Atypical presentation of Ewing's sino-nasal tumor: case report

Shweta Sharma[1], Shantnu Singh Chauhan[2], Mir Rizwan Aziz[2], Nalin Chugh[2], Bharat Bhushan Sharma[2]*, Nitish Virmani[3]

European Journal of Medical Case Reports
Volume 3(1):37–40

https://doi.org/10.24911/ejmcr/173-1539503065

ABSTRACT

Background: Ewing's family tumors are malignant and occur in children and young adults. Sinonasal region is slightly rare in occurrence and can present in various types of symptomatology. Ewing sarcoma and primitive neuroectodermal tumor are the most common entities in this family. The preliminary diagnosis is almost always confused because of delayed symptoms and improper histopathological and immunohistochemistry (IHC). Most of the complaints are because of tumor mass pressing and displacing the neighboring structures.

Case Presentation: A 36-years-old male adult reported with progressively increasing facial swelling on the bridge of the nose and forehead, predominantly on the left side of 3 years duration. There was pain and blockage of left side nose first followed by the other side. There were also off and on episodes of epistaxis. Ultrasonography and color flow imaging confirmed the swelling of solid consistency with hypervascularity. The patient underwent contrast-enhanced computerized tomography and magnetic resonance imaging studies which delineated the entity which was confirmed by histopathology.

Conclusion: Sinonasal masses require detailed cross-sectional imaging studies, histopathological and IHC confirmation for proper and in time management strategies.

Keywords: Ewing's family, PNET, immunohistochemistry, CFI, CECT, MRI.

Received: 14 October 2018 **Accepted:** 21 December 2018

Type of Article: CASE REPORT

Funding: None

Declaration of conflicting interests: None

Specialty: Radiology

Correspondence Author: Bharat Bhushan Sharma
*Department of Radiodiagnosis, SGT Medical College, Gurgaon, India.
Email: bbhushan986@gmail.com
Full list of author information is available at the end of the article.

Background

The incidence of sino-nasal tumors is around 3% of the upper respiratory tract tumors. These are more common in the white population and have male predominance. Sino-nasal Ewing's family tumors are rare in occurrence in head and neck region. The presence of these tumors in the sino-nasal region is a rarity. Ewing's family of tumors constitutes Ewing sarcoma (ES) and primitive neuroectodermal tumors (PNET). Arthur Stout in 1918 described PNET in the ulnar nerve. It was James Ewing who described this tumor and was named after his name as ES. Now, these tumors are considered as different entities because of differences in histology, immunohistochemistry (IHC), and molecular biology. This issue is important for the diagnosis and follow-up point of view [1]. Previously, many cases of ES had been described but without molecular confirmation and long-term follow-up.

Case Presentation

A 36-years-old male presented to the Otorhinolaryngology outpatient department with a complaint of pain, mouth breathing, diplopia, and face swelling over the bridge of the nose and the left infraorbital region of 1-year duration. The swelling had progressively increased in size over the past 3 years. Now, he had reported because of the pain and difficulty in breathing through the nose. There were also frequent episodes of epistaxis from the same side in the last 3 months. Superficial examination, it revealed swelling 5.8 cm × 3.7 cm over the bridge of the nose which extended to the left eye and infraorbital region. The overlying skin was of reddish in coloration but without any ulceration (Figure 1).

Local examination had shown soft fleshy mass blocking the left nostril. Right nostril was patent. Systemic examination was unremarkable. Oral cavity and oropharynx were without any other pathology. Laboratory investigations were within normal limits. Ultrasonography (USG) revealed soft tissue hypoechoic mass with increased vascularity with few regions of necrosis (Figure 2).

Patient was subjected to plain and contrast-enhanced computerized tomography (CECT) for further evaluation. The mass was slightly hypodense, heterogonous in nature and showed post-contrast enhancement

