

Hodgkin's Lymphoma: A Rare Ophthalmic Case Report

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Abstract

Hodgkin's lymphoma (HL) is a disease originating from lymphoid tissues; however, it may pose a diagnostic challenge. Ocular involvement is more prevalent in non-HL compared to HL. We have reported a rare case of Hodgkin's lymphoma presented with forwarding bulging of left eye with pain, redness, and watering. The patient also complained of painless swelling of the submandibular lymph node when admitted into the National Institute of Ophthalmology and Hospital (NIOH), Dhaka, Bangladesh; later, incision and biopsy was done at ENT & Head-Neck Cancer Hospital, Dhaka, Bangladesh. Histopathology report revealed Hodgkin's lymphoma. Hodgkin's lymphoma is a malignancy with unknown aetiology. With timely diagnosis and early treatment, our patient began to show gradual improvement with her symptoms. Prompt oncologic treatment and immunotherapy can be beneficial, if instituted early in the course of the disease.

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Introduction

Ocular and adnexal lymphoma requires a meticulous ophthalmic and systemic evaluation, advanced imaging array helps in localizing the disease, open biopsy is preferable than fine-needle aspiration biopsy (FNAB). Complete excision is often impossible. Histopathology and immunohistochemistry analysis is recommended for the final diagnosis. Non-Hodgkin lymphoma (NHL) is expected in the orbit and adnexal area; however, Hodgkin's lymphoma (HL) rarely occurs in the orbit and adnexal area.¹⁻³ Hodgkin's lymphoma is a potentially curable disease, and the World Health Organization classifies HL into four distinct types: nodular sclerosing, mixed cellularity, lymphocyte depleted, lymphocyte rich, nodular lymphocyte predominant. Clinical features include asymptomatic, unexplained fever and weight loss, cough, shortness of breath, pruritis, and palpable nontender lymphadenopathy.^{3,4} Here, we have presented a rare case on Hodgkin's Lymphoma involving the orbit.

Case Presentation

A 14-year-old girl presented with complaints of forwarding bulging of the left eye for one month. She also complains of pain, lacrimation, redness of her left eye, and painless swelling in the left submandibular lymph node area for the same duration. After admission into the National Institute of Ophthalmology and Hospital (NIOH), Dhaka, Bangladesh, we evaluated the patient thoroughly. Her visual acuity was counting fingers 2 feet in the right eye and Perception of light in the left eye. She had severe axial proptosis (12 mm) of the left eye. Left eyelids were swollen, conjunctiva of the left eye was hugely chemosed (Grade-II). The left cornea was severely eroded

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and ulcerated. Ocular motility was grossly restricted in all gazes of the left eye. Tender feelings on palpating the left eye were experienced. Corneal sensation was absent in the left eye. Aphakia was observed in the right eye. The left eyeball was not compressed. Valsalva manoeuvre was negative. She had been experiencing a similar condition 1 month back, and the patient was admitted to another tertiary based multi-specialized hospital and diagnosed as an orbital pseudotumor that was treated conservatively. She also gave a history of an ocular (cataract) surgery in her right eye seven months ago. She didn't give a history of consanguinity of Marriage. She has properly fulfilled her immunization Schedule. Imaging investigations were performed. The thyroid function test showed normal values. MRI of brain and orbit revealed that ill-defined enlargement of extraocular muscles involving the tendon involving left orbit rather than right orbit with forwarding protrusion of the left globe suggestive of non-specific orbital inflammatory disease. The features of a CT scan of the brain and orbit are also suggestive of non-specific orbital inflammatory disease of both eyes.

Swollen left submandibular lymph node underwent incision biopsy by an ENT surgeon at ENT & Head-Neck Cancer Hospital, Dhaka, Bangladesh, and histopathology report revealed Hodgkin's lymphoma. Differential diagnoses included anaplastic large cell lymphoma. The histopathologic appearance of the right-sided lymph node also favours Hodgkin lymphoma.

We treated her with Systemic (intravenous) Methylprednisolone (500mg) for three consecutive days, with the addition of topical and systemic broad-spectrum antibiotics. Conservative treatment was given to protect the

cornea and treat the corneal ulcer. Then, we referred the patient to an oncologist to manage the Hodgkin's Lymphoma. The patient was treated with chemotherapy and improved on the treatment during the follow up. The corneal ulcer was regressed and leaves a corneal scar.



