University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 2019, Vol. 5(3)

Extra skeletal ewings sarcoma of sinonasal tract-A rare case report.

DHIVYA S
Department of OTO-RHINO-LARYNGOLOGY,MADURAI MEDICAL COLLEGE AND HOSPITAL

Abstract : Extraskeletal ewings sarcoma is rarely found arising in the head and neck region. Involvement of facial bones and paranasal sinuses are exceptionally rare. In literature very few cases have been reported (ref 1-6).Here we present a case of ewings sarcoma of maxillary sinusand nose in a 28 year old male. Based on radiological imaging and histopathological analysis, the diagnosis of ewings sarcoma was made. The patient underwent a combined modality of treatment which included surgery and chemoradiotherapy.

Keyword :"Extraskeletal" , "ewings sarcoma", " Paranasal sinus","Rare"

INTRODUCTION :

Ewing's sarcoma first described in 1921 by James Ewing as a 'diffuse endothelioma of bone' is highly malignant, small round cell tumor, originating from primitive neuroectodermal cells. It is a locally aggressive tumor occurring predominantly in males during first three decades. Ewing's sarcoma have both skeletal and extraskeletal forms. Skeletal forms are commonly seen arising from mid shafts and metaphysis of long bones like femur, tibia, humerus and fibula. Extraskeletal forms usually arise from soft tissues of lower extremities and paravertebral region ,chest wall, retroperitoneum and rarely in head and neck(ref 7). Extraskeletal forms involving paranasal sinuses is a rare entity and poses significant diagnostic challenges. However with combined radiological, histopathological, immunohistochemistry tools it can be well differentiated from other round cell tumors.

CASE REPORT:

A 28 year old male patient came with complaints of left sided nasal obstruction for two months with history of intermittent episodes of epistaxis. There was associated headache and hyposmia. On examination, the external contour of nose was normal and there was a mild swelling over the left maxillary region associated with tenderness. On anterior rhinoscopy examination there was a proliferative mass occupying the left nasal cavity(figure1)

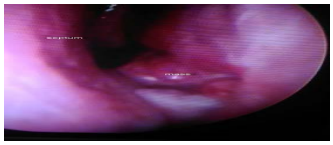

figure 1-Proliferative mass in Lt.nasal cavity

On diagnostic nasal endoscopy the mass was occupying the inferior and middle meatus of left nasal cavity .The coana was free. The right nasal cavity examination was within normal limits. In CT-PNS a soft tissue density was noted in the left maxillary sinus and nasal cavity with bony destructions noted in the inferior wall of maxillary sinus and floor of orbit(figure 2)

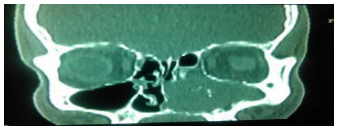

figure 2-soft tissue density in lt.maxillary sinus and nasal cavity

The biopsy taken from the mass was sent for histopathological examination which was reported as ewing's sarcoma. Immunohistochemistry was carried and the tumor was CD-99 and vimentin positive(figure 3). The patient underwent left total maxillectomy with post operative adjuvant chemoradiotherapy. The chemotherapy comprised the VAC regimen consisting vincristine(1.2 mg\meter square),cyclophosphamide (600mg\meter square),adriamycin (50mg\meter square) for six cycles once in twenty days. The radiotherapy dosage was 6000cGY , given in divided fractions,200cGY per fraction using intensity modulated radiotherapy(IMRT) in linear accelerator.

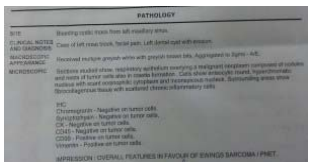

figure 3 -Histopathological report

DISCUSSION:

Ewing's sarcoma and primitive neuroectodermal tumor (PNET) have been unified into a single category called ewing's sarcoma family tumor(ESFT) as they show similar clinical ,morphological, biochemical, and molecular features although PNET shows more neuroectodermal differentiation than ewing's sarcoma. The extraskeletal form of ewing's sarcoma involving head and neck region is just 2-7%.Maxilla and mandible are most common sites (ref 6).

molecular genetics:

