Case Report

Ewing's Sarcoma of the Nose

Muwafaq Algihiwi, [1] Nada Shahaltough, [1] Mohamd Ali Hiari, [1] Belal Elhawwari, [2] Samer Kharma [3] and Mahmoud Al-Raqqad [4]*

Abstract

Nasal bleeding is a common complaint familiar to all otolaryngologists. But, sino-nasal primary Ewing's Sarcoma is extremely rare. This is a case of a 16-year old male patient who suffers from epistaxis, in addition to his left eye proptosis. After examination, it has been revealed that he had a nasal mass in the left side of his nose. The case was managed initially as a vascular mass, but then, under surgery, a frozen section was subjected to examination using histology; the examination revealed the existance of malignant cells. After all, Definite histopathological diagnosis by immunohistochemical stains proved it to be Ewing's Sarcoma.

(J Med J 2006; Vol. 40 (4): 315- 320)

Received	Accepted
July 13, 2005	November 16, 2006

Introduction

Ewing's sarcoma is a rare malignancy as it represents only 5% of all childhood cancers; it mostly affects Caucasians, while extremely rare in blacks. Most commonly, it appears as undifferentiated primary bone tumour, less commonly arises in soft tissue (Extraosseous Ewing Sarcoma (EES)), both types are part of a spectrum of neoplastic diseases known as Ewing's sarcoma Family of Tumour (EFT).

Because these tumours share similar histological and immunohistochemical characteristics, in This family also includes: the more differentiated peripheral Primitive Neuroectodermal Tumour (PNET) that was previously called neuroepithelioma; adult neuroblastoma; malignant small cell tumour of the thoracopulmonary region; askins tumour; paravertebral small cell tumour and atypical Ewing's Sarcoma (ES). PNET can also appear in bone or soft tissue.

Radiological studies reported that there is a mass lesion that causes destruction to the medial wall

1. MD ENT JB.
2. MD Radiation Oncologist JB.
3. MD FRCOphth JB.
4. MD Orthopedics JB.
*** Correspondence should be addressed to:**
Muwafaq Algihiwi, MD
ENT Department, Royal Medical Services, King Hussein Medical Center, Amman- Jordan.

addition to unique nonrandom translocations (t (11;22),t (1;16)); they are considered to have a common origin. They also share common clinical features. Peak incidence occurs between ages 10-20, and 70% of all incidences occur under the age of 20. They have rapid metastasis to lungs, bone and bone marrow. Moreover, all of these tumours have Responsiveness to the same chemotherapeutic regimens and radiotherapy. [1- 5] We present a 16-year old male patient with Ewing's sarcoma of the nose who was initially suspected to have a vascular mass.

Case Report

A 16-year old male patient from Ma'an, admitted to our hospital on 23rd Feb. 2005, was referred from the OPD as a case of nasal mass.

His history started as a case of epistaxis of 4 months duration prior to his admission to the hospital, then, this was followed by a protrusion of his left eye of 2 months duration. Upon examination, he was found to have a mass filling the whole left nasal cavity causing left eye proptosis and limitation of its gaze in all directions, while the other cranial nerves were intact.

On the day of referral, the patient was admitted and biopsy was taken from the mass that seemed to be none vascular. Vision at that time was 6\6 in both eyes with mild left eye proptosis. Also, his kidney function's test was normal; liver enzymes normal apart from alkaline phosphatase 491u/l with the expected value (90-290u/l), total protein and albumin within the normal range, white blood cells' number was 9400, haematocrit percentage was 33% with microcytosis and platelets' number as 390000.

of the left maxillary sinus and lamina papyracea; this lesion extends to the medial left aspect of the orbit and engulfs the medial rectus muscle, causing by this deviation of the left optic nerve to lateral side and resulting in protrusion of the left eye ball. Appearances suggest that it is an aggressive lesion either infective or malignant (Fig. 1(a, b) and 2 (a, b)).

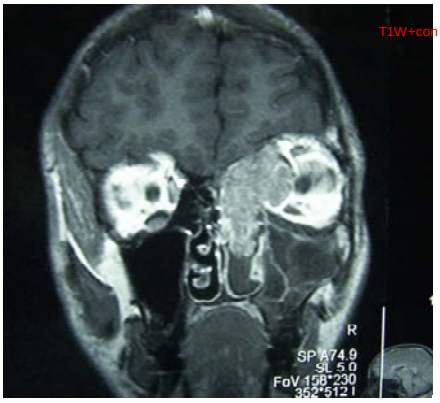

Fig. 1 (a).

