DOI: https://doi.org/10.18320/JIMD/201704.0142

Case report

Journal of International Medicine and Dentistry 2017; 4(1): 42-46

JOURNAL OF INTERNATIONAL MEDICINE AND DENTISTRY

To search.....to know.....to share

JIMD

p-ISSN: 2454-8847

e-ISSN: 2350-045X

Ewing's sarcoma of maxilla: A case report

Punyasloka Pati[1], Suryanarayan Das[2], Biranchi Narayan Biswal[3], Rachna Rath[4]

Abstract:

Background: Ewing's sarcoma (ES) is a malignancy primarily affecting bone tissue that is commonly diagnosed in adolescents and young adults. Its occurrence in the head and neck region is unusual and generally involves the mandible and maxilla. Radiographic finding in ES reflect many destructive nature of the lesion, like osteolysis, cortical erosion and soft tissue mass**.**

Case report: We report a rare case of ES of the maxilla in a 10-year-old boy with clinical, radiological, histopathological and immunohistochemical findings. Early detection of such lesions is difficult because the signs and the symptoms do not appear until the lesion has progressed considerably.

Conclusion: This case explicates the importance of professional knowledge of the relevant aspects of malignant lesions such as Ewing's sarcoma.

Keywords: Ewing's sarcoma, immunohistochemistry, mandible

Introduction:

Ewing's sarcoma (ES) is a rare malignant small round cell tumor that primarily affects the skeletal system[1]. It accounts for 4 to10% of all types of bone cancer, with long bones and pelvis being the most common locations.[1,2] It affects mainly adolescents and young adults and is rarely seen before the age of 5 years and after the age of 30 years. Clinically, this tumor has an aggressive behaviour characterized by rapid growth and high probability of micrometastasis at diagnosis.[3] The occurrence of ES in the head and neck region is unusual and, when it occurs, it generally involves the mandible and less frequently the maxilla. [4,5]

Clinically, patients are usually young adults. Typical symptoms are bone swelling & often pain, progressing over a period of months. Paraesthesia and loosening of teeth are common findings while, low grade fever, leukocytosis and increased ESR are other findings which may be present. Radiographically, it is an irregular, lytic, ill-defined radiolucent lesion. Appearance of jaw swelling is rapid along with intra-oral mass which may soon become ulcerated.[6,7,8] Here, we present a case report of Ewing's sarcoma in a 10-year old male patient.

Case report:

A 10-year old male reported with chief complaints of a slow growing swelling on left side of midface region (**Figure Ia, b**). The swelling gradually increased in size over last six months without pain. The medical history was non-contributory. The clinical examination showed a swelling of 6X4 cm in the left side midface region extending from base of nose to posterior part of maxilla posteriorly and inferior border of orbit superiorly. The swelling was firm, non-tender, non-fluctuant, non-pulsatile and smooth on palpation. The overlying mucosa was normal in colour & texture. Intra-oral examination revealed a diffuse swelling on hard palate extending from 12 tooth region to the left maxillary tuberosity obliterating the buccal vestibule. CT scan showed an expansile, osteolytic lesion involving anterior and left side of maxilla(**Figure Ic, d**). The lesion was provisionally diagnosed as bone neoplasm and an incisional biopsy was planned.

Figure I (a, b): Firm, non-tender swelling over left midface/ intra-orally
Figure I (c, d): CT scan showing expansile, osteolytic lesion

