

Case Report-Rhabdomyosarcoma Masquerading Lymphoma

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1. Introduction

Rhabdomyosarcoma

- Rhabdomyosarcoma (RMS), accounting for 4.5% of all childhood cancers, is the most common soft tissue sarcoma in children. They are of 4 major types-embryonal, botryoid, alveolar and pleomorphic.
- It is a malignant tumor of mesenchymal origin which is included in the group of small round blue cell tumors along with neuroblastoma, lymphoma and primitive neuroectodermaltumors.
- It usually manifests as a slow growing expanding mass.

Ocular Lymphoma

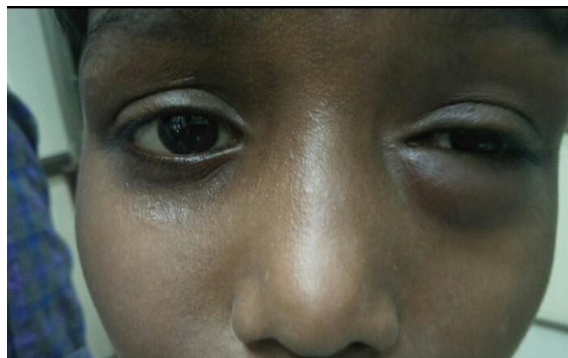
- B cell Non-Hodgkin lymphoma, being the most common variant of ocular lymphoma, constitutes 50% of all primary orbital malignancies in adults between 50 and 70 years of age.
- It presents as a palpable mass with a typical 'salmon-patch' appearance of the swollen conjunctiva.

2. Case History

- An otherwise healthy 12 year old Hindu male, presented to our tertiary healthcare center with an abrupt onset, *painless, gradually progressing swelling* of left eye lower lid since past 1 month.
- There was no history of diminution of vision, trauma, fever, weight loss or anorexia. There was no past history of similar complains and no significant family history.
- General examination was unremarkable. On ocular examination, a well defined hard mass was palpated

underlying lower lid. There was mild restriction of movements in superior & inferior direction.

- Visual acuity was 6/6 in both eyes
- Slit lamp examination showed a normal anterior segment with both eye pupils normally reacting to light and a normal fundus.



3. Materials and Methods

- The patient was then subjected to routine blood investigations, which revealed a high WBC count showing 67.9 % of polymorph on DLC and an absolute neutrophil count of 9,500/uL, *with normal lymphocyte count.*
- CT Scan of Orbit showed a homogeneous dense soft tissue swelling (HU:45-50) in the left infraorbital region with intraorbital extension, with a maximum thickness of 10mm. The lesion abuts the anterior most part of left lower rectus muscle & *was not encircling the globe.*

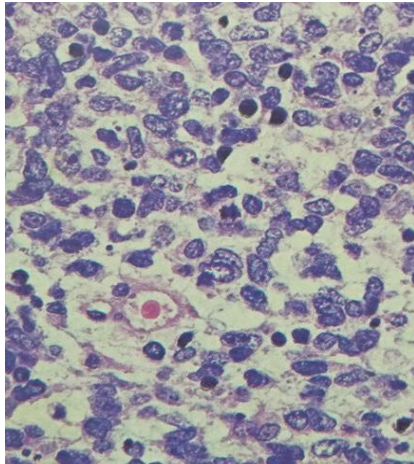


- An incisional biopsy was done via inferior conjunctival approach.
- Histopathological examination disclosed an undifferentiated *malignant round cell tumor* having small

round to ovoid tumor cells having hyperchromatic round nuclei, scanty cytoplasm and occasional mitoses.

- For confirmation of diagnosis and further typing of tumor, Immunohistochemistry (IHC) was done which showed positive – TDT, PAX-5, CD79a, CD20, vimentin weak positive – CD99
Ki67 – 80-85%
negative – myogenin, synaptophysin, desmin

Hereby stamping a diagnosis of *high grade Precursor B Lymphoblastic Lymphoma*.



- Further, for ruling out metastasis, a panel of investigations was done showing a normal chest X Ray (PA view), few nodes in neck and axilla with preserved fatty hilum, in USG (all region), normal 2D ECHO and Doppler study, normocellular marrow uninvolved by any malignancy on Bone Marrow biopsy, CSF cytology was negative for any malignant cells.

4. Discussion

- Our case presented a diagnostic dilemma.
- Given the commonality, the age of the patient and slow progression of the tumor, the diagnosis of Embryonal Rhabdomyosarcoma was favoured initially in the light of its clinical presentation and histopathological examination, but it eventually turned out to be Lymphoma despite the absence of any redness, salmon patch appearance, significant movement restriction, visual symptoms or a typical CT scan finding of the lesion encircling the globe. The age group was also highly unfavourable for it to have raised a clinical suspicion of lymphoma.
- Patient underwent chemotherapy with 1 cycle of cytoreductive CVP (cyclophosphamide+ vincristine+ prednisolone) followed by MCP 841 protocol which includes VCR and prednisolone in 2 phases.

5. Conclusion



After 2 months of chemotherapy

- An RMS-mimicking tumor was finally diagnosed and treated as lymphoma. So it lays stress on the importance of typing and immunological markers for accurate diagnosis.
- Also it is evident that by not excising the tumor in its entirety, the underlying rectus muscle could be saved thereby not interfering with the ocular motility. Subsequent chemotherapy helped in destroying residual tumor.