Majority of the patients have t(11;22)(q24;q12), i.e, fusion between the 5' end of the EWS gene from chromosome band 22q12 with the 3' portion of the 11q24 FLI1 gene, a member of the ETS family of transcription factors. This EWS/ETS fusion protein blocks the differentiation of pluripotent marrow stromal cells. Rest 10-15% of the cases have t(21;22)(q22;q12) fusing EWS to a closely related ETS gene, ERG from chromosome band21q22. In less than 1% of cases, t(7;22), t(17;22), t(2;22) and inv(22) have been found that give rise to fusions between EWS and the ETS genes like ETV1, E1AF, FEV, and ZSG, respectively. Mutations associated with P53 or P16/p14 ARF have high aggressive behavior and poor chemotherapeutic response(ref 7,11).

microscopic appearance:

Tumor macroscopically appears soft, tan-white, and contains areas of hemorrhage and necrosis. Microscopically the tumor is composed of sheets of uniform small round cell ,slightly larger and more cohesive than lymphocyte. The cells have scant cytoplasm ,which may appear clear;(figure 4)

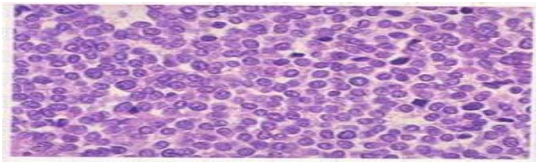

figure- 4 histopathological image

The presence of homer-wright rosettes(round groupings of cells with a central fibrillary core) indicates greater degree of neuroectodermal differentiation. Immunohistochemistry markers help to differentiate ewing's sarcoma from other small round cell tumors. The markers include CD-99,FLI 1 and caveolin. In addition markers of neural differentiation like NSE,CD-57 are also present. The miscellaneous markers include desmin, vimentin. In recent years CD 133 have been isolated from ewing's sarcoma family tumors that are more resistant to standard chemo-radiation therapies and are considered as a marker for chemoresistance (ref 11).

differential diagnosis:

The differential diagnosis for round cell tumors include olfactory neuroblastoma , sinonasal melanoma, embryonal rhabdomyosarcoma, mesenchymal chondrosarcoma, small cell osteosarcoma, and lymphoma.

olfactory neuroblastoma:

In Olfactory neuroblastoma , the prominent fibrillary or reticular background with a strong positivity for NSE and keratin differentiates it from ewing's sarcoma.

sinonasal melanoma:

In Sinonasal melanoma the growth itself is pigmented and microscopically the cells show nesting growth pattern and stains positive for HMB-45.

embryonal rhabdomyosarcoma

It is common in children and microscopically shows small nucleoli distributed in the sub epithelial layer(cambium layer) with presence of rhabdomyoblasts and stains positive for MYO D1.

lymphoma:

Lymphomas involving sinonasal tracts are commonly non-hodgkin type and are usually lymphoblastic and can be ruled out by absence of prominent nucleoli and CD 45 negativity.

mesenchymal chondrosarcoma:

Mesenchymal chondrosarcoma and small cell osteosarcoma were ruled out due to absence of either cartilage or osteoid tissue and most importantly FLI1 immunopositivity.

diagnosis and treatment:

Although small round cell tumors of the sinonasal tract pose significant diagnostic challenges, majority can be diagnosed by light microscopy. However, some cases need ancillary methods like immunohistochemistry and molecular genetics(discussed already) for definitive diagnosis. The CT findings are regardless of primary site and it is a well enhancing soft tissue density mass without calcification. When there is necrosis or hemorrhage , it appears heterogenous. Invasion of subcutaneous tissue and bony destruction are common (ref 9)(figure 2). In MRI the tumor appears hypointense to isodense on T1W1 and varying signal intense on T2W1.It is markedly enhanced by gadolinium. European intergroup co-operative ES study protocol recommends 14 cycles of etoposide, vincristine ,actinomycin D, ifosfamide and adriamycin. According to this protocol, chemotherapy is repeated every three weeks (1 cycle) and in each cycle either adriamycin or actinomycin D is used alternatively. In case of insufficient bone marrow recovery (white blood cell count <2.0 ×109/l or platelets <80 ×109/l), the next cycle of chemotherapy is postponed and granulocyte-colony stimulating factor (G-CSF) was added to subsequent cycles. Chemotherapy was interrupted for surgery which includes wide excision of the tumor after four to six cycles followed by radiotherapy. If histological examination of a radically resected tumor revealed more than 10% of viable tumor cells, radiotherapy was also administered postoperatively (ref 10). Since in our case the tumor was surgically resectable the patient underwent surgical excision of the tumor(total maxillectomy) followed by adjuvant chemoradiotherapy consisting vincristine, Adriamycin and cyclophosphamide for six cycles and radiotherapy of 60GY to the tumor site.

