Primary Soft Tissue Orbital Ewing's Sarcoma Causing Unilateral Visual Loss: A Case Report

M. ASHRAF SHAHEEN, ABDUL HAMEED, MUHAMMAD IRFAN
Kiran Niraula, M. Tariq, Suresh Sapkota, Joshan Neupane, Diggaj Shrestha
Ram Chandra Shrestha, Shah Zaib Tasdique
Department of Neurosurgery, King Edward Medical University / Mayo Hospital, Lahore

ABSTRACT

A case of primary Ewing's sarcoma of right orbit causing unilateral visual loss in a 3 year old female child is reported. The child presented with 1 year history of painless unilateral proptosis and loss of vision. Loss of vision due to primary Ewing's sarcoma in orbit is extremely rare and it should be considered as one of the differential diagnoses while dealing the patient with these sorts of clinical presentation.

INTRODUCTION

Ewing's sarcoma, primarily a bony lesion[1] belonging to a group of malignant small cell neoplasm has various degrees of neuroectodermal differentiation.[2,3] It commonly arises from diaphysis of a long bone of pelvis, or vertebrae or the ribs.[3] In 80% of the cases the tumor typically occurs in the first and second decade of life.[2, 4] Extraosseous[5] and primary Ewing's of head and neck[6] regions are rare entities. Although described by Dr James Ewing's in 1921[7] the diagnosis is still debatable due to lack of consistency in description of a certain cell of origin.

Here, we describe a case involving right orbit and extending to right temporal lobe in a 3 year old female child with 1 year history of painless unilateral proptosis and loss of vision.

CASE REPORT

A 3 – year – old female child from Peshawar admitted to department of Neurosurgery, King Edward Medical University / Mayo Hospital, a referred patient from department of Ophthalmology of the same hospital. The child presented with features of redness of right eye ball which was painless and the proptosis was increasing for last 1 year causing disfiguring (Figure 1 a, b). There was associated loss of vision but there were no complaints of headache or vomiting. She was operated in Khyber Teaching Hospital, hospital to her locality where right lateral orbitotomy was done though lid crease incision under General Anesthesia. Grossly, lesion was soft jelly like in the retrobulbar region. Biopsy revealed spindle cell lesion suggestive of aggressive tumor. Then the teaching hospital referred the case to our center in Lahore.

The child was oriented in time, place and person and was vitally stable. Abdominal examination revealed no organomegaly; it was soft, non tender with audible bowel sounds. Her chest was bilaterally clear with normal vesicular breath sound with no added sounds. Cardiovascular examination was normal with audible 1st and 2nd heart sounds with no audible murmurs. Her local examination revealed a right sided proptosis with restriction of extraocular movements. Her visual acuity was 6/6 on left eye whereas on right eye there was no perception of light and loss of direct and consensual light reflex. Her right eye was full of pus with damaged cornea. She didn't have lymphadenopathy on examination of cervical, inguinal, axillary and epitrochlear lymph nodes. Her hematological analysis was within normal limits and chest x-ray, abdominal ultrasound were normal. Isotope Bone scan revealed no signs of metastatic disease.

Computed Tomography (CT) scan revealed a lesion involving both intra and extraconal territories of right orbit which was pushing the globe superio-