Fig. 1. a 14-years-old girl presented with huge proptosis with corneal ulceration of the left eye



Fig. 2. Axial CT image shows ill-defines enlargement of the left medial rectus muscle involving the tendon

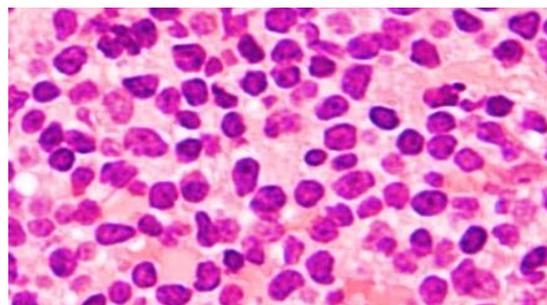


Fig. 3. Photomicrograph of Hodgkin's Lymphoma with Hodgkin lymphoma (NLPHL), with a Reed-Sternberg cell.

Discussion

Non-Hodgkin Lymphoma (NHL) is the most common ocular adnexal malignancy, and it includes 2% of all lymphomas and approximately 10% of all extranodal lymphomas and comprises 30% of orbital tumours. Most orbital lymphomas are B-cell NHL. The relative frequencies of OAL are in orbit, 37%; conjunctiva, 29%; lacrimal, 20%; and eyelid, 14%. Unilateral 85% and the primary tumour is 72%.⁵

HL very rarely involves the eye and adnexa, whereas NHL is the most common ocular and adnexal lymphoma occurrence.⁶ Hodgkin lymphoma (HL) most commonly occurs in people between 14 and 30 years old and those over 55. Family history of Hodgkin lymphoma and history of NHL are also risk factors of Hodgkin lymphoma. Classic HL is the most common and comprises 90 per cent of HL.⁷⁻⁹ Clinical features of OAL are painless proptosis, ptosis, palpable rubbery eyelid growth, Motility deficits (diplopia), and salmon patch appearance of the conjunctiva. The painful lesion may present in 10% to 30% of high-grade lymphoma.^{10,11} CT scan of the orbit not only helps in identifying the extent and location of the lesion but also helps in planning out the management strategies.

OAL is typically seen as well-circumscribed, homogeneous, hyper to iso dense lesion and mould the globe, and other orbital tissues that may appear as the pancake-like configuration. Tissue invasion and bone erosion is usually seen in high-grade lymphoma. Lymphomas usually are iso-intense on both T1 and T2 weighted images of MRI and show moderate enhancement with gadolinium. Newer imaging techniques like PET-CT scan may be performed to identify the initial onset of malignant tumour, determine whether cancer has spread or metastasized in the body,

and assess the effectiveness of the treatment plan. PET-CT scans are used to determine how far cancer has spread to other organ systems.¹⁰⁻¹² Open biopsy is preferable rather than fine-needle aspiration biopsy (FNAB) to obtain representable tissue specimen. Tissues for histopathological examination are sent in formalin for histopathology and as fresh tissue for flow cytometry and gene rearrangement studies. TNM Staging helps in documenting the disease, prognosticating, and planning the treatment approach and follow-up strategies. Proper initial Staging is important and should include total-body positron emission tomography bone marrow biopsy.¹²⁻¹⁴ Cancer cells may involve not only the lymph nodes but also one or more organs and tissues.²⁻⁴ Hodgkin's lymphoma is a curable disease. The goal of treatment is to kill the malignant cells and for remission of the disease. Chemotherapy is the treatment of choice. Radiotherapy can be used after chemotherapy or alone for early-stage nodular lymphoma. Bone marrow transplant is reserved for recurrent disease. Recent modality treatment includes immunotherapy and targeted therapy.³⁻⁵

Conclusion

Hodgkin's lymphoma is rarely involving the orbit and adnexa. Histopathology is the main diagnostic tool to confirm the Hodgkin's lymphoma. It is potentially curable, if detect and manage early. Chemotherapy is still the mainstay of treatment.

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