CASE REPORT

Stage IV Burkitt Lymphoma Presenting with Eyelid Swelling

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ABSTRACT

Burkitt lymphoma is a rare and aggressive tumour which predominantly affects children. We report a case of Stage IV Burkitt lymphoma (non-endemic type) in a 5-year-old immunocompetent Malay child who initially presented with left upper eyelid swelling. Clinical examination revealed a soft solid swelling over the left upper lid, extending temporally up to the lacrimal gland. Computed tomography (CT) scan showed a homogenous mass of soft tissue density with intracranial and left orbital involvement. Incisional biopsy of the mass revealed histological changes suggestive of B lymphoblastic lymphoma or leukemia. She was treated with 6 cycles of chemotherapy. Post chemotherapy, the left temporal and upper eye lid swelling subsided. Clinicians need to be on alert when encountering children with eyelid swelling, as this could be an initial presenting feature of Burkitt lymphoma without other systemic symptoms. A delay in diagnosis can lead to aggressive orbital involvement followed by permanent visual loss.

KEYWORDS: Stage IV Burkitt Lymphoma, Eye lid swelling

INTRODUCTION

Burkitt lymphoma is a rapidly growing B-Cell lymphoma which predominantly affects children. It accounts for two-thirds of B-cell non-Hodgkin lymphoma cases. It can be divided into endemic, sporadic (non-endemic type) and immunodeficiency-associated types. The most common presenting features in Burkitt lymphoma are facial bone involvement (in the endemic type) or abdominal mass and ileo-coecal region involvement (in the sporadic type). In immunodeficiency-associated Burkitt lymphoma, patients usually present with organ and nodal involvement[1,2] although there have been some rare case reports of occurrence in gastric[3] and pancreas.[4] To date, only one report of Burkitt lymphoma occurring in conjunctiva[5] has been found.

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We discuss a case of stage IV Burkitt lymphoma presenting with an upper eyelid swelling in an immunocompetent Malay child.

Case Report

A 5-year-old Malay girl presented with one month history of swelling on the temporal side of her left eye, which progressed to involve the upper lid. There was mild pain but no eye discharge, eye redness, blurring of vision or pain upon moving the eye. There were no fever, loss of appetite, loss of weight or other constitutional symptoms. She was initially suspected to have a left temporal abscess and treated by the paediatric team with a course of antibiotics. However, there was no improvement. She was then referred to the ophthalmology department for further investigation.

Clinical examination revealed a soft swelling over the left upper lid, extending temporally to include the lacrimal gland. There was mild tenderness. Visual acuity was 6/9 bilaterally and extra-ocular eye movements were full. The anterior segment and fundi were normal in both eyes. There was no proptosis. An urgent computed tomography (CT)

scan showed a mass with intracranial and left orbital involvement with the given provisional diagnosis of rhabdomyosarcoma.

Incisional biopsy of the left upper lid and temporal swelling then revealed histological changes suggestive of B lymphoblastic lymphoma or leukemia (Figure 3). Magnetic resonance imaging (MRI) further revealed a diffuse, ill-defined mass at the left temporal region, involving mainly the temporal muscle (Figure 1). The mass was directly infiltrating the left lateral wall of orbit and lateral rectus muscle, with the involvement of the left lacrimal gland. There was also intracranial extension into the left temporal lobe. CT scan of the thorax, abdomen and pelvis found multiple hypodense lesions in both kidneys, with an enlarged left medial supraclavicular lymph node. Bone marrow aspiration and all other blood investigations were normal. Test for antibody towards Epstein Barr virus in this patient was not done due to unavailability of the test in our centre.

She was treated as stage IV Burkitt lymphoma with orbital metastasis and started on chemotherapy. After she completed 6 cycles of chemo protocol 902 (COPADM3) which are cyclophosphamide, vincristine, hydrocortisone, doxurubian and methotrexate, the left temporal and upper eye lid swelling subsided (Figure 1). The latest CT scan six months post treatment showed a reduction in the size of the temporal mass (Figure 2) and resolved renal masses bilaterally.

