



Diffuse Large B-Cell Lymphoma Masquerading as an Indolent and Seemingly Benign Skin Lesion

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Abstract

A 55-year-old woman complained of increased swelling in her left periorbital area and forehead. Her medical history told us that over the past two years, she had repeated occurrences of mild, painless, non-pulsatile scalp swellings that were only over her head and unrelated to any underlying tissue. Over the last three months, the scalp lesions have grown to cover most of the scalp and become undulating, giving the head an uneven appearance. She developed complete left ophthalmoplegia upon evaluation, with no history of any constitutional symptom. A brain scan with contrast-enhanced MRI showed diffuse pachymeningitis and hypo-intense thickening of soft tissues near the left orbital apex and left cavernous sinus. All blood reports pertaining to the differentials and cerebrospinal fluid analysis were within normal ranges. A biopsy confirmed that the lesion in the skull was a diffuse large B-cell lymphoma of the GCB type. Finally diagnosed with NHL, our patient presented with an unusually isolated skin illness that had been indolent for two years, without any constitutional symptoms. The delay in diagnosis could have been avoided if we had set a lower criterion for skin biopsy for recurrent skin lesions at the time of initial presentation. Our case strongly conveys that we should not underestimate any skin lesion, consider all possible secondary differentials during the initial presentation, and act as soon as possible.

Keywords: B-Cell Lymphoma Masquerading; Periorbital Area; Lymphoma; Ophthalmoplegia

Abbreviations

MRI: Magnetic Resonance Imaging; NHL: Non-Hodgkin's Lymphoma; CSF: Cerebrospinal Fluid; GCB: Germinal Center B-cell; ABVD: Adriamycin, bleomycin, vinblastin, and dacarbazine.

Introduction

Lymphoma is the sixth most common cause of malignancy, and diffuse large B-cell lymphoma accounts for 20–30%

of non-Hodgkin's lymphoma (NHL). Fever, night sweats, appetite loss, weight loss, pedal edema, exhaustion, pain in the lymph nodes or organs, itching, chest discomfort, shortness of breath, lump in the neck, armpit, or groin, and other symptoms specific to a particular organ are all signs of B-cell lymphoma. We report here an uncommon presentation of the disease, where the lesion that initially appeared benign turned out to be a sign of an impending disaster. Our case is worthy of reporting as it sends a strong message to the medical community that we should not underestimate any skin lesion and consider all possible differentials secondary

to it.

Case Report

A 55-year-old female presented with complaints of progressive left periorbital and forehead swelling for seven days. We got history that for the last two years, she used to have recurrent episodes of multiple small swellings over the scalp that were painless, non-pulsatile, and not fixed to underlying structures. They used to come as single lesions initially, but later cropped up at multiple sites only on the scalp; the lesions started as plaque, gradually increased in size over a few weeks, and then regressed, only to reappear a few weeks later. Since the last three months, the scalp lesions have become undulating and covered most of the scalp to make it an irregular surface. The new-onset periorbital swelling was associated with drooping of the left eyelid along with pain, watering, and restriction of left ocular movement. On examination, she had complete left ophthalmoplegia; the left pupil was dilated and sluggishly reacting to light; the fundus and other examination findings were within normal limits. She had no history of fever, night sweats, or weight loss. MRI of the brain with contrast revealed T2 iso to hypointense soft tissue thickening next to the left cavernous sinus and left orbital apex, involving the posterior part extraocular muscle and optic nerve (had post contrast enhancement) along with diffuse pachymeningitis. Considering orbital apex syndrome, we kept possibilities of infective and non-infective granulomatous diseases (eg. tuberculosis, fungal infection, sarcoidosis), lymphoproliferative disorder, histiocytic disorder, IgG4-related diseases, and vasculitis. His blood report (related to the differentials) was unremarkable except mild anaemia. The routine blood test revealed: haemoglobin of 11 gram/ decilitre, total leucocyte counts of 5,000/ cubic millimetre, platelet count of 1.5 lacs /microlitre, ESR (Erythrocyte Sedimentation Rate) of 20 millimetre/ hour, C-reactive protein of one milligram/decilitre and normal peripheral smear examination. Antinuclear antibody test, Serum Angiotensin Converting Enzyme level were within normal level. The cerebrospinal fluid (CSF) picture revealed: cell count of 3/ cubic millimetre (100% lymphocytes), protein of 40 milligram/ decilitre and glucose 73 milligram/decilitre (corresponding blood glucose of 120 milligram/decilitre), CSF adenosine deaminase of 0.5 Unit/ Litre. The comprehensive central nervous system infection panel and Gene X-pert for tuberculosis were non-contributory.

An excisional biopsy of the skull lesion revealed diffuse large B-cell lymphoma, GCB (Germinal Center B-cell) type (Figure 1). The diagnosis was confirmed with immunohistochemistry to detect specific antigens CD3, CD20, CD34, EMA, CK, CD10, BCL2, BCL6, CMYC, MUM1, KI67. Amongst them, antigens CD20, CD10, BCL2 (50%) and KI67 (60%) were found to be positive. The microphotograph of the lesion is shown in

Figure 2.

