Associate Professor, Department of Surgical Oncology, King George's Medical University, Lucknow, UP, India

*Corresponding Author:

Name: Channabasappa Kori

Email: channabasappakori@gmail.com

Abstract: Ewing sarcoma is malignant round cell tumour that primarily affects bones. Primary Extra osseous Ewing sarcoma (EES) is extremely rare. Clinical and radiologic features are non specific. Diagnosis is mainly based on immunohistochemistry. This article presents a rare case of Primary extrosseous soft tissue Ewing sarcoma of right antero-lateral abdomen wall in a 30 year old lady and was successfully treated with multi agent chemotherapy and local treatment in the form of aggressive surgery and radiation.

Keywords: Ewing sarcoma, Abdominal Wall.

INTRODUCTION

Ewing sarcoma is most commonly a bone tumour and may extend into the soft tissues at the time of presentation. It commonly presents in children and young adults [1]. James Ewing described Ewing's sarcoma in 1921 as a tumour arising from undifferentiated osseous mesenchymal cells. Based on recent studies; Ewing sarcoma may be of neuroectodermal origin, arising from primitive neural crest cells [2]. Ewing's sarcoma having extra skeletal origin is highly uncommon [3]. There are only few case reports of the Extra osseous Ewing sarcoma (EES) being reported in the literature [4, 5]. Most commonly affected sites are paravertebral spaces, lower extremities, head and neck, pelvis; but rarely affects abdominal wall [6]. To the best of our knowledge, few cases of EES affecting abdominal wall are reported. Here, we describe an unusual case of extra skeletal Ewing sarcoma primarily arising from right flank region and treated successfully with multimodality treatment.

CASE REPORT

A 30 year old lady presented with 6 month history of swelling over right lumbar region with recent onset of pain. Physical examination showed a bosselated swelling on the right anterolateral aspect of abdomen wall involving lumbar and iliac region measuring 25 x 15cm in dimensions (Fig. 1).

Contrast enhanced computed tomography (CECT) of abdomen showed well lobulated, heterogeneously enhancing soft tissue attenuation lesion

measuring approximately 23 x 18.5 x 9 cm in size with cystic and necrotic area changes is seen involving right lateral abdomen wall in lumbar and iliac region. The lesion was involving underlying muscles, overlying skin and subcutaneous tissue (Fig. 2). A whole body bone scintigraphy study revealed no evidence of skeletal metastasis with normal uptake of tracer. Chest radiography was within normal limit.

Trucut biopsy from the swelling revealed round cell sarcoma probably Ewing sarcoma. Patient received neoadjuvant chemotherapy including VAC regimen (Vincristine, Adriamycin, Cyclophosphamide) and IE regimen (Ifosfamide, Etoposide) alternatively. Patient showed stable disease after 3 cycles of IE and VAC regimen. Patient underwent wide local excision of the lesion with primary closure and meshplasty. Postoperative course was uneventful. Gross examination showed infiltrative growth measuring 24 x 17 x 6cm (Fig. 3). Microscopy revealed sheets of monomorphic round cells with focal alveolar pattern, eosinophilic cytoplasm and hyperchromatic nuclei (Fig. 4). Immunohistochemistry study showed tumour cells positive for CD 99 (Fig. 5) and FLI-1, negative for desmin, cytokeratin and synaptophysin. Hence final diagnosis of Ewing sarcoma was made. Patient completed adjuvant chemotherapy and is disease free at one year follow up.



Fig. 1: Physical examination showed a bosselated swelling on the right anterolateral aspect of abdomen wall involving lumbar and iliac region measuring 25 x 15cm



Fig. 2: CECT scan showed well lobulated, heterogeneously enhancing soft tissue lesion measuring approximately 23 x 18.5 cm



Fig. 3: Gross examination showed infiltrative growth measuring 24 x 17 x 6cm

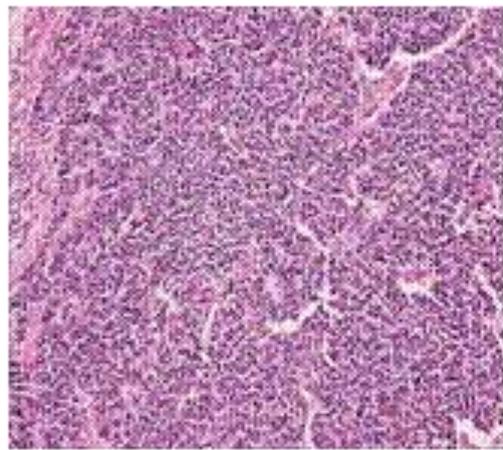


Fig. 4: Microscopy revealed sheets of monomorphic round cells with focal alveolar pattern, eosinophilic cytoplasm and hyperchromatic nuclei