prognosis:

Important prognostic factors include age of the patient, stage, anatomic location and size of the tumor. Patients younger than 15 have a better outcome. The five-year survival is 55% without metastases and it reduces to 22% with metastasis. Pediatric studies have also reported a better prognosis for patients with lung metastases as compared with those of bone metastases. Progressive or recurrent disease has a poor outcome, despite aggressive therapy including high dose chemotherapy and peripheral stem cell transplantation(ref 11).

conclusion:

This case of extraskeletal ewing's sarcoma of maxillary sinus is presented due to its rarity and a
careful evaluation and knowledge of microscopic features with ancillary techniques like
immunohistochemistry and cytogenetics will help in accu-rate diagnosis and appropriate management
of the patient.

REFERENCES:

1.Primary extraskeletal ewing's sarcoma of maxillary sinus with intraorbital extension. (PMID:23120451) Varshney S, Bist SS, Gupta N, Bhatia R.Indian J Otolaryngol Head Neck Surg [2007].

2. Primary soft tissue Ewing's sarcoma of the maxillary sinus in elderly patients: presentation, management and prognosis. (PMID:24442190) Dutta M, Ghatak S, Biswas G, Sen A. Singapore med j(2014).

3.Ewing's sarcoma of maxilla: A rare case report.(PMID:25328307) Jairamdas Nagpal DK, Prabhu PR, Palaskar SJ, Patil S.J Oral Maxillofac Pathol [2014]
4 Ewing's family of tumors of the sinonasal tract and maxillary bone. (PMID:21107767) Hafezi S, Seethala RR, Stelow EB, Mills SE, Leong IT, MacDuff E, Hunt JL, Perez-Ordonez B, Weinreb I.Head Neck Pathol [2011]
5.Peripheral primitive neuroectodermal tumour of the left maxillary sinus:Isam Alobid , Manuel Bernal- Sprekelsen , Llucia Alós , Pedro Benítez , Jose Traserra , Joaquim Mullol Acta Oto-Laryngologica Vol. 123, Iss. 6, 2003.
6. Primitive Neuroectodermal Tumor of the Head and Neck: Incidence, Diagnosis, and Management- Jochen P. Windfuhr, MD, Dept of Otorhinolaryngology–Plastic Head and Neck Surgery, St Anna Hospital, 47259 Duisburg, Germany.
7. Yeshvanth SK, Ninan K, Bhandary SK, Lakshinarayana KH, Shetty JK, Makannavar JH. Rare case of extraskeletal Ewings sarcoma of the sinonasal tract. J Can Res Ther 2012;8:142-4.
8. Alfio Ferlito (1978). Primary Ewing's sarcoma of the maxilla: a clinicopathological study of four cases. The Journal of Laryngology & Otology, 92, pp 1007-1024. doi:10.1017/S0022215100086436.
9. Primary Ewing's sarcoma of the paranasal sinus with intracranial extension: imaging features M Harman, F Krolu, M Kösem, and Ö Ünal Dentomaxillofacial Radiology 2003 32:5, 343-346.
10. Wexler, L. H., Kacker, A., Piro, J. D., Haddad, J. and Close, L. G. (2003), Combined modality treatment of Ewing's sarcoma of the maxilla. Head Neck, 25: 168–172. doi: 10.1002/hed.10156.
11.Alfredo Pinto, Paul Dickman, and David Parham, “Pathobiologic Markers of the Ewing Sarcoma Family of Tumors: State of the Art and Prediction of Behaviour,” Sarcoma, vol. 2011, Article ID 856190, 15 pages, 2011. doi:10.1155/2011/856190

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Surgery and Surgical Specialities