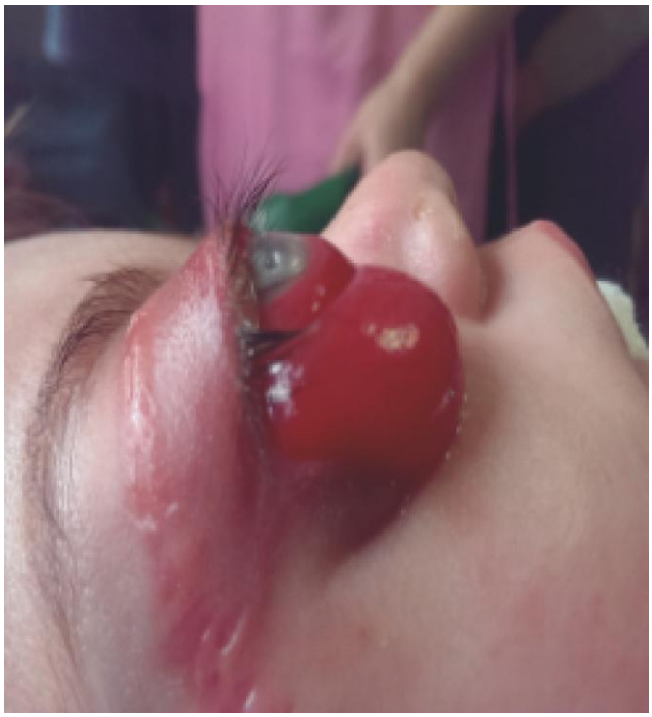


Figure 1a: *Lesion from Lateral Side.*

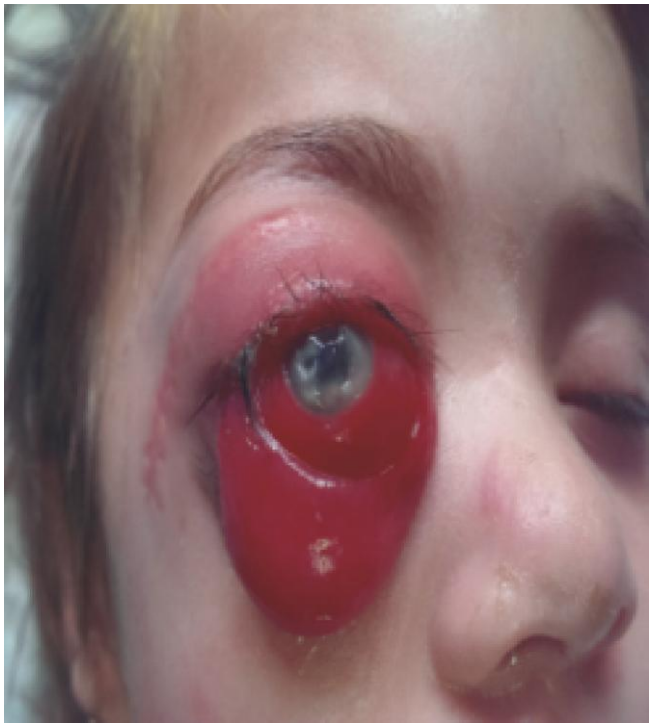


Figure 1b: *Lesion from Front.*

laterally. It also had infiltrated extradurally along the right middle cranial fossa (Figure 2). Her magnetic

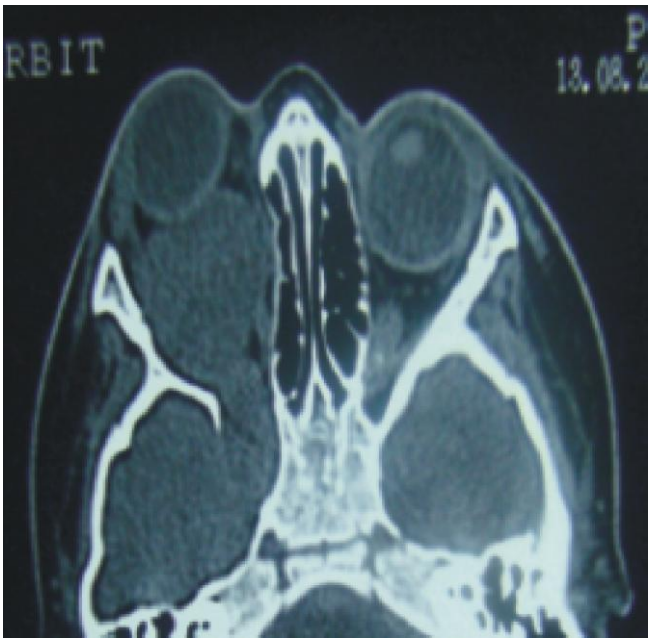


Figure 2: *Computed Tomography (CT) scan revealing the lesion involving both intra and extra-conal territories of right orbit pushing the globe superiolaterally and infiltrating extra-durally along the right middle cranial fossa.*

