

CASE REPORT

Burkitt's Lymphoma of Anterior Skull Base associated with Acute Loss of Vision: A Rare Case Report

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ABSTRACT

Burkitt's lymphoma is a malignant neoplasm rarely found in paranasal sinuses and skull base. We report a rare case of Burkitt's lymphoma of anterior skull base associated with acute loss of vision. A 10 years old boy presented with history of headache, diminution of vision in right eye since 15 days. Biopsy showed diffuse NHL of Burkitt's type. After treatment with chemotherapy there was complete recovery in vision.

Keywords: Burkitt's lymphoma, Skull base, Visual loss.

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CASE REPORT

A 10-year-old boy presented to the Department of Otorhinolaryngology, Sardar Patel Medical College, Bikaner, with history of headache, diminished vision in right eye since 15 days. On examination, patient had no perception of light in right eye and was associated with afferent pupillary defect. There was slight proptosis of right eye with normal extraocular movements. No other abnormality was found on diffuse light and slit lamp examination. Fundus examination showed a pale disk with attenuated vessels. Left eye examination was essentially normal.

On nasal endoscopy, bilateral concha bullosa was seen, mild DNS toward the left side. Adenoids were enlarged. CT PNS showed mass lesion in bilateral posterior ethmoid and sphenoid sinuses. T2 MRI showed hypointensities in

sphenoid sinus and posterior ethmoid cells. Involvement of body, lesser and greater wings of sphenoid was seen, showing marrow infiltration, cortical erosion and extraosseous soft tissue components. Lesion also involved the bony wall of right optic canal with compression of right optic nerve (Figs 1 and 2). Endoscopic biopsy was taken from sphenoidal and posterior ethmoidal mass and sent for histopathological examination. Histopathology revealed diffuse non-Hodgkins lymphoma of Burkitt's type (Burkitt's Lymphoma). Immunohistochemistry was positive for CD 20, CD 10 and KI 67, negative for CD 3 and BCL 2, suggestive of Burkitt's lymphoma. PBF report showed microcytic hypochromic anemia. No immature lymphoid or myeloid cells seen. CSF cytological examination showed few lymphocytes and no malignant cells.

Treatment was started with intensive combination chemotherapy. Regimens involving methotrexate, cytarabine, cyclophosphamide, vincristine, Adriamycin were given cyclically. After that radiotherapy was given in 10 fractions of 200 cGy. He responded dramatically to treatment with improvement of vision. CT scan repeated after full course of chemotherapy and RT and it showed disappearance of skull-base lesion.

DISCUSSION

Burkitt's lymphoma (BL) is a malignant endemic neoplasia with a mandibular localization, described for the first time in 1958, in African children. BL represents one of the highly aggressive subtypes of NHL. BL occurs as endemic (African), sporadic and immunodeficiency-associated forms and atypical form in WHO scheme.

Endemic BL: It presents in childhood/adolescence in Africa with large extranodal tumors in jaw or abdominal viscera; 90% associated with EBV infection; aggressive but curable disease.

Sporadic BL: Often children, rare in adults (median age 31); presents with rapidly growing lymphadenopathy, often intra-abdominal mass arising from a Peyer's patch or mesenteric node; BM, CNS and blood involvement frequent; 30% associated with EBV infection; also associated with HIV infection; aggressive but curable disease in non-HIV associated cases.