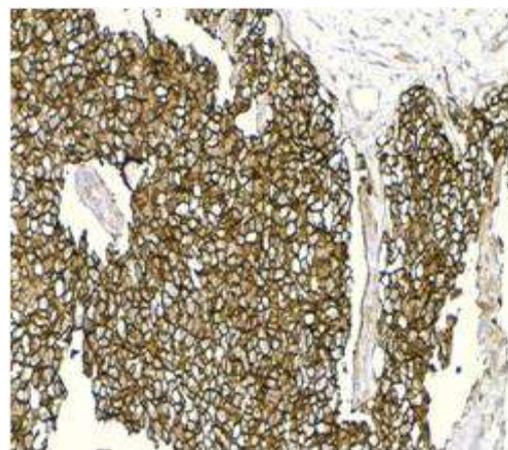


Fig. 5: IHC study showed tumour cells positive for CD 99

DISCUSSION

Extrasosseous Ewing sarcoma (EES) is an unusual rapidly growing soft tissue sarcoma. It is composed of small undifferentiated round to oval cells histologically resemble skeletal ewing sarcoma [1]. Diagnosis is based mainly on the basis of histological findings with no evidence of bony involvement at the time of presentation. Extra osseous Ewing sarcoma is a subtype of Ewing sarcoma/ Primitive neuroectodermal (PNET) family. EES predominantly affects young adolescents and adults between the age group of 10- 30 years with male predilection [7]. EES observed in about 15% of cases and classically involves the soft tissues of the chest wall, pelvis, paravertebral region, retroperitoneal region, orbit, skin, head & neck and extremities, but it uncommonly affects abdominal wall [8, 9]. Based on extensive literature, very few cases of EES affecting abdominal wall are reported [10]. Based on recent report by Applebaum et al; patients with EES had slightly higher mean age as compared with patients of skeletal Ewing sarcoma and most frequent site of EES is axial skeleton.

Most common presenting symptom is rapidly growing mass and pain, rarely with constitutional symptoms like fever, anorexia and weight loss [11]. Radiologic features were nonspecific. Ultrasonography (USG) and computed tomography (CT scan) reveals large, sharply delineated heterogeneous mass with post contrast enhancement. On magnetic resonance imaging (MRI), tumour shows low to intermediate signal intensity on T-1 weighted images and high signal intensity on T-2 weighted images and exhibits heterogeneous contrast enhancement [11, 12]. Rarely, tumour may cause cortical erosion and periosteal reaction as secondary changes on imaging. Imaging is also useful to assess the rate of resectability and tumour response to chemotherapy. Based on radiologic findings, differential diagnosis includes Rhabdomyosarcoma, Malignant fibrous histiocytoma and Dedifferentiated sarcoma. Diagnosis is confirmed by histopathological examination of biopsy specimen and immunohistochemistry study (IHC) [13]. EES should be treated with a multidisciplinary approach comprising of multi agent chemotherapy and local treatment in the form of aggressive surgery and radiotherapy. Systemic chemotherapy includes VAC (Cyclophosphamide, Adriamycin, Vincristine), alternating with IE (Ifosfamide, Etoposide) regimen [14]. Local radiation is used postoperatively for close or positive margin and as definitive modality if the primary cannot be excised completely. Although Ewing tumour is radiosensitive, but radiotherapy does not seem to be primary modality. Role of multi-agent chemotherapy is well established and have improved the 5 and 10 year survival rates from 50 to 60% [15].

Localised EES had a better 5 year survival rate as compared to osseous Ewing sarcoma as per study by Applebaum *et al.* [16]. Based on study by Ahmed *et al.*, EES is a curable disease and has the best prognosis in younger patients treated with surgical resection in addition to chemotherapy and local radiation therapy [7]. The study also showed that tumour size did not have a significant effect on overall 5 year survival or disease free survival. Most common sites for distant metastasis are lung, liver, brain and bone.