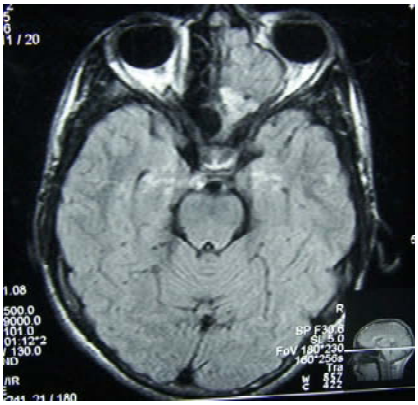

Fig.1 (b).

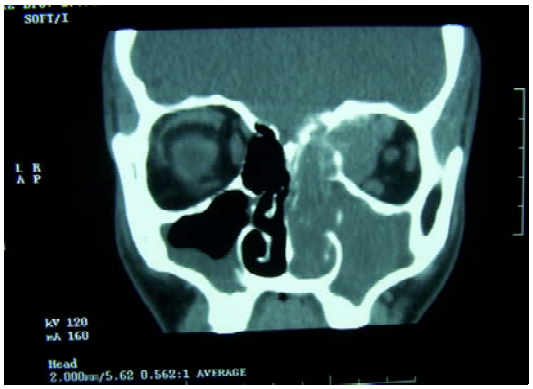

Fig. 2 (a).

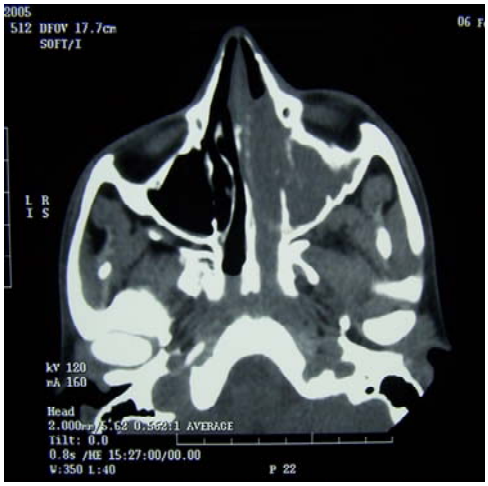

Fig.2 (b).

The patient was discharged. Few days latter, histopathology of the taken biopsy reported the mass to be an infarcted hemorrhagic benign lesion, so, angiography was planned and it was done showing an avascular mass.

After 3 weeks, we started with endoscopic removal of a soft tissue mass from the left nasal cavity, and there was spontaneous drainage of profuse purulent blackish discharge from the sinuses. Frozen section taken under surgery was sent and defined as malignant cells. The patient was discharged two days later.

Few days later, Histopathology report revealed Ewing's sarcoma, which was confirmed by immunohistochemical stains that were positive for vimentin, CD99, S-100, while negative for CK LCA, chromogranin and synaptophysin. Other stains were requested with a supplementary report issued with positive stain for NSE, negative stain for CD56 which is consistent with Ewing's / PNET sarcoma.

When the Patient came back a week later, he was sent to the haematology and oncology department, prior to that, ophthalmic assessment showed severe proptosis with optic nerve compression. The optic disc was swollen in the left eye and visual acuity was 6/6 in the right eye and 6/18 in the left, visual field examination was normal in the right eye and not possible in the left due to blurred vision. (Fig. 3 (a, b) and Fig. 4).

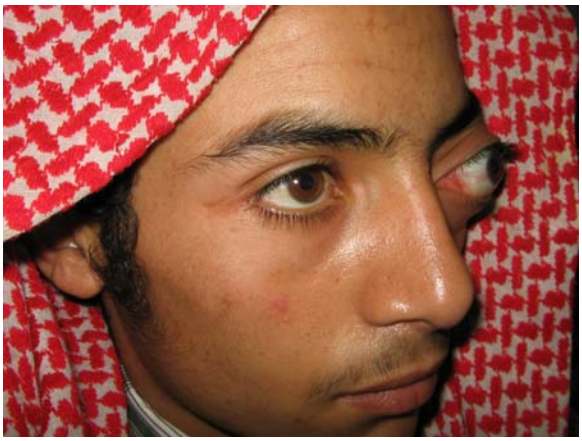

Fig. 3 (a).

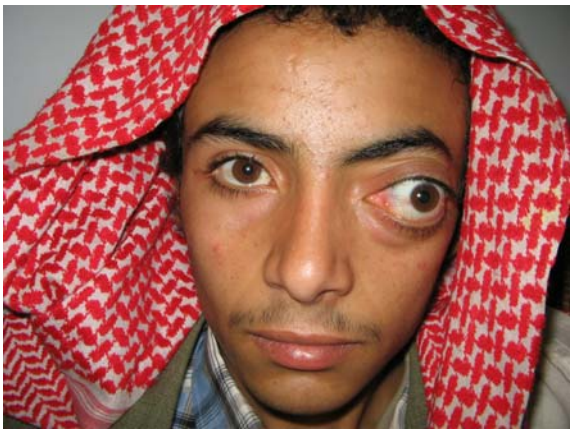

Fig. 3 (b).