resonance imaging (MRI) revealed a T_1 – hypointensity with homogenous contrast enhancement in the right orbit with extradural extension to right middle cranial fossa pushing right temporal lobe laterally (Figure 3 a, b, c). The child underwent a Superolateral Orbitotomy with fronto temporal craniotomy and gross total excision of mass along with enucleation of the right eyeball. Eye ball was removed due to empyema of the globe and corneal damage. Soft, mild vascular, partly suckable, greyish, extra dural tumor occupying right retro-orbital region displacing the eye ball outwards was the operative finding. The tumor was not very vascular. It was pushing the right temporal lobe laterally. Gross total resection of tumor along with enucleation of visionless right eye ball was done. Partially involved right optic nerve was excised with the tumor and the stump was left to retract back (Figure 4 a, b, c). Patient recovered uneventfully from anesthesia. The histopathological (sent to two different pathological laboratories) examinations revealed a malignant neoplasm. Neoplastic cells were round blue cell tumor cell with hyperchromatin nuclei and scanty cytoplasm (Figure 5 a, b). On immunohistochemistry, specimen was positive for vimentin and CD_{99}. The lesion was negative for Desmin and Epithelial

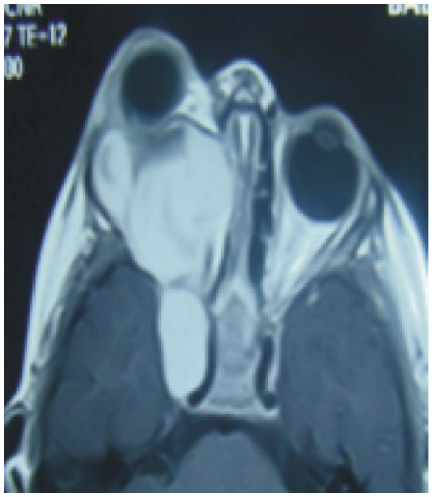


Figure 3a: *T_1 Axial MRI.*

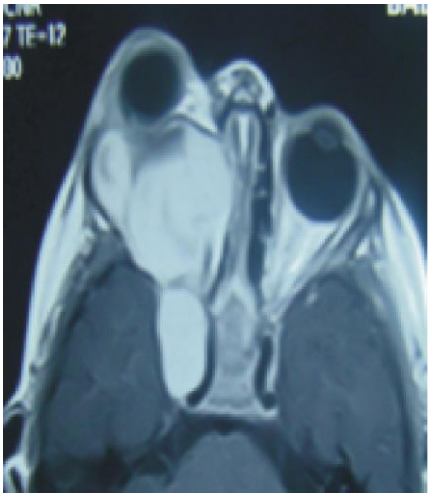


Figure 3b: *Contrast Axial MRI.*

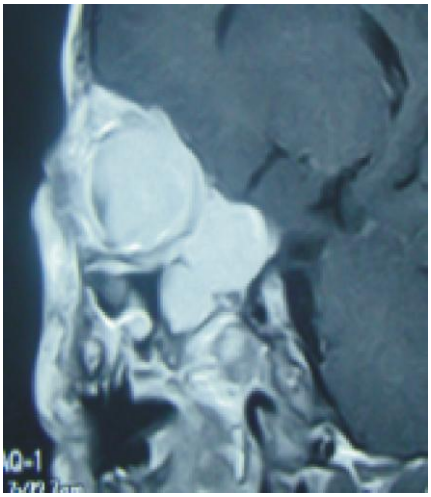


Figure 3c: *Contrast Sagittal MRI.*

Figure 3: *MRI revealing a T_1 – hypointensity with homogenous contrast enhancement in the right orbit with extradural extension to right middle cranial fossa pushing right temporal lobe laterally.*