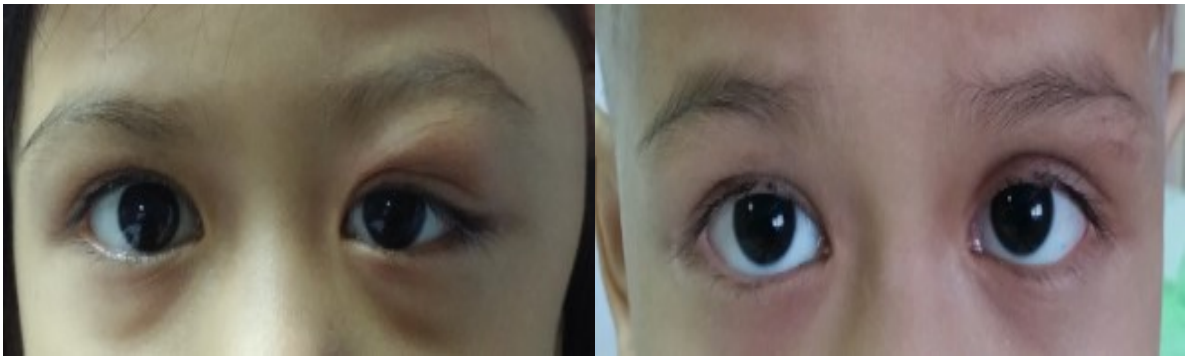

Figure 1: Left sided photo showed swelling over the left upper lid and right sided photo showed resolving left upper lid swelling after chemotherapy

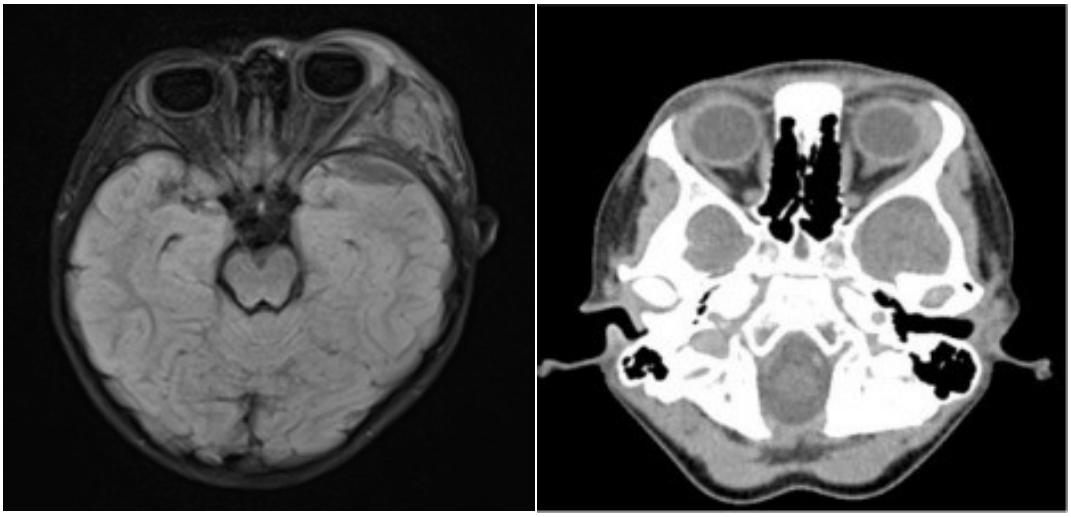

Figure 2: MRI over left sided photo shows a diffuse, ill-defined mass was seen at the left temporal region, involving mainly the temporal muscle and infiltrating the left lateral wall of orbit, lateral rectus muscle, and left lacrimal gland. There was intracranial extension into the left temporal lobe. After 6 cycles of chemotherapy, the CT scan over right sided photo shows a reduction in the size of the temporal mass.