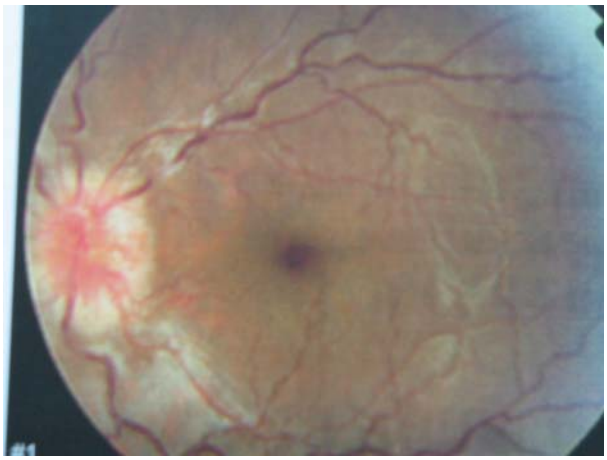

Fig. 4.

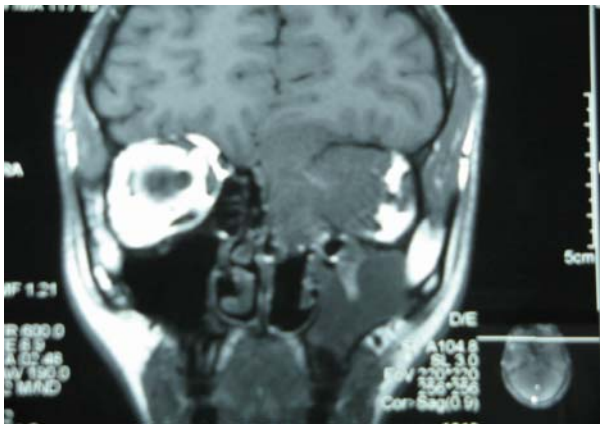

Fig. 5 (a).

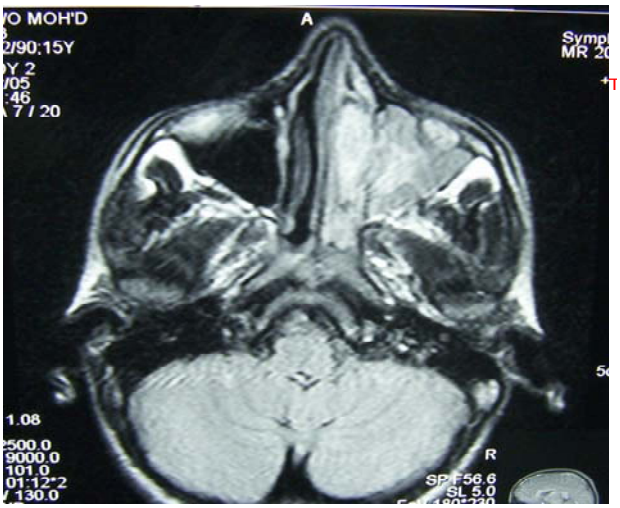

Fig. 5 (b).

He was, then, admitted urgently on the same day and given pulse therapy of methylprednisolone. Brain and orbit MRI showed a degree of progression of the tumour with intracranial extension (Fig. 5 (a, b)).

He was seen by the oncologist and radiotherapist and, according to that; he was planned for radiotherapy and chemotherapy. He received cyclophosphamide, Adriamycin, vincristine and mesna.

Despite having some improvement regarding his ophthalmic condition, the patient left the hospital against medical advice. He lost the follow up for 3 months, and then came back to proceed his chemotherapy, the tumour was controlled with no evidence of metastasis.

Discussion

Despite the rarity of the case, the very common presentation to the ENT clinic, which was epistaxis, had a very sinister pathology. The incidence in US is only 2.1 cases in one million children; but still, it is the second most primary tumour affecting children. It appears in any bone in the body, most frequently in the diaphysis of the long tubular bones and in the bones of the pelvis and, in only 2.3% of the cases, it develops on the face. [6] Diagnosis depends on the initial evaluation by plain films, CT and MRI scans, [7] following the biopsy microscopy shows sheets of small round blue cells with hyperchromatic nucleoli, scanty cytoplasm and poorly defined boundaries, nuclear atypia, palisading and formation of rosettes. [8]

In our case, the preliminary histological diagnosis was in favor of a vascular mass, but during surgery, being aware that this lesion could be malignant as the radiological studies were more conclusive towards that, a frozen section was sent. The presence of small round cells directed the diagnosis towards PNET; this was confirmed later by immunohistochemical stains.

The affected tissue in Ewing sarcoma is grossly described in most cases as grayish-white-soft and pus like, as it was apparent in the lesion of our case.