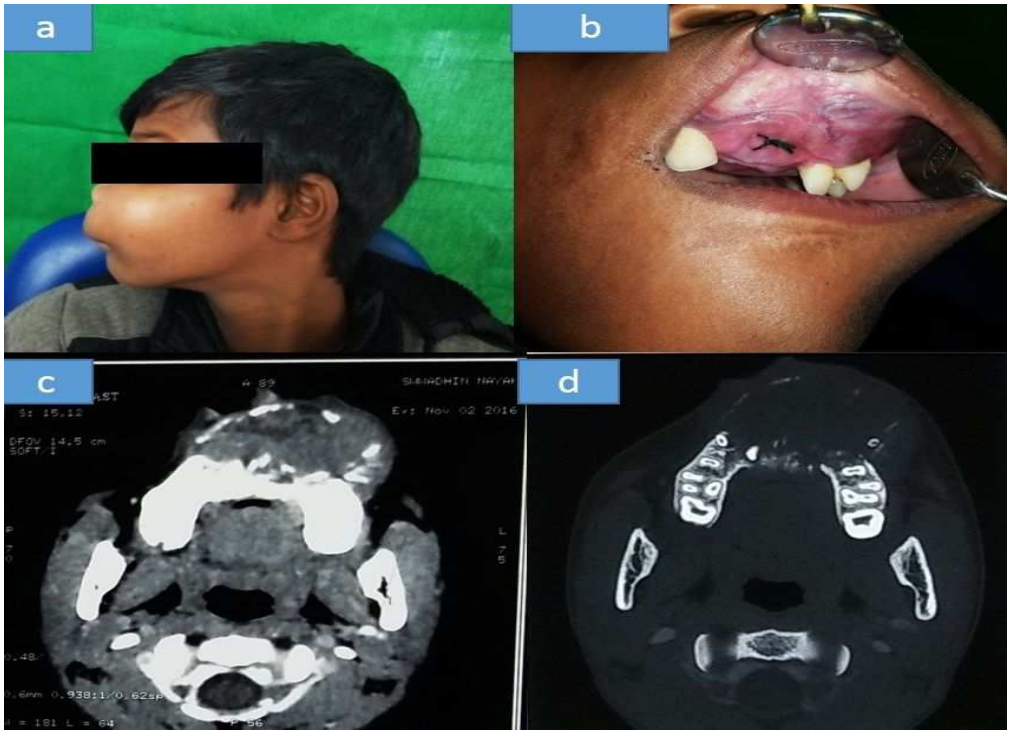
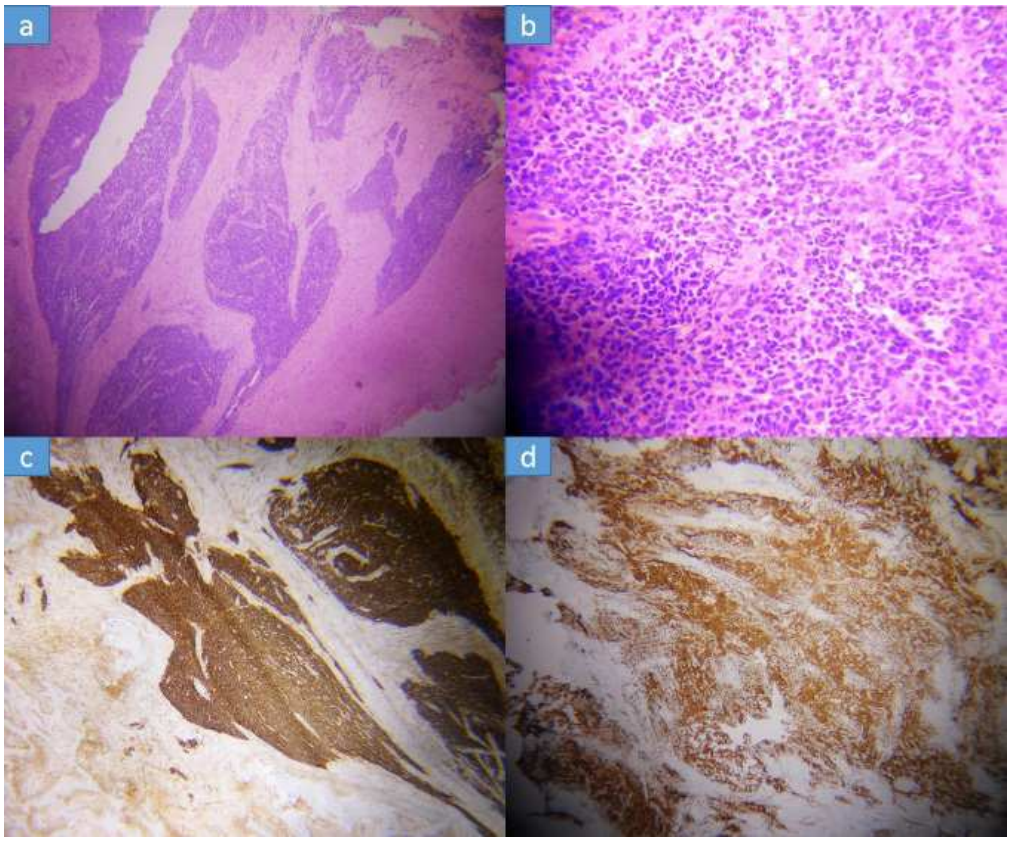


Figure II (a, b): Histopathology of tissue(H & E stain x40, x400)
Figure II (c, d): Immunohistochemistry –CD99+++, FLI 1+++