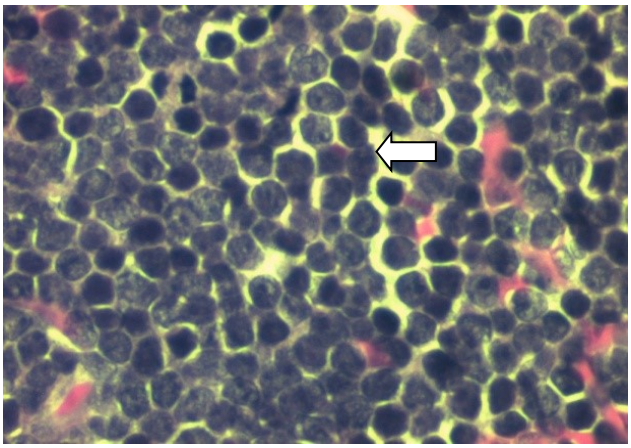

Figure 3: Histopathology examination with immunohistochemistry staining showed atypical lymphoid cells with increased mitotic figures.

DISCUSSION

Burkitt lymphoma, also known as B-cell non-Hodgkin lymphoma, is a rare and extremely aggressive tumour first described by Denis Parsons Burkitt in 1958 in African children.[1] It is divided into endemic, sporadic and immunodeficiency-associated type.[1,2] Our patient's clinical history and blood / bone marrow investigation did not reveal any evidence of immune-deficiency and hence is classified to have sporadic disease.

The adult cases have been identified in the sporadic form of Burkitt lymphoma as compared to the endemic form of Burkitt lymphoma, which mainly affects children. In sporadic form, patients usually present with symptoms of abdominal mass or intestinal obstruction due to the involvement of lymphoid tissues of the mesenteric and bowel lymph nodes. Besides this, the bone marrow is also frequently involved. The central nervous system and the mediastinal area can be affected later in endemic and sporadic form. However, the orbital involvement in sporadic form is rarely encountered.[5]

There has been more than 10 cases of sporadic Burkitt lymphoma reported so far.[5,6,7,8,9] Orbital involvement is uncommon in Burkitt lymphoma. In most cases of orbital involvement, patients usually present with proptosis associated with ocular complaints like blurred vision.[6,7,8,9] Our patient is unique in a way that the main pathology was located within the temporal's area subcutaneous tissue extending into the eyelid without other orbital or visual symptoms and with no systemic complaints. In such cases, clinical diagnosis is a challenge thus rendering tissue biopsy and diagnosis invaluable. Despite having detected to have stage IV disease, patient is well prior to and during the presentation. There was no history of symptoms suggestive of infectious mononucleosis. However, no test towards the Epstein Barr virus antibody was carried out in this child as the test is not available in our hospital.

We advise clinicians to be on alert when encountering patients with eyelid swelling, as this could come as presenting feature of Burkitt lymphoma without other systemic symptoms. A delay in diagnosis may lead to aggressive orbital involvement with devastating vision loss, as Burkitt lymphoma tends to progress rapidly. In patients with orbital involvement, irreversible vision loss can be prevented with early detection and prompt treatment, as Burkitt lymphoma responds well to treatment.[6,7,9,10] Burkitt lymphoma, once considered an aggressive tumour, has enjoyed an increase in 5 year survival of over 90 percent with the recent introduction of refined inexpensive chemotherapy in patients with age group of less than 15 years old.[10]

CONCLUSION

Burkitt lymphoma should be a differential diagnosis in a paediatric patient presenting with rapid onset painless eyelid swelling, despite being in a non-endemic country. Urgent biopsy is required to allow further staging and initiation of treatment. Early prompt multidisciplinary approach and effective chemotherapy would ensure good clinical and visual outcome of this rare condition.

Conflict Of Interest: No conflict of interest

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