CONCLUSION

Extraskeletal Ewing sarcoma is highly malignant round cell tumour arising from soft tissues is an unusual entity. Clinical and radiologic features are non specific. Imaging can be used to assess tumour response to chemotherapy and resectability of the tumour. Diagnosis is mainly based on immunohistochemistry. Neoadjuvant chemotherapy, aggressive surgical resection, followed by adjuvant radiotherapy resulted in a favourable clinical outcome for EES. EES has better disease free survival as compared to osseous sarcoma. Diagnosis of EES should

always be kept in mind in the differential diagnosis of soft tissue tumours in young patients.

REFERENCES

1. Paoletti H, Colineau X, Acalet L, Tourrette JH, Civatte M, Fesselet J *et al.*; Ewing's sarcoma of the soft tissues: apropos of 3 cases and review of the literature. *J Radiol.*, 1999; 80(5): 477-482.
2. Yalcin S, Turoglu HT, Ozdamar S, Sadikoglu Y, Gurbuzer B, Yenici O; Ewing's tumor of the mandible. *Oral Surgery Oral Med Oral Pathol.*, 1993; 76(3): 362-367.
3. Mukkunda R, Venkitaraman R, Thway K, Min T, Fisher C, Horwich A *et al.*; Primary adult renal Ewing's sarcoma: A rare entity. *Sarcoma*, 2009; 2009: 504654.
4. Chow E, Merchant TE, Pappo A, Jenkins JJ, Shah AB, Kun LE; Cutaneous and subcutaneous Ewing's sarcoma: an indolent disease. *Int J Radiat Oncol Biol Phys.*, 2000; 46(2): 433-438.
5. Somers GR, Shago M, Zielenska M, Chan HS, Ngan BY; Primary subcutaneous primitive neuroectodermal tumor with aggressive behavior and an unusual karyotype: case report. *Pediatr Dev Pathol.*, 2004; 7(5): 538-545.
6. Askri A, Farhat LB, Ghariani B, Rabeh A, Dali N, Said W *et al.*; Extraskeletal Ewing sarcoma of the abdominal wall. *Cancer Imaging*, 2008; 8(1): 156-158.
7. Ahmad R, Mayol BR, Davis M, Rougraff BT; Extraskeletal Ewing's sarcoma. *Cancer*, 1999; 85(3): 725-731.
8. O'Keeffe F, Lorigan JG, Wallace S; Radiological features of extraskeletal Ewing sarcoma. *Br J Radiol.*, 1990; 63(750): 456-460.
9. Cheung CC, Kandel RA, Bell RS, Mathews RE, Ghazarian MD; Extraskeletal Ewing sarcoma in a 77-year-old woman. *Arch Pathol Lab Med.*, 2001; 125(10): 1358-1360.
10. Aydinli B, Ozturk G, Yildirgan MI, Polat KY, Basoglu M, Gundogdu C *et al.*; Extraskeletal Ewing's sarcoma in the abdominal wall: a case report. *Acta Oncol.*, 2006; 45(4): 484-486.
11. Shirkhoda A, Peuchot M; Extraosseous Ewing's sarcoma. Computed tomography evaluation before and after chemotherapy. *Clin Imaging*, 1989; 13(2): 142-146.
12. Perouli E, Chrysikopoulos H, Vlachos A, Koskinas A, Batistatou A, Polyzoidis K; Imaging findings in paraspinal extraosseous Ewing sarcoma. *JBR-BTR*, 2006; 89(6): 310-312.
13. Hasegawa SL, Davison JM, Rutten A, Fletcher JA, Fletcher CD; Primary cutaneous Ewing's sarcoma: immunophenotypic and molecular

- cytogenetic. Evaluation of five cases. *Am J Surg Pathol.*, 1998; 22(3): 310-318.
14. Ladenstein R, Pötschger U, Le Deley MC, Whelan J, Paulussen M, Oberlin O *et al.*; Primary disseminated multifocal Ewing sarcoma: results of the Euro- EWING 99 trial. *J Clin Oncol.*, 2010; 28(20): 3284-3291.
 15. Ullmann C, Beck JD, Holter W, Petsch S, Dunst J, Sauer R *et al.*; Long-term results following multidisciplinary treatment of localized Ewing's sarcoma in children and adolescents. *Strahlenther Onkol.*, 2008; 184(3):137-144.
 16. Applebaum MA, Worch J, Matthay KK, Goldsby R, Neuhaus J, West DC *et al.*; Clinical features and outcomes in patients with extraskeletal Ewing sarcoma. *Cancer.* 2011; 117(13): 3027-3032.