H & E stained sections revealed solidly packed, lobular pattern of uniform round cells. The individual cells had a round or ovoid nucleus with a distinct nuclear membrane in a fibrovascular stroma. Most of the individual cells exhibited ill-defined scant cytoplasm, round and oval hyperchromatic nuclei and inconspicuous nucleoli (**Figure IIa, b**). Few mitotic figures and also areas of necrosis were seen. Based on histopathological features differential diagnosis of small round blue, cell tumor possibly Ewing's sarcoma, rhabdomyosarcoma, lymphoma, neuroblastoma was considered. Immunohistochemistry study showed the tumor cells were negative for epithelial differentiation (Cytokeratin), lymphoblastic markers (CD45), rhabdoid differentiation (Desmin) and neuroendocrine differentiation. The tumor cells were strongly positive for CD99 & FLI1 (**Figure IIc, d**). Based on the IHC finding a final diagnosis of Ewing's sarcoma was made.

Discussion:

ES is a malignant neoplasm that primarily affects long bones of the extremities with nearly 50% of reported cases involving the femur and pelvis. It exhibits a marked predilection for whites and is rarely seen among blacks. The majority of the patients affected are between the ages 5 and 20 years, whereas the disease is distinctly uncommon in individuals before age 5 and after age 30 years. ES arising from the bones of the head and neck region is exceedingly uncommon. When it occurs in the jaw, mandible is more frequently affected than the maxilla. But in our case, it was the maxilla.[2,3,5, 9]

In the head and neck region the clinical findings are not specific for ES, although most of the patients complaints at the time of presentation are commonly associated with mass effect of the tumor and include rapid growth, swelling of the affected area and pain. [10,11] The present case showed clinical aspects similar to those reported in the literature for cases in the head and neck region: it was located in the maxilla & presented with a rapidly progressive growth.

Radiographically, ES appears as a poorly defined osteolytic lesion that may be frequently associated with cortical erosion and soft tissue mass adjacent to the destructive site.[11] The presence of sun-ray spicules of periosteal bone and presence of the laminar periosteal response ("onion skin" reaction) described as the commonest radiological features for lots of ES of the long bones, is rarely seen in jaw lesions for ES affecting jaw bones. [10] In the present case, radiographically there was an osteolytic lesion, neither sun-ray spicules of periosteal bone nor an "onion skin reaction" of bone cortical region were observed.

Histopathologically, ES is composed of small, poorly differentiated cells with medium-size, round or oval nuclei exhibiting a fine chromatin pattern, small nucleoli and scanty cytoplasm. [11] The intracytoplasmic glycogen may be demonstrated by PAS stain in75% of the cases, but it is not pathognomonic and conclusive because other small round cell may show the presence of glycogen as well.[12,13]

The use of immunohistochemistry helped in the diagnosis of this tumor. In general, the tumor cells are positive for vimentin and CD99 and negative for neural, skeletal, vascular and lymphoid cell markers. More than 90% of cases show a characteristic translocation t (11;22) (q24; q12) resulting in the fusion of the EWS and FLI-1 genes. This gene rearrangement causes a fusion product which functions as an oncogenic aberrant transcription factor with structural variability and potentially prognostic impact. FLI 1 nuclear positivity has been reported in 71%–84% cases of ES and polyclonal antibodies to this protein have been developed. So, FLI-1 positivity can aid in distinguishing ES

from other CD99 positive round cell tumours and antibodies to FLI 1 may play a valuable adjunctive role in the diagnosis of ES.[14,15] The present case was positive for CD99, FLI 1 and negative for other immunomarkers, leading to a diagnosis of ES.

It has been reported that combined therapy including surgery, radiotherapy and chemotherapy is the best approach for ES. The treatment should include wide surgical resection and neoadjuvant chemotherapy.[14]

Conclusion:

Ewing's sarcoma is a rare malignancy with diagnostic challenge because of manyoverlapping clinical, radiographic, histopathological and immunohistochemical features with other malignant round cell tumors. So, the distinction between these tumours is important as they require management via multi-disciplinary approach. Early diagnosis by advancing techniques such as immunohistochemistry and cytogenetic studies, with initiation of treatment can improve the overall survival rate.