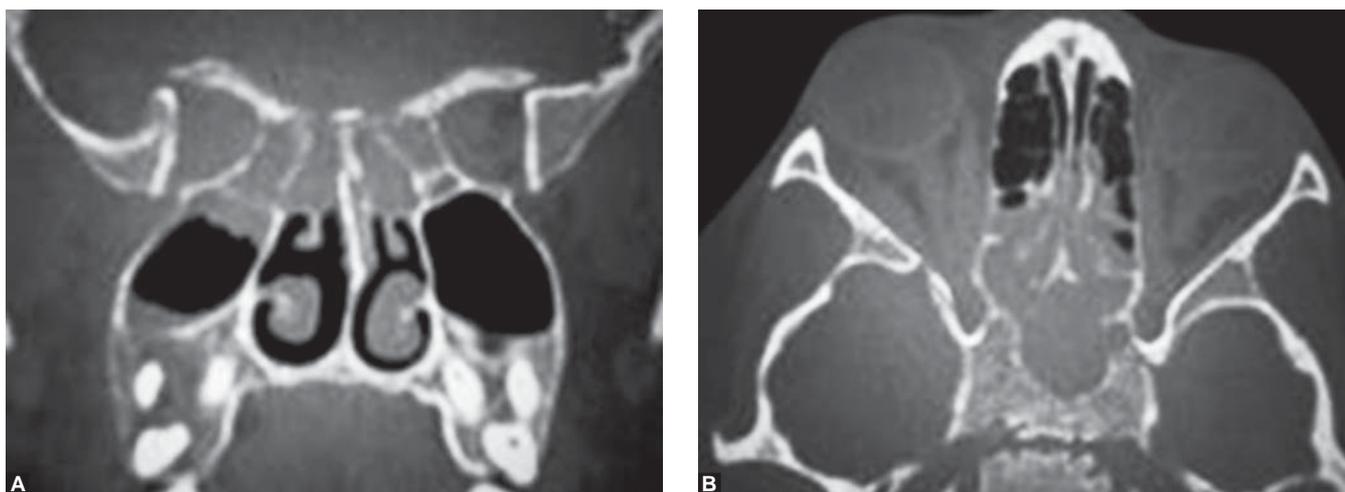
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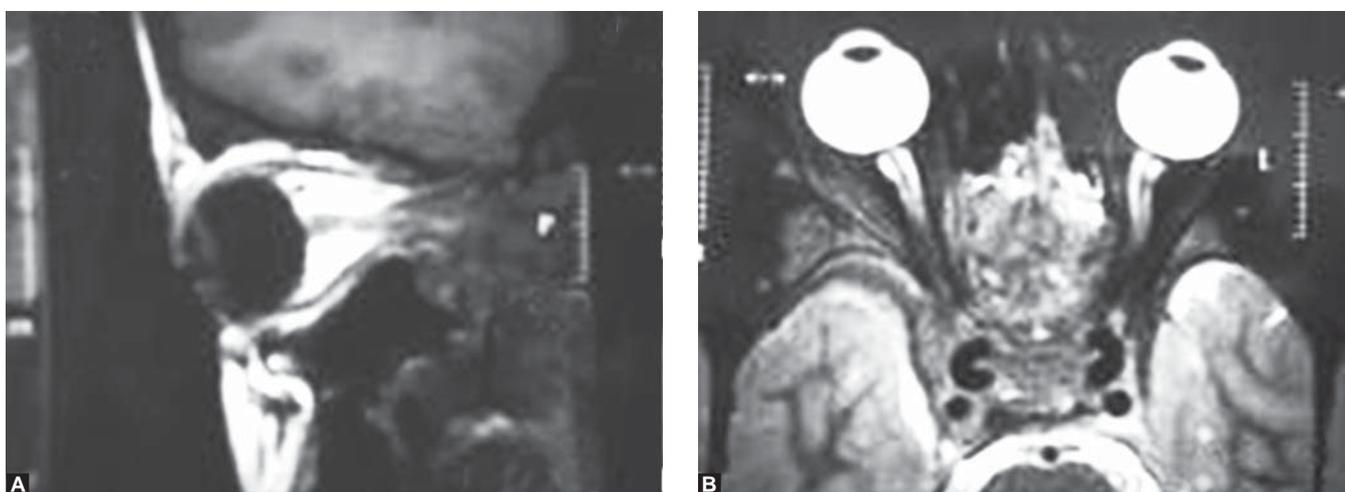
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Figs 1A and B: CT scan (coronal and axial cuts) showing mass lesion in bilateral posterior ethmoid and sphenoid sinuses along with involvement of bony wall of right optic canal



Figs 2A and B: MRI scan (sagittal and axial cuts) showing mass in bilateral posterior ethmoid and sphenoid sinuses along with compression of right optic nerve

NHL is rare in skull base. The most common symptom of sinonasal lymphomas is nasal obstruction; others are headache, rhinorrhea and facial pain, mostly consistent with chronic rhino sinusitis.¹ Endoscopically, sinonasal lymphoma can present as a polypoid mass-like lesion. Thus, it can be easily confused with paranasal sinusitis, polypoid disease or other tumorous conditions. Therefore, in the early stages, Burkitt's lymphomas in this region may be misdiagnosed as other diseases. Our patient was misdiagnosed with fungal rhinosinusitis and underwent medical treatment for 15 days. The initiation of chemotherapy was delayed due to misdiagnosis.

Early diagnosis and prompt chemotherapy is important in Burkitt's lymphoma because it is one of the fastest growing tumors, with a cell doubling time of only 24 to 48 hours.^{2,3} If treatment is delayed, Burkitt's lymphoma can spread to other parts of the body, leading to poor prognosis. If Burkitt's lymphoma involves the paranasal sinuses, it can cause facial deformity. Involvement of the facial bones and orbit

is less common in the North American cases, but invasion of the orbit from the sinuses may occur.^{4,5} Generally, Ann Arbor staging system has been used in both Hodgkin's and non-Hodgkin lymphoma, and Burkitt's lymphoma could be classified by same staging system.⁶ In children, survival rates of 80 to 90% are being achieved with intensive, short duration chemotherapeutic protocols.⁷

CONCLUSION

Burkitt's lymphoma is rare in skull base and paranasal sinuses. Lesion adjacent to orbit can lead to sudden and permanent loss of vision. So early diagnosis and prompt chemotherapy is important to spare vision and other complications in skull base and sphenoidal lymphomas.

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