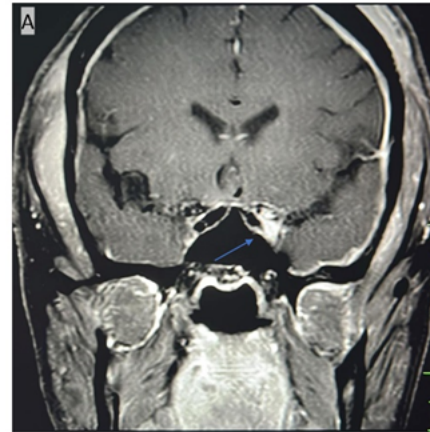


Figure 1A: MRI of the brain with contrast revealed soft tissue thickening next to left cavernous sinus and left orbital apex with contrast enhancement (blue arrow), along with diffuse pachymeningitis.

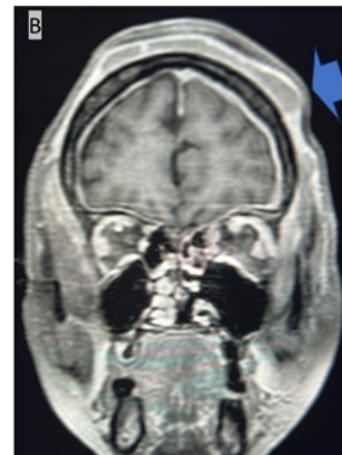


Figure 1B: Undulating calvarium due to lymphomatous deposits (Thick blue arrow).

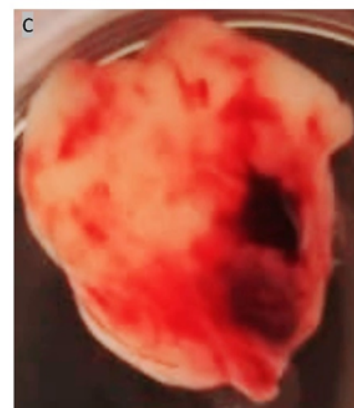


Figure 1C: The soft tissue material obtained by excisional

biopsy.

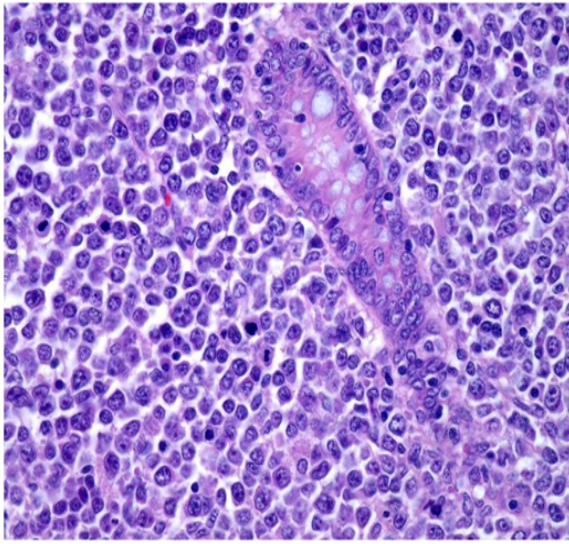


Figure 2: Microphotograph of the biopsied sample.

Discussion

About 25–40% of non-Hodgkin's lymphoma has primary extranodal disease as a presentation [1]. Our case of NHL presented with isolated skin disease for 2 years without any constitutional symptoms.

Though malignant lymphoma may mimick benign skin lesions [2,3], our patient's skin lesions appeared only in the scalp a rare presentation. The delay in diagnosis in our case could have been prevented if we had a lower threshold for skin biopsy for the recurrent skin lesions during the initial presentation. Darine Hakkou et al. reported that a 30-year-old man had a severe nodular lesion on his skin. Immunohistochemical staining and biopsy results showed that the lesion was consistent with classic Hodgkin's lymphoma. Adriamycin, bleomycin, vinblastin, and dacarbazine (ABVD) treatment led to the patient's complete recovery [4].

NHL usually spreads hematogenously, unlike Hodgkin lymphoma, where dissemination is via lymphatics. Because

of the hematogenous spread, most cases of NHL present with advanced disease (stage III/IV). Diffuse large B-cell lymphoma is a type of NHL with a very aggressive presentation and often presents with a rapidly growing mass (unlike our case). We learned why multisystemic manifestations of all neurological disorders need to be identified even when the neurological symptoms have not surfaced.

We conclude that we should not disregard any clinical sign, as even a seemingly insignificant lesion could serve as the initial indication of a serious illness. To prevent a disastrous outcome, a thorough assessment of the lesion is required (much like a skin biopsy of the lesion in our situation).

Ethical Statement

Informed verbal consent has been obtained from the patient regarding the case report. Ethical approval was obtained for this case report and clearance obtained from the Institutional Ethical Committee (Apollo Multispeciality Hospitals, Kolkata, West Bengal, India).

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