International Journal of Otorhinolaryngology and Head and Neck Surgery
Satish D et al. Int J Otorhinolaryngol Head Neck Surg. 2018 Jan;4(1):304-307
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

Case Report

DOI: http://dx.doi.org/10.18203/issn.2454-5929.ijohns20175649

A rare case of Ewing's sarcoma in the sinonasal tract

Deepthi Satish*, Nanadakumar R., Balasubramanya A. M., Nitty Mathew

Department of ENT, St John's Medical College hospital, Bangalore, Karnataka, India

Received: 16 October 2017
Revised: 23 November 2017
Accepted: 24 November 2017

***Correspondence:**
Dr. Deepthi Satish,
E-mail: deepthishashikanth@gmail.com

ABSTRACT

Ewing's sarcoma (ES) first described by James Ewing in 1921, is a primary neoplasm of the skeletal system. Extraskeletal Ewing's sarcoma (EES) is a rare, rapidly growing, round cell malignant tumour that can develop in the soft tissue at any location. Extraskeletal Ewings sarcoma of the head and neck is uncommon. Nearly 80% of patients are younger than 20 years with peak incidence in the second decade. Ewings sarcoma rarely affects the sinonasal tract. Diagnosis is after histopathological examination immunohistochemical studies and cytogenetic studies. Treatment will include a multidisciplinary approach with surgery as the first line followed by chemotherapy and radiotherapy. Ewings sarcoma in the head and neck region do not metastazise early hence carries a better prognosis. Evaluation of lesion using imaging, biopsy followed by histopathology and immunohistochemistry and cytogenetic analysis are necessary for early diagnosis and treatment. We present a case of an 18-year-old male patient with left sided epistaxis and left sided nasal obstruction, on examination a mass was seen the nasal cavity, endoscopic excision and biopsy of the mass was suggestive of an Ewings sarcoma. He received post-operative chemotherapy and radiotherapy. 2 years on regular follow up patient is fine without signs of metastasis or recurrence. We are presenting this case due to the rarity of its presentation in the sinonasal tract.

Keywords: Ewings sarcoma, Extraskeletal Ewing's sarcoma, Ewings family tumours

INTRODUCTION

Ewing's sarcoma (ES), is a primary neoplasm of the skeletal system. Extraskeletal Ewing's sarcoma (EES) is rare, it usually occurs in the soft tissue of lower extremities, paravertebral tissues, chest wall, retroperitonium and rarely in the head and neck region, primary sinonasal ES is even rarer and represents only a small subset of these head and neck cancers. The diagnosis of these entities requires a histopathological examination, immunohistochemistry, and a cytogenetic analysis along with a contrast enhanced CT scan of the paranasal sinuses. The effective treatment for ES includes combined surgical excision and chemotherapy/ radiotherapy.[1] We report a case of extraskeletal ES of the sinonasal tract in an 18 yr old male who presented with nasal obstruction and epistaxis. The patient was treated with surgery followed by chemotherapy and radiotherapy.

CASE REPORT

An 18 year old male patient presented to ENT OPD of St John's medical college hospital a tertiary referral centre in Bengaluru, South India with a history of left sided epistaxis of five months duration associated with left sided nasal block. On anterior rhinoscopy a fleshy reddish mass was seen in the left nasal cavity.

A contrast enhanced computed tomography scan of nose and paranasal sinuses done in our hospital showed a 2 cm ×3 cms heterogeneously enhancing soft tissue lesion in the floor of the left nasal cavity extending into the left maxillary sinus and eroding the medial and inferior wall

of left maxillary sinus, erosion of the inferior turbinate and left half of hard palate was also noted. A differential diagnosis of juvenile nasopharyngeal angiofibroma was made considering the patient's age, history and the imaging findings (Figure 1).