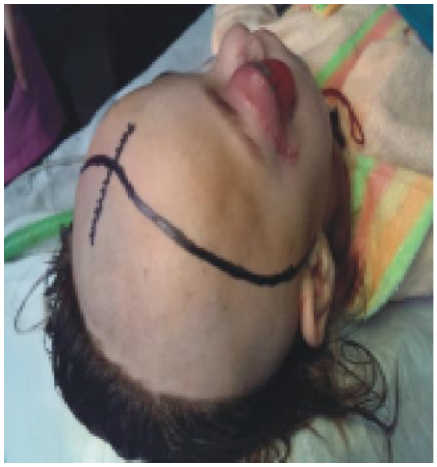


Figure 4a: *Incision Mark.*

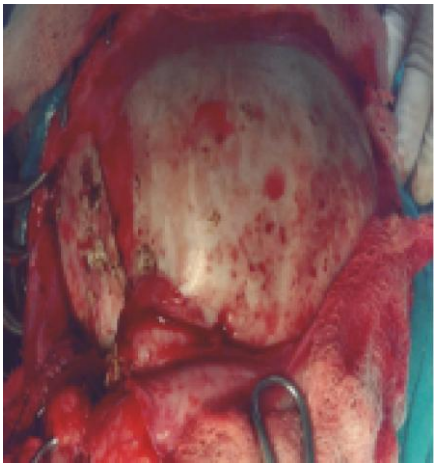


Figure 4b: *Superio-lateral Orbitotomy with Fronto Temporal Craniotomy.*

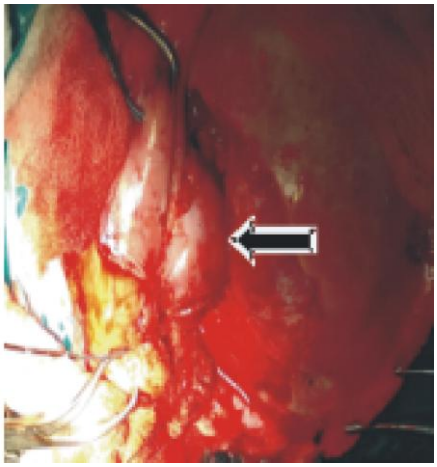


Figure 4.c: *Tumor Visualized.*

Membrane Antigen (EMA). Overall findings revealed Soft tissue Ewing's sarcoma of Orbit. Overall post operative period was uneventful. A week after the surgery the stitches were removed and scare mark over the scalp and over the right upper eye lid is visible. She was referred to Khyber Medical College at her home town for radiotherapy and chemotherapy on her parents request (Figure 6).

DISCUSSION

Primary orbital Ewing's sarcoma is extremely rare for proptosis and visual loss[8]; rather in this age group orbital tumors like rhabdomyosarcoma[9] could be suspected. Ewing's sarcoma in pediatric age group involving orbit with intracranial extension together, is rarely reported. Separate cases of intracranial involvement and orbit have been reported.[10-14] Intracranial Ewing's of the calvarium is reported causing mass effect.[15] Bhatoe had discovered primary orbital Ewing's sarcoma, could extend the cranium as well.[16]

Normally distance between lateral orbital rim to the corneal apex is 16 – 21 mm and proptosis is defined if it exceeds 22 mm.[11] Also asymmetry of more than 2 mm between two eyes is also regarded as proptosis. In our patient, the distance between lateral orbital rim to the corneal apex was approximately 25 mm in the right eye. Asymmetry of more than 2 mm bet-