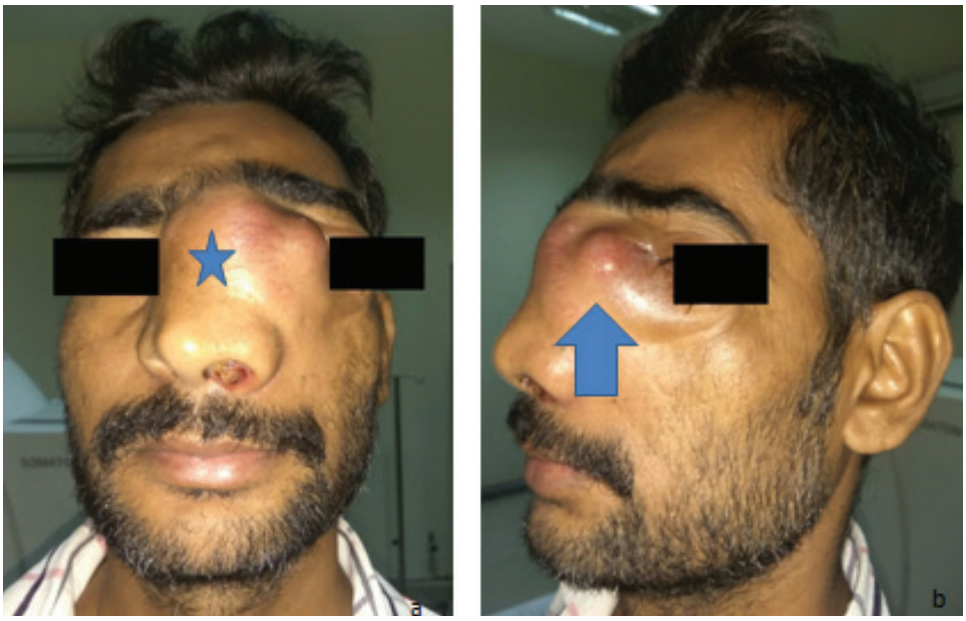

Figure 1. *Photograph of a 36-years-old male in enface (a) and profile (b) views. The swelling is evident at the bridge of the nose extending to the left infraorbital region. There is pink coloration over the swelling.*

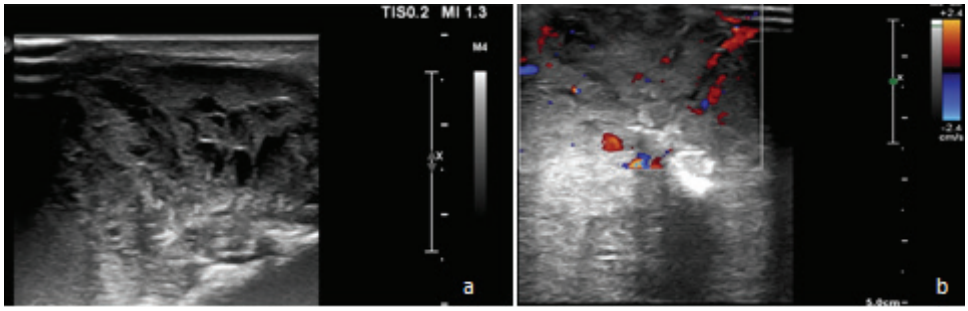

Figure 2. *Ultrasound examination. (a) Grayscale image shows slightly heterogenous mass with few necrotic areas. (b) The lesion shows increased vascularity within the mass.*

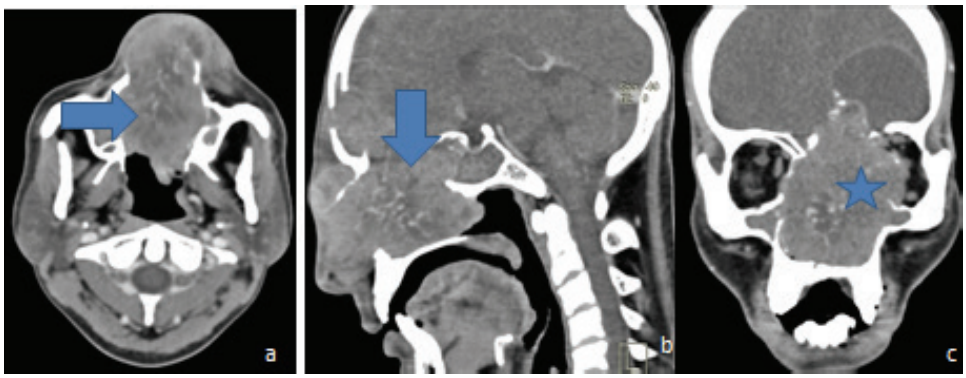

Figure 3. *CECT of the neck and facial region. (a) Axial section shows the enhancing mass in the left sino-nasal region (horizontal arrow) which is pressing upon the adjoining structures. (b) Sagittal section reveals the same mass showing extension in the base of the skull (vertical arrow) and posteriorly up to the sphenoid sinus. There is a total involvement of ethmoid and sphenoid regions. (c) Well-marginated enhancing mass with visible vessels (star). The mass is causing displacement of the left orbit laterally.*

(Figure 3). The bone window revealed expansion of the tumor causing thinning out of the bone with partial destruction (Figure4).

Magnetic resonance imaging (MRI) was performed to know more about the characterization of the mass. The mass was well-defined and of mixed intensity (Figure 5).

