when the lens vesicle is single and it is the laxity and abnormal stretching which causes the deep furrow and possibly duplication. Duplication may be the extreme form of such a bilobed, furrowed lens. It is interesting to note that both the cases previously reported had lens place in a straight axis at opposite ends. This gives further credence to our theory of the fissure dividing the lens into two and the remaining zonules pulling them away from each other.

Since lens coloboma occurs frequently such deep furrows (bilobed lens) are uncommon. We believe that it represents the missing link between the colobomatous lens and duplicated lens. We understand that a single case report may not be suggestive of any mechanism but nevertheless it does indicate a possible mechanism of lens duplication.

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Atypical presentation and diagnostic pitfalls: A case of rapidly progressive bilateral proptosis in a child aged 18 months

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We report an atypical presentation of non-Hodgkin lymphoma (NHL) in a child aged 18 months who presented with rapidly progressive bilateral proptosis. Computerized search using Medline did not reveal a similar presentation of NHL in such a young child. It stresses the need for an early histopathological study including immunohistochemistry and demonstrates the dramatic local response to combined radiotherapy and chemotherapy even in advanced stages without any ocular side-effects.

Key words: Bilateral, non-Hodgkin lymphoma, pediatric, proptosis

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There has been a progressive increase in the incidence of lymphoma over the years with frequent reports of varying and atypical presentations and a bewildering number of confusing classifications which still do not fit all the cases. This is especially true with respect to non-Hodgkin lymphoma (NHL). This case report discusses the difficulties encountered in diagnosis, the need for an early incisional biopsy and a complete immunohistopathological study with all the markers. It also highlights the fact that lymphomas respond dramatically to combined radiotherapy and chemotherapy.

Case History

An 18-month-old female child presented with a history of rapidly increasing prominence of both eyes of two months duration. It was first noticed in the left eye, the right eye was involved two to three days later. The child had occasional fever and did not have any major eye or systemic disease before the onset of prominence of the eyes. Her brother, five years old, was healthy.

The child weighed 9 kg and was afebrile. Examination of the eyes revealed an axial proptosis which measured 22 mm right and 23 mm left eye at an outer intercanthal distance of 82 mm. Lid edema, prominent vessels over the lids and lagophthalmos were more pronounced in the left than the right eye [Fig. 1]. There was gross congestion and chemosis of the conjunctiva and exposure keratitis in the left eye. The conjunctiva was mildly congested in right eye and the cornea was clear. Ocular motility was restricted in all directions of gaze in both eyes. Pupils were normal and reactive in both the eyes and vision appeared normal. Fundus examination of right eye revealed a normal optic disc but the left eye showed an edematous disc. Based on clinical findings, neuroblastoma, granulocytic sarcoma, multifocal eosinophilic granuloma and inflammatory pseudotumor were considered in the differential diagnosis.

Blood examination revealed hemoglobin of 10.9 gm%, erythrocyte sedimentation rate (ESR) of 35 mm, bleeding time of 1 min 25 sec and clotting time of 7 min 02 sec and blood counts were normal. Peripheral blood smear showed microcytic hypochromic picture without any abnormal or atypical cells and adequate platelets. Routine ENT and pediatric referral did not reveal any abnormality. Chest X-ray was normal. Computerized tomography [Fig. 2] showed diffuse infiltration of the orbit and erosion of medial wall of the orbit bordering the nose and lateral wall of the nose bordering the infra-temporal fossa on both sides. Adjacent paranasal sinuses were normal. A malignant lesion was suspected and orbital biopsy was advised.

An incisional biopsy was done from both orbits to confirm a single bilateral etiology. Histopathological sections [Fig. 3] study showed solid sheets of predominantly undifferentiated
cells along with a few interspersed elongated and fusiform cells with central nucleus and eosinophilic cytoplasm. The undifferentiated cells were round to elongated with large pleomorphic nucleus and prominent single to multiple nucleoli with numerous atypical mitosis. Amidst these there were also a few vacuolated cells. These features favored the diagnosis of undifferentiated rhabdomyosarcoma. Immunohistochemistry was advised to confirm the diagnosis. Bone marrow was positive for malignant cells and cerebrospinal fluid showed metastasis to the brain. Ultrasound scan of the abdomen and pelvis showed hepatomegaly with multiple small hypoechoic lesions. Renal function tests were normal. Serum calcium (12 mg/dl) and phosphorus (7 mg/dl) were slightly higher than normal. Liver function tests were normal except serum alanine transaminase (50 U/L).

In consultation with an oncologist a multimodal treatment was planned with radiotherapy and chemotherapy. As the patient’s left eye was at risk due to exposure keratitis and papilledema, it was decided to start local external beam irradiation followed by chemotherapy. A total of 46 cGy with 180 cGy per fraction were given over a period of five weeks. Lens was shielded from exposure to radiation. Chemotherapy was planned as per IRS-IV protocol with injection vincristine 1.5 mg/m² IV weekly for 12 weeks and on Week 16, injection dactinomycin 0.015 mg/Kg/day from Day 1 to Day 5, every three weeks up to 16 weeks, injection cyclophosphamide 2.2 g/m² with mesna 440mg/m² for one day per cycle every three weeks up to 16 weeks. Chemotherapy was interrupted due to anemia and low neutrophil counts requiring blood transfusion. Proptosis completely disappeared, the exposure keratitis of the left eye healed and disc edema totally subsided 2.5 months after starting therapy [Fig. 1]. Patient did not have any visual deficit in either eye. Unfortunately, patient expired 6.5 months after she was first seen in our hospital due to central nervous system infiltration. She did not develop any complications due to radiation during her follow-up.