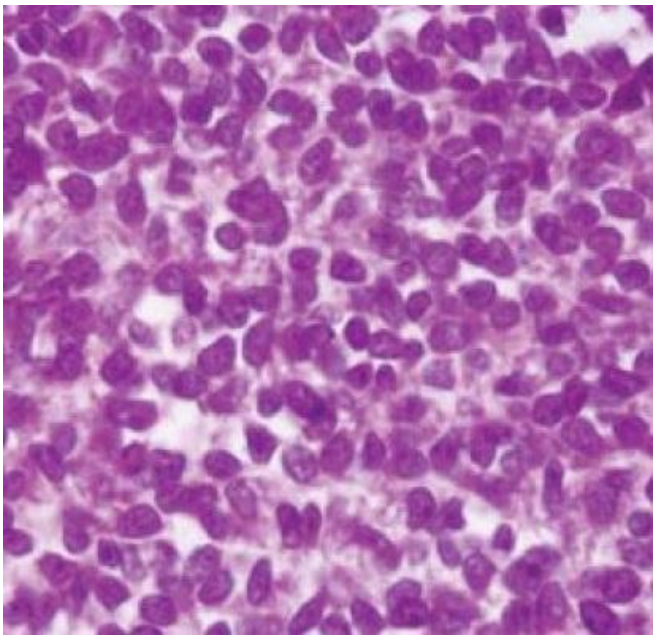
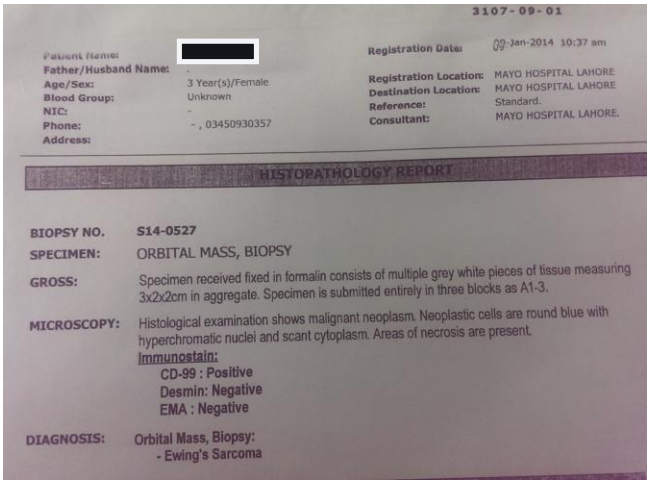


Figure 5a: *Neoplastic cells were round blue cell tumor cell with hyperchromatin nuclei and scanty cytoplasm.*



3107-09-01

Patient Name: [redacted]
Father/Husband Name:
Age/Sex: 3 Year(s)/Female
Blood Group: Unknown
NIC: -
Phone: - , 03450930357
Address:

Registration Date: 09-Jan-2014 10:37 am
Registration Location: MAYO HOSPITAL LAHORE
Destination Location: MAYO HOSPITAL LAHORE
Reference: Standard.
Consultant: MAYO HOSPITAL LAHORE.

HISTOPATHOLOGY REPORT

BIOPSY NO. S14-0527
SPECIMEN: ORBITAL MASS, BIOPSY
GROSS: Specimen received fixed in formalin consists of multiple grey white pieces of tissue measuring 3x2x2cm in aggregate. Specimen is submitted entirely in three blocks as A1-3.
MICROSCOPY: Histological examination shows malignant neoplasm. Neoplastic cells are round blue with hyperchromatic nuclei and scant cytoplasm. Areas of necrosis are present.
Immunostain:
CD-99 : Positive
Desmin: Negative
EMA : Negative
DIAGNOSIS: Orbital Mass, Biopsy:
- Ewing's Sarcoma

Figure 5b: *Histopathology Report.*

ween two eyes was also present in our patient. Both the criterias favors proptosis. The eye ball was pushed superio-laterally and had restriction to eye movement; hence intraconal pathology or intraconal along with extraconal pathology was to be sought. Considering relatively short history, lack of fever, rapid progression, non-pulsatile nature a provisional diagnosis of orbital malignancy was made. Incisional Biopsy revealed spindle cell lesion suggestive of aggressive tumor. The rapidity of growth, opacity of cornea, lack of vision, gave us mandated to perform surgical decompresssion in this patient.