References:

1. Prasad BV, Mujib BRA, Bastian TS, Tauro PD. Ewing`s sarcoma of the maxilla. Indian J Dent Res 2008; 19: 66-69; https://doi.org/10.4103/0970-9290.38935

2. Infante-Cossio P, Gutierrez-Perez JL, Garcia-Perla A, Noguer Mediavilla M, Gavilan-Carrasco F. Primary Ewing's sarcoma of the maxilla and zygoma: report of a case. J Oral Maxillofac Surg 2005; 63:1539-1542; https://doi.org/10.1016/j.joms.2005.06.011

3. Heare T, Hensley MA, Dell'Orfano S. Bone tumors: osteosarcoma and Ewing's sarcoma. Curr Opin Pediatr 2009; 21:365-372;https://doi.org/10.1097/MOP.0b013e32832b1111

4.Schultze-Mosgau S, Thorwarth M, Wehrhan F, Holter W, Stachel KD, Grabenbauer G, et al. Ewing's sarcoma of the mandible in a child: interdisciplinary treatment concepts and surgical reconstruction. J Craniofac Surg 2005; 16:1140-146; https://doi.org/10.1097/01.scs.0000180005.52986.19

5. Lopes SL, Almeida SM, Costa AL, Zanardi VA, Cendes F. Imaging findings of Ewing's sarcoma in the mandible. J Oral Sci 2007; 49:167-71; http://doi.org/10.2334/josnusd.49.167

6. Rajendran R. Benign and Malignant tumors of oral cavity. In: Shafer 's Textbook of Oral Pathology. 5th ed. New Delhi: Mosby; 2007: p. 113-308.

7. Parija T, Shirley S; Uma S, Rajalekshmy KR, Ayyappan S and Rajkumar T. Type 1(11; 22)(q24:q12) translocation is common in Ewing's Sarcoma/Peripheral Neuroectodermal tumor in south Indian patients. J Biosci 2005; 30: 371-76; https://doi.org/10.1007/BF02703674

8. Kumar V, Abbas AK, Kllusto N. Bones, Joints and Soft tissue tumors. In: Pathologic Basis of diseases. 7th ed. New Delhi: Elsevier; 2004: p. 1273-1324.

9. Vaccani JP, Forte V, de Jong AL, Taylor G. Ewing's sarcoma of the head and neck in children. Int J Pediatr Otorhinolaryngol 1999; 48: 209-216; https://doi.org/10.1016/S0165-5876(99)00030-0

10. Wood RE, Nortje CJ, Hesseling P, Grotepass F. Ewing's tumor of the jaw. Oral Surg Oral Med Oral Pathol 1990; 69:120-127; https://doi.org/10.1016/0030-4220(90)90280-6

11. Gorospe L, Fernández-Gil MA, García-Raya P, Royo A, López Barea F, García-Miguel P. Ewing`s sarcoma of the mandible: radiologic features with emphasis on magnetic resonance appearance. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2001; 91: 728-734; https://doi.org/10.1067/moe.2001.113546

12. Quesada JL, Alcalde JM, Espinosa JM, García-Tapia R. Ewings' sarcoma of the mandible. J Laryngol Otol 2003;

117:736-738; https://doi.org/10.1258/002221503322334639
13.Talesh KT, Motamedi MH, Jeihounian M. Ewing's sarcoma of the mandibular condyle: report of a case. J Oral Maxillofac Surg 2003; 61:1216-1219; httsp://doi.org/10.1016/S0278-2391(03)00686-4
14.Solomon LW, Frustino JL, Loree TR, Brecher ML, Alberico RA, Sullivan M. Ewing's sarcoma of the mandibular condyle: Multidisciplinary management optimizes outcome. Head Neck 2008; 30: 405-410; https://doi.org/10.1002/hed.20692
15. Folpe AL, Hill CE, Parham DM, O'Shea PA, Weiss SW. Immunohistochemical detection of FLI-1 protein expression: a study of 132 round cell tumors with emphasis on CD99-positive mimics of Ewing's sarcoma/primitive neuroectodermal tumor. Am J Surg Pathol 2000 ; 24(12):1657-62; https://doi.org/10.1097/00000478-200012000-00010

****

Conflict of interests: None declared
Source of funding: Nil
Date of submission: 03-04-2017
Date of acceptance: 18-04-2017

Authors details:

1. **Corresponding author:** M.D.S. Post-graduate student, Department of Oral Pathology and Microbiology, S.C.B. Dental College and Hospital, Cuttack- 753007, Odisha, India; Email: punyasloka.pati@gmail.com
2. Associate Professor**,** Department of Oral Pathology and Microbiology, S.C.B. Dental College and Hospital, Cuttack, Odisha
3. M.D.S. Post-graduate student, Department of Oral and Maxillofacial Pathology, S.C.B. Dental College and Hospital, Cuttack, Odisha
4. Assistant Professor**,** Department of Oral Pathology and Microbiology, S.C.B. Dental College and Hospital, Cuttack , Odisha