Conjunctival MALT lymphoma: an unusual cause of red eye

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Summary

We describe a patient presenting with a red eye who was found to have conjunctival non-Hodgkin's lymphoma of the mucosa-associated lymphoid tissue (MALT) type.

Keywords: conjunctiva; lymphoma; MALT; red eye

A 79-year-old man presented with a 3-week history of a painless, red left eye. There was no discharge and minimal irritation. The vision in his left eye had deteriorated over several months secondary to cataract. His ophthalmic history included right cataract surgery one year previously. There was no relevant medical history.

On examination, right visual acuity was 6/9 and left was 6/18. He was found to have an infected left eye with an enlarged left caruncle and a fleshy 'salmon-pink' swelling that extended into the inferior forniceal conjunctiva (figure 1). The remainder of the ocular examination revealed a left cataract but no other abnormalities. There was no preauricular or submandibular lymphadenopathy and general examination confirmed localised disease. A biopsy of the inferior conjunctiva was performed.

Orbital computed tomography (CT) scans showed a soft tissue swelling anterior to the left globe. Conjunctival biopsy revealed a lymphoid infiltrate of the connective tissue and epithelium (figure 2). Immunostaining showed predominantly B cells with small numbers of T cells. This was consistent with a diagnosis of low grade non-Hodgkin's lymphoma of the MALT type (mucosa-associated lymphoid tissue). Blood tests including bone profile, serum immunoglobulins, marrow biopsy and

Figure 2 Biopsy specimen showing conjunctival epithelium and underlying stroma infiltrated by small lymphoid cells. Haematoxylin & eosin; orig magnification x 100

CT scans of head, neck, chest, abdomen and pelvis confirmed that the disease was localised.

He was referred to the radiotherapists for treatment. He was reviewed in the eye clinic 3 months later and found to have new conjunctival lesions in the right eye. A second biopsy of the right inferior forniceal conjunctiva showed a focal infiltrate of B and T cells with lymphoepithelial lesions consistent with the working diagnosis.

In view of the multifocal nature of his disease chemotherapy was commenced with oral chlorambucil. After six courses his lesions improved and the conjunctival infection resolved. He remains under regular review.

Discussion

In the early 1980s the term MALT was coined in order to describe a characteristic arrangement of lymphoid tissue found in certain mucosal surfaces.¹ Mucosa-associated lymphoid tissue is now recognised as giving rise to a specific form of tumour with distinct features, distinguishing MALT lymphoma from other forms of primary, non-Hodgkin's, extranodal lymphoma.^{2,3}

The commonest site of MALT in humans is the gastrointestinal tract, specifically stomach, terminal ileum and appendix.² It has also been described in the bronchus, salivary, thyroid and thymus glands. It was described in the conjunctiva in 1980 by Knowles and Jakobiec.³ Lymphoid tumours of the conjunctiva can pose problems in diagnosis since there is considerable overlap between benign and malignant lesions. Under normal circumstances the conjunctiva contains small numbers of

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Accepted 7 December 1998

Figure 1 Photograph of the left eye showing a fleshy, 'salmon-pink' lesion arising from the caruncle

MALT: characteristic histological features

- submucosal B cell follicles
- mantle zone of small lymphocytes
- region of B cells extending from mantle to overlying epithelium with centrocytic appearance
- lympho-epithelial lesions: extension of B cells into overlying epithelium

Box 1

lymphocytes together with a few mast cells and Langerhans' cells. This conjunctival-associated lymphoid tissue is associated with the production of local immune responses.^{4 5}

The majority of lymphoid tumours at this site are low grade B cell neoplasms.⁴ They are composed mainly of small lymphocytes with occasional larger blasts. Some tumours have a plasmacytoid morphology and pure plasma cell neoplasms are found rarely. High-grade B cell lymphomas are usually of the lymphoblastic type.⁶ Conjunctival lymphoma may be limited to this site or it may be associated with systemic disease. The characteristic histological features of MALT are listed in box 1.^{7 8}

The behaviour of MALT is different from other B cell extranodal non-Hodgkin's lymphoma.^{6 9} It is less aggressive and tends to

Summary points

- in cases of 'red eye' a careful examination of all ocular structures should be made
- the differential diagnosis of red eye includes conjunctival lymphoma
- a low threshold for biopsying suspicious lesions is required
- MALT are not aggressive, respond well to radiotherapy, and carry a good prognosis

Box 2

remain localised to mucosal surfaces. It also has