As this was an unusual presentation, tissue block was sent for immunohistochemistry with an academic interest. The markers used were Cytokeratin (epithelial tumors), Vimentin (mesenchyme-derived neoplasms including lymphomas, melanomas, some sarcomas and certain neural tumors), Leukocyte Common Antigen (lymphoma), CD20 and CD79A (B-cell neoplasms), CD3, CD43 and CD99 (T-cell neoplasms), CD30 (anaplastic large cell lymphoma and Hodgkin disease), Desmin (rhabdomyosarcoma), Synaptophysin (neuroblastoma), Myeloperoxidase (granulocytic sarcoma), HMB45 (melanoma), TDT (lymphoblastic lymphomas and leukemia) and Ki-67 (proliferation-related marker). It was negative for all markers except Vimentin and Leukocyte Common Antigen [Fig. 3]. KI 67 was 60 to 70% positive. The immunohistochemistry report was unclassifiable high-grade NHL.

**Discussion**

Lymphomas are solid malignant neoplasms that originate from leukocytes, mostly lymphocytes, dendritic histiocytes and plasma cells. Lymphoma occurs most commonly in lymph nodes and primary lymphoid tissue but also in extranodal sites, including the orbit, skin oropharynx, gastrointestinal tract and bone marrow. Systemic NHL in adults is usually seen in the fifth or sixth decade and presentation in childhood is extremely rare.
Most orbital lymphomas are NHL and tend to present in the sixth and seventh decade and present anteriorly in the orbit.\textsuperscript{4} Presentation before the age of 20 is rare.\textsuperscript{4} In one reported case series,\textsuperscript{1} although NHL was the most frequent malignant tumor of the eye and ocular adnexae (55%), the youngest patient was 15-years-old (15 to 96) and the median age was 71 years.

In another review\textsuperscript{5} of 145 patients of lymphoma orbit and/ or adnexa and/or ocular, the youngest patient was three-years-old (three to 96 years) and the median age was 66 years and 10% were bilateral. Exophthalmia was seen in 27%. Kodsi \textit{et al.} in their review of 340 orbital tumors in children found only 18.2% of them to be malignant\textsuperscript{6} (11.5% primary and 6.8% metastatic). Among 62 malignant tumors rhabdomyosarcoma (24) was the most common malignant neoplasm, with lymphoma (five) much less frequent.

Most of the lymphomas of the eye and its adnexa are NHL of B-cell type.\textsuperscript{7} Computed tomography demonstrates the molding of the mass to orbital structures, such as the globe and orbital bones, without bony erosion except in large cell lymphoma.\textsuperscript{4}

Radiotherapy is effective in indolent forms and chemotherapy is applied in aggressive lymphomas and a combination of radio- and chemotherapy may even be more effective.\textsuperscript{8} The presence of bilateral involvement, extension to the subcutaneous tissue or temporalis fossa is associated with increased risk of dissemination.\textsuperscript{9} Higher grade and destruction of orbital wall suggest a poor prognosis.\textsuperscript{10}

In conclusion, it was a rare presentation of a primary orbital NHL in a child aged 18 months presenting as rapidly progressive bilateral axial proptosis with an early metastasis. In this case the tumor was so poorly differentiated that it couldn’t be classified.\textsuperscript{3} Bony erosion of orbital and nasal wall instead of molding to the adjoining structures was seen. There was no radiation-induced complication in the eye with the dosage given, over a follow-up period of 6.5 months.

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\textbf{Bilateral conjunctival retention cysts in the aftermath of Stevens-Johnson syndrome}

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In this case report, we describe the rare occurrence of bilateral conjunctival retention cysts in a child with Stevens-Johnson syndrome. The case was managed conservatively as there were no functional or cosmetic problems.

\textbf{Key words:} Bilateral, conjunctival retention cyst, Stevens-Johnson syndrome

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Extensive ocular surface scarring is the common denominator in Stevens-Johnson syndrome (SJS) causing complications such as symblepharon, dry eye, ocular surface keratinization, lacrimal duct obstruction and lid deformities.\textsuperscript{1} Conjunctival and orbital cysts were reported occasionally.\textsuperscript{7} We describe the occurrence of bilateral conjunctival retention cysts in a child with SJS.

\textbf{Case History}

An 11-year-old male child was referred for epiphora and bilateral conjunctival swelling. He was admitted in a hospital and treated for Stevens-Johnson syndrome. The case was managed conservatively as there were no functional or cosmetic problems.

\textbf{Key words:} Bilateral, conjunctival retention cyst, Stevens-Johnson syndrome

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Extensive ocular surface scarring is the common denominator in Stevens-Johnson syndrome (SJS) causing complications such as symblepharon, dry eye, ocular surface keratinization, lacrimal duct obstruction and lid deformities.\textsuperscript{1} Conjunctival and orbital cysts were reported occasionally.\textsuperscript{7} We describe the occurrence of bilateral conjunctival retention cysts in a child with SJS.

\textbf{Case History}

An 11-year-old male child was referred for epiphora and bilateral conjunctival swelling. He was admitted in a hospital and treated for Stevens-Johnson syndrome after an attack of brief fever, three years