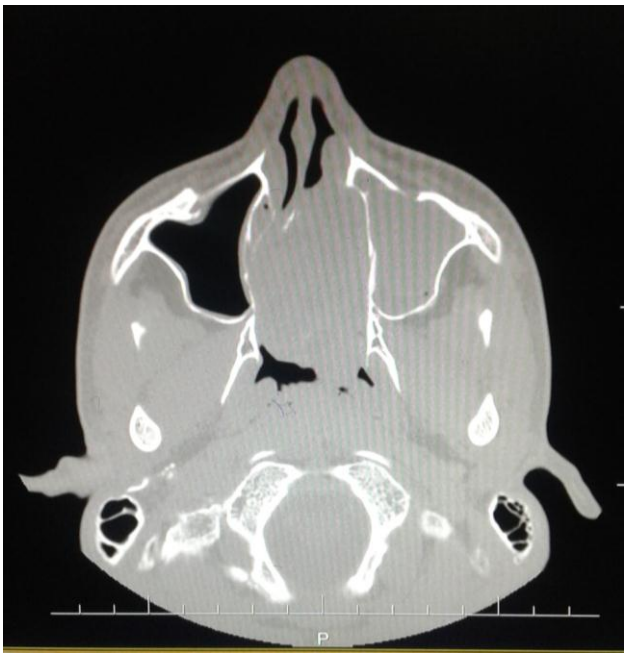

Figure 1: CT with contrast of nose and PNS showing an irregular, well defined, soft tissue lesion with heterogeneous enhancement in the left nasal cavity. There is extension into the left maxillary sinus with erosion of the medial and inferior wall of the maxillary sinus and erosion of the bony nasal septum.

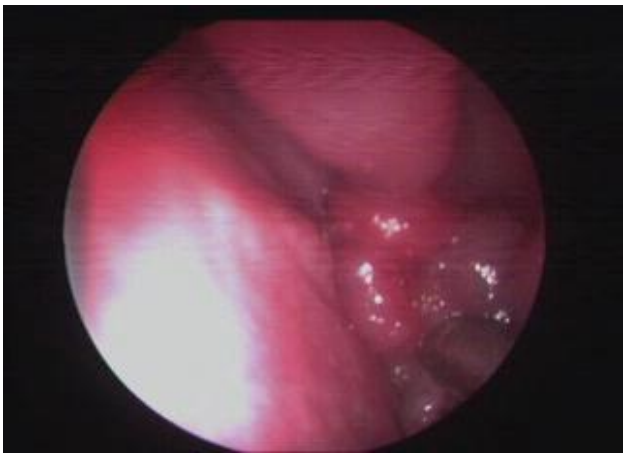

Figure 2: Intraoperative picture showing a bluish red polypoidal mass in the left nasal cavity arising from the left maxillary sinus.

Endoscopic excision and biopsy of the mass was planned after embolisation. Intraoperatively a reddish mass was seen in the left nasal cavity arising from the left maxillary sinus extending to the floor, a wide middle meatal antrostomy was done. The mass was vascular and dark fluid was seen extruding out of the mass. Subtotal excision of the mass was done and sent for biopsy (Figure 2).

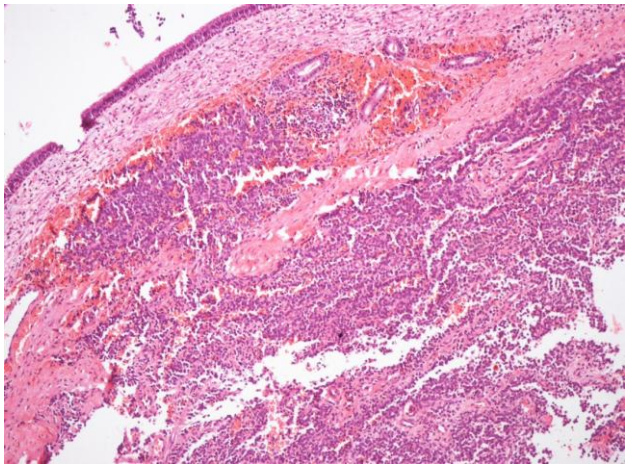

Figure 3: Fragments of nasal mucosa with an infiltrating neoplasm composed of small round cells arranged in sheets.