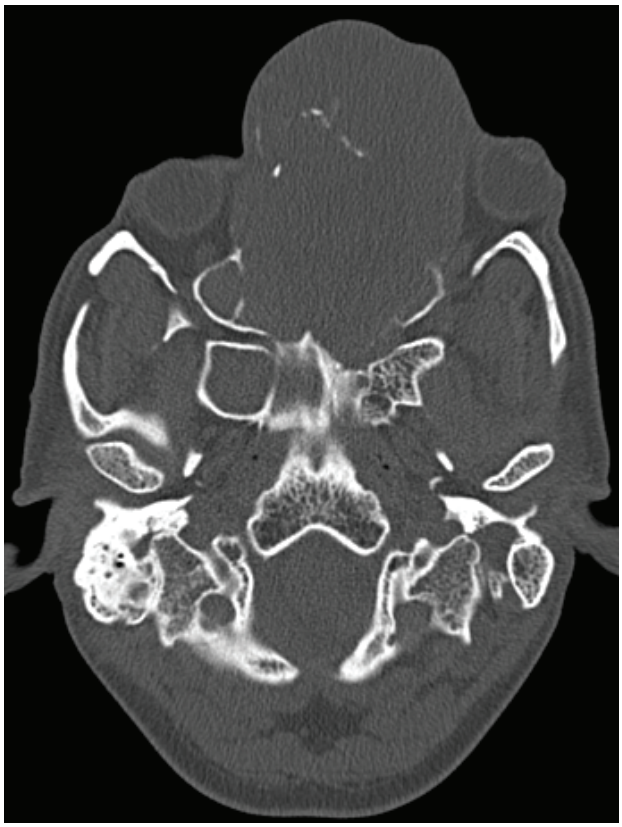

Figure 4. *Axial section CT in the bone window at the tumor level. This shows the outstretched eaten up bone with the tumor expansion.*

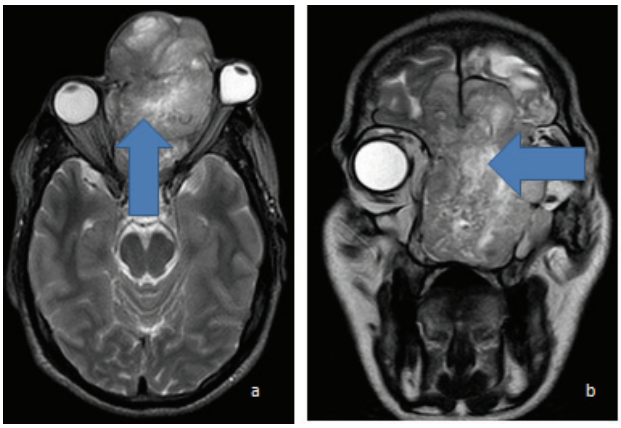

Figure 5. *MRI images at the level of the bridge of the nose. (a) Axial T2WI shows mixed intensity mass with a well defined outline (vertical arrow). The mass is displacing the left eye globe rather evading it. (b) Coronal T2WI shows the supero-inferior extent of the mass (horizontal arrow).*

The patient underwent complete excision of the mass under general anesthesia. The histopathological specimen revealed lobules of monomorphic tumor cells with occasional mitosis. The cells are immunopositive for MIC-2, while negative for synaptophysin, chromogranin, pancytokeratin; CD56 and myogenin. Findings were consistent with Ewing's sarcoma/peripheral PNET. This was followed by six cycles of Vincristine-Ifosfamide-Doxorubicin-Etoposide chemotherapy. The patient had an uneventful recovery and is on yearly follow-up.

Discussion

Malignant tumors with monomorphic population of undifferentiated cells with scanty cytoplasm and small nuclei form a heterogeneous group of small round blue cell neoplasms. Ewing sino-nasal is a high-grade round-cell tumor of childhood and adolescent with a predilection for male [2]. This has got higher malignant potential with both skeletal and extraskeletal manifestations. This gives the clinical appearance of osteomyelitis [3]. The manifestations in the head and neck regions are slightly rare but our present case had both skeletal as well as soft tissue involvement. The long bones are frequently involved. Soft tissue of lower extremities, paravertebral tissues, retroperitoneum, and chest wall are frequently involved. Maxilla and mandible are frequently involved as compared to sino-nasal region [4]. The extension of the tumor was assessed on the clinical features and computed tomography (CT). CECT is also helpful in evaluating for distant metastasis. Superficial tumors can also be assessed by USG and color flow imaging as in our present case. MRI is also helpful in some cases to know about the tissue characterization and asses the bleeding within the mass. Bone marrow biopsy and bone scintigraphy are also helpful in hidden metastatic foci. The approach is for the earliest accurate diagnosis and management. But unfortunately, the exact diagnosis made on H&E light microscope picture is also not the answer. This has to be coupled with IHC. The appearance is white in color with polypoidal and lobulated outline. It is mainly less than 10 cm in size. Histologically, this is in the form of densely packed small round cells in lobular form. There are intracellular deposits of glycogen which makes the cytoplasm as pale and ill-defined [5]. The differential diagnosis includes all small round blue cell pathologies. Glycogen in tumor cells can be highlighted with periodic acid-Schiff. In one study twenty-five percent of the patients had skeletal or visceral metastasis when diagnosed [6]. The management requires a long list depending upon the type of tumor. This extends from surgical excision followed by chemotherapy or radio-chemotherapy if the tumor is non-operable [7,8]. The age of the patient, stage, anatomic location, and size of the tumor had been taken as the prognostic factors as per the study conducted by Yeshvanth et al. The 5 years survival was 22% with metastasis and 55% without it [9].