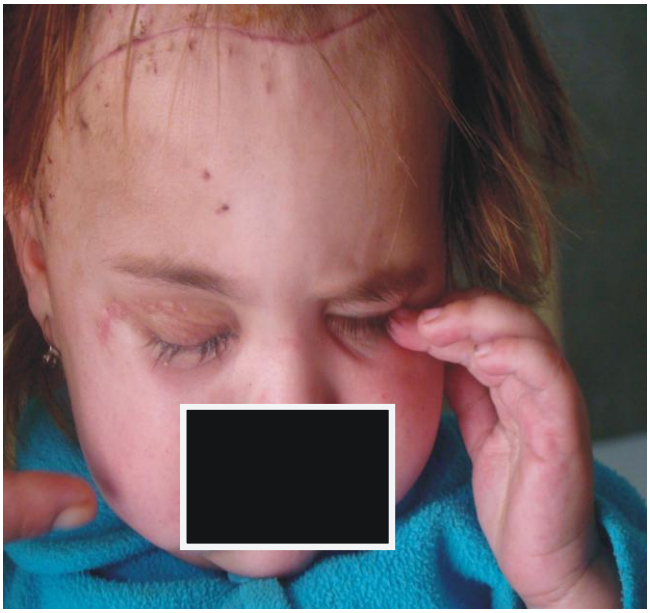


Figure 6: *Scar mark Over the Scalp and Right Upper Eye Lid 1 Week after the Surgery.*

In our patient, histological findings were Neoplastic round blue cell tumor cell with hyperchromatin nuclei and scanty cytoplasm. This type of features is also present in other small, round cell tumors of neuroectodermal origins, such as rhabdomyosarcoma, neuroblastoma and lymphoma.[17] For accurate diagnosis immunohistochemistry studies were important in this case.[18] Our patient's specimen was positive for vimentin and CD_{99} and negative for Desmin and Epithelial Membrane Antigen (EMA). In Ewing's sarcoma, the most consistent finding in immunohistochemistry is positive vimentin. The tumor cells usually contain large amount of cytoplasmic glycogen which is demonstrated by periodic acid – Schiff stain with diastase control. Tests like LCA, chromogranins A and B and osteonectin are negative for Ewing's sarcoma.[19,20] The detection of monoclonal antibodies to the fusion gene product, termed as CD_{99} (MIC_2), is also contributory to the diagnosis of Ewing's sarcoma. Translocation of t (11; 22) chromosome and the resulting EWS – FL_{11} gene fusion also contributes in diagnosis.[21]

Our patient has been referred to Khyber Medical college hospital for chemotherapy and radiotherapy on her parent's request. Chemotherapeutic agents like ifosfamide, etoposide, doxorubicin show promising

results in this tumor. Treatment of Ewing's sarcoma with a combination of surgery, radiotherapy and chemotherapy results in 5 year survival rate of approximately 65%.[17,18] After 1993, the 5 year survival rate increased from 44 to 68%, whereas for metastatic disease it increased from 16 to 32%.[1]

With timely and proper treatment quality and expectancy of life can be increased in this rare pediatric malignancy.

CONCLUSION

Ewing's sarcoma in the pediatric age group presenting as proptosis, vision loss with intracranial extension is an extremely rare manifestation. With appropriate management both quality and expectancy of life can be increased.

RECOMMENDATION

This rare diagnosis need to be taken into consideration as a differential diagnosis in patient with unilateral painless proptotic vision loss in pediatric age group.

Conflict of Interest

Authors declares no competing financial or non financial conflict of interest.

The study is not funded by any pharmaceutical company directly or indirectly involved in production of medication, instruments used for the management of this tumor.

Corresponding Author
Dr. Muhammad Irfan
MS – Neurosurgery
Department of Neurosurgery
King Edward Medical University/
Mayo Hospital, Lahore
E-mail: nsirfansheikh@gmail.com

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