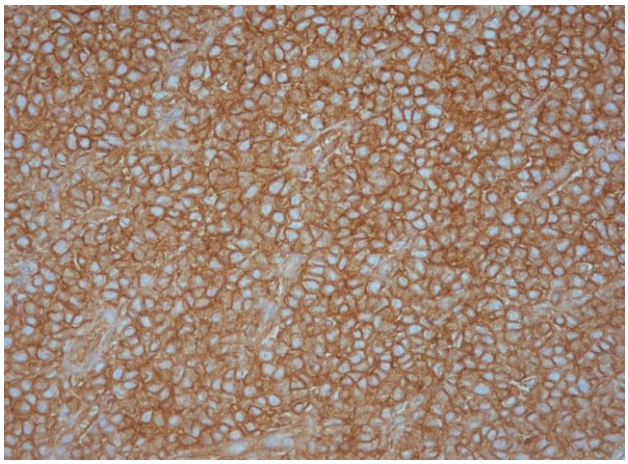

Figure 4: CD 99 immunostain showing membranous positivity in the neoplastic cells.

On histopathological examination fragments of nasal mucosa with an infiltrating neoplasm composed of small round cells arranged in sheets were seen and on immunohistochemistry the neoplastic cells expressed CD99 (3+, 100%). A FISH study revealed that the sample was positive for ESWR translocation t (11; 22), with this immunoprofile a diagnosis of Ewings sarcoma was made. (Figure 3 and 4).

Patient was referred to the department of paediatric oncology for further management. A PET CT was done which showed a non FDG avid heterogeneously enhancing mass lesion with epicentre in the left maxillary sinus eroding its medial wall and inferiorly eroding into the hard palate and left maxillary alveolus (Figure 5).

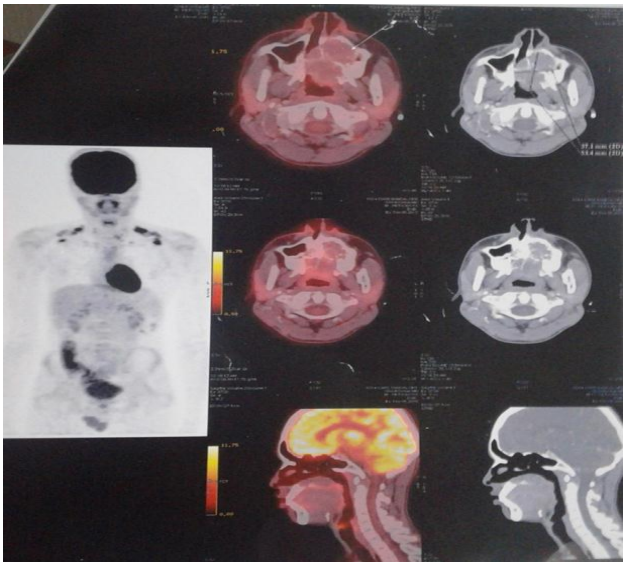

Figure 5: PET CT showing a non FDG avid primary tumour in the left nasal cavity with local extension and infiltration. No definite regional nodal or distant metastasis.

He was started on chemotherapy (Vincristine, Adriamycin, cyclophosphamide and actinomycin) alternating with ifosfamide and etoposide (VACA/IE) with radical radiation therapy on linear accelerator (total 55.80Gy in fractions over 6 weeks). He completed his treatment and is on regular follow up, 2 years post operatively patient is asymptomatic with no signs of metastasis or recurrence.

DISCUSSION

Ewing's sarcoma (ES) first described by James Ewing in 1921, is a primary neoplasm of the extra skeletal Ewings sarcoma (EES).[2] EES is a rare, rapidly growing, round cell malignant tumour that can develop in the soft tissue at any location.

Primary Ewings sarcoma of the head and neck is uncommon. Amongst bone Ewings sarcoma, the head and neck (skull) accounts for 3.8% of cases.[3] Primary sinonasal ES is even rarer and represents only a small subset of these head and neck cancers.[4] In the sinonasal tract the various differential diagnosis of small round cell tumours are Rhabdomyosarcoma, Lymphoma, Poorly-differentiated carcinomas, Melanoma, Olfactory neuroblastoma (ONB) and Ewings family tumours (EFT).[5]

Among these tumors, EFTs are rare in this location and have not been extensively reported in the literature.