The differential diagnosis in children involves a large array of various poorly differentiated neoplasms of the sinonasal region. [7]

The essential diagnostic test is for the specific immunocytochemical CD99 or P30/32 MIC2 marker. [9, 10]

Modern treatment plans include: neoadjuvent chemotherapy followed by local control; either by radiotherapy or surgical resection of a potentially resectable tumor or both, then additional chemotherapy. With these treatment plans the 5 year survival have been raised to 56%. [11]

Prognosis depends primarily on the presence of metastasis at the time of diagnosis. Ewing's sarcoma have rapid metastasis to lungs, bone and bone marrow. With the majority of cases having subclinical metastasis and 25% having overt metastasis at the time of diagnosis, and if these are treated only locally, they have a relapse rate of 80-90%; making the 5 year survival decrease by 22%. [12, 13] Extraosseous Ewing sarcoma and PNET respond to the same chemotherapy regimens as Ewing sarcoma. [1]

Standard chemotherapy treatment options include: conventional dozes of Doxorubicin plus Cyclophosphamide or Vincristine, Doxorubicin and Cyclophosphamide with or without Actinomycin D (VDC or VDCA), or alternating courses of VDC and Isofosfamide plus Etoposide .None of these regimens had yet proven its superiority. [14]

Conclusion

Ewing's sarcoma is rare, but it is always possible that a seemingly more common condition can be an unusual pathology, so; there should always be a correlation between the clinical findings, radiological studies and the histopathological reports.

References

1. Smith MA, Gurney jg, Ries LA. Cancer in adolescents 15-19 years old. In: cancer incidence and survival among children and adolescents: United States SEER program 1975-1995(pub#99-4649).
2. Fraumeni JF, Glass AG. Rarity of Ewing's Sarcoma Among U.S Negro Children. Lancet 1970; 1: 366.
3. Delattre O, Zucman J, Melot T et al. The Ewing Family of Tumors: a Sub Group of Small Round Cell Tumors Defined by Specific Chimeric Transcripts. N Engl J 1994; 331:294.
4. Denny CT. Gene Rearrangements in Ewing's Sarcoma. Cancer invest. 1996; 14: 83.
5. Gururangan S, Marina NM, LVO X. et al. Treatment in Children with Peripheral Primitive Neuroectodermal Tumor or Extraosseous Ewing's Tumor with Directed Therapy. J Pediat Hematol Oncol. 1998; 20:55.
6. Lajos V, CSOKONAI, Balint L, Gabriella A, Budapest, Hungary. Ewing's Sarcoma in the Nasal Cavity. Otolaryngology Head and Neck Surgery. 2001; 125:665-7.
7. Howarth KL, Khodaei I, Karkanevatos A and Clarke RW. A Sinonasal Primary Ewing's Sarcoma. International Journal of Pediatric Otorhinolaryngology. 2004; 68:221-224.
8. de Alava E, Gerald WL. Molecular Biology of Ewing's Sarcoma/ Primitive Neuroectodermal Tumor Family. J Clin Oncol. 2000; 18:204.
9. Ambros I, Ambros P, Strehl et al. MIC2 is a Specific Marker for Ewing's Sarcoma and Primitive Neuroectodermal Tumors. Evidence of Common Histogenesis of Ewing's Sarcoma and PNET from MIC2 Expression and Specific Chromosome Aberration. Cancer. 1999; 67:1886.
10. Fillinger EJ, Garen-Chesa P, Triche TJ, et al. Immunohistochemical Analysis of Ewing's Sarcoma Cell Surface Antigen P30/32MIC2. Am J Path 1991; 139:317.
11. Nesbit ME Jr, Grehan EA, Burget EO Jr, et al. Multimodal Therapy for the Management of Primary Nonemetastatic Ewing's Sarcoma of Bone: A long Term Follow Up of the First Intergroup Study. J Ciin Oncoi 1990; 8:1664.
12. Jaffe N, Paed D, Traggis D et al. Improved Outlook for Ewing's Sarcoma with Combination Chemotherapy (Vincistine Actinomycin- D and Cyclophosphamide and Radiation Therapy. Cancer 1976; 38:1925.

13. Kinsella TJ, Miser JS, Walter B et al. Long Term Follow Up of Ewing's Sarcoma of Bone Treated with Combined Modality Therapy. Int J Radiat Oncol Biol Phys 1991; 21: 1501.
14. Ferrari S, Mercuri M, Rosito et al. Isofosfamide and Actinomycin-D, Added in the Induction Phase to Vincristine, Cyclophosphamide and Doxorubacin, Impove Histologic Response and Prognosis in Patients with None Metastatic Ewing's Sarcoma of the Extremity. J Chemotherapy 1998; 10: 484.