Conclusion

ES of the sino-nasal region is rare with a wide spectrum and requires multi-modality diagnosis by cross-sectional imaging. This is required to be confirmed by histopathology and IHC. It is important to know about the extent of the tumor and to know if this had metastasized to some

other organs. The management solely depends upon the type of neoplasm.

Acknowledgment

We are thankful to Mr. Prahlad and Kasif for providing us the CT and MRI images and helping in conducting the investigations.

Ethical Approval

Not required

List of Abbreviations

CECT	Contrast-enhanced computerized tomography
CT	Computed tomography
ES	Ewing sarcoma
PNET	Primitive neuroectodermal tumors

Consent for publication

Written consent of the patient was taken.

Author details

Shweta Sharma[1], Shantnu Singh Chauhan[2], Mir Rizwan Aziz[2], Nalin Chugh[2], Bharat Bhushan Sharma[2], Nitish Virmani[3]

1. ENT Speciality registrar, Warwick Hospital Flat No 9.Lakin House, Warwick CV345BW UK
2. Department of Radiodiagnosis, SGT Medical College, Gurgaon, India
3. Faculty of Allied Health Sciences, SGT University, Gurgaon, India

References

1. Bridge JA, Bowen JM, Smith RB. The small round cell tumors of the sinonasal area. Head Neck Pathol. 2010;4(1):84–93. https://doi.org/10.1007/s12105-009-0158-6
2. Kleihues P, Sobin LH. World Health Oraganization classification of tumors. Cancer. 2000;88(12):2887. https://doi.org/10.1002/1097-0142(20000615)88:12<2887::AID-CNCR32>3.0.CO;2-F
3. Cotterill SJ, Ahrens S, Paulussen M, Jurgens HF, Voute 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. https://doi.org/10.1200/JCO.2000.18.17.3108
4. Patil AM, Yendigeri SM, Sajjanar BB, Kubadi SS. Ewing sarcoma/PNET of sinonasal region: a rare case report. Sci J. 2013;2(2):251–5.
5. Desai SS, Jambhekar N. A pathology of ewing's sarcoma/PNET current option and emerging concepts. Indian J Orthop. 2010;44(4):363–7. https://doi.org/10.4103/0019-5413.69304
6. Thompson LDR. Ewing sarcoma and primitive neuroectodermal tumor. ENT J. 2007;86(2):79.
7. Granowetter L, Womer R, Devidas M, Krailo M, Wang C, Bernstein M, et al. Dose-intensified compared with standard chemotherapy for non metastatic Ewing sarcoma family of tumors :a Children's Oncology Group Study. J Clin Oncol. 2009,27(15):2536–41. https://doi.org/10.1200/JCO.2008.19.1478
8. Grier HE, Krailo MD, Tarbell NJ, Link MP, Fryer CJH, Pritchard DJ, et al. Addition of ifosamide and Etoposide to standard chemotherapy for Ewing's sarcoma and primitive neuro-ectodermal tumor of bone. N Engl J Med. 2003;348(8):694–701. https://doi.org/10.1056/NEJMoa020890
9. Yeshvanth SK, Ninan K, Bhandary SK, Lakshinarayana KP, Shetty JK, Makannavar JH. Rare case of extraskeltal Ewings sarcoma of the sinonasal tract. J Cancer Res Ther. 2012;8(1):142. https://doi.org/10.4103/0973-1482.95197

Summary of the case

Patient	1	36-years-old male
Final diagnosis	2	Ewing's sino-nasal tumor
Symptoms	3	Swelling over the nose and epistaxis
Medications (Generic)	4	Symptomatic with post-operative chemotherapy
Clinical procedure	5	Surgical excision
Specialty	6	Radio-diagnosis