It is difficult to differentiate ES from similar tumours arising from the sinonasal tract based on clinical and radiological examination alone hence it requires a histopathological examination, immunohistochemistry, and a cytogenetic analysis to make the diagnosis. The essential diagnostic examination to differentiate ES from the many small round neoplasms is the CD99 marker, which can be detected in a specific immunohistochemical examination.[6] However rhabdomyosarcoma (RMS) and Lymphoma can also show positivity for CD99, hence further testing with CK, S100 and Desmin are required for which ES is negative which further differentiates it from RMS and Lymphoma. Nearly 80% of patients are younger than 20 years with peak incidence in the second decade. There is male preponderance with bone being the primary site in 60% of cases.

Patients usually present with symptoms of nasal obstruction and epistaxis. A contrast enhanced CT scan is used in the radiological evaluation of ES of maxillary sinus, which shows an enhancing soft tissue mass with bone destruction. Usually the ES shows a characteristic radiographic picture described by some authors as “onion skin appearance” especially in the long bones, but such pattern is less commonly seen in the sinuses.

The effective treatment for ES includes combined surgical excision and chemotherapy/radiotherapy which has increased the five-year survival rate. The recommended protocol consists of 14 cycles of Etoposide, Vincristine, Actinomycin D, Ifosfamide and Adriamycin.[7] According to this protocol, chemotherapy is repeated every three weeks (1 cycle) and in each cycle either Adriamycin or Actinomycin D is used alternatively. Due to advances in the treatment of Ewings sarcoma in recent years, survival rates are reported to have improved and have reached up to 86% in patients with non-metastatic disease at initial presentation. Those with metastases have poorer outcomes.[8]

CONCLUSION

Ewings sarcoma rarely affects the sinonasal tract. Diagnosis is after histopathological examination immunohistochemical studies and cytogenetic studies. Treatment will include a multidisciplinary approach with surgery as the first line followed by chemotherapy and radiotherapy. Ewings sarcoma in the head and neck region do not metastazise early hence carries a better prognosis.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Yeshvanth SK, Ninan K, Bhandary SK, Lakshinarayana KPH, Shetty JK, Makannavar JH. Rare case of extraskeletal Ewings sarcoma of the sinonasal tract. J Cancer Res Therap. 2012;8(10):142-4.
2. Gupta S, Gupta OP, Mehrotra S, Mehrotra D. Ewing sarcoma of maxilla: A rare presentation. Quintessence Int. 2009;40:13.

3. Cotterill SJ, Ahrens S, Paulussen M, Ju¨rgens HF, Vouˆte PA, Gadner H, et al. Prognostic factors in Ewing's tumor of bone: analysis of 975 patients from the European Intergroup Cooperative Ewing's Sarcoma study group. J Clin Oncol. 2000;18(17):3108–14.
4. Wenig BM, Dulguerov P, Kapadia SP, Prasad ML, Fanburgsmith JC Thompson LD. Neuroectodermal tumors. In: Barnes EL, Eveson JW, Reichart P, Sidransky D, editors. World Health Organization classification of tumours. Pathology and genetics of head and neck tumours Lyon. France: IARC Press; 2005: 65–70.
5. Wenig BM. Undifferentiated malignant neoplasms of the sinonasal tract. Arch Pathol Lab Med. 2009;133(5):699–712.
6. Suzuki T, Yasumatsu R, Nakashima T et al. Primary Ewing's Sarcoma of the Sinonasal Tract: A Case Report, Case Rep Oncol. 2017;10(1):91-7.
7. Hafezi S, Seethala RR, Stelow EB, Mills SE, Leong IT, MacDuff E, et al. Ewing's family of tumors of the sinonasal tract and maxillary bone. Head Neck Pathol. 2011;5:8-16.
8. Raney RB, Asmar L, Newton WA Jr, Bagwell C, Breneman JC, Crist W, et al. Ewing's sarcoma of soft tissues in childhood: a report from the Intergroup Rhabdomyosarcoma Study, 1972 to 1991. J Clin Oncol. 1997;15(2):574–82.

Cite this article as: Satish D, Nanadakumar R, Balasubramanya AM, Mathew N. A rare case of Ewing's sarcoma in the sinonasal tract. Int J Otorhinolaryngol Head Neck Surg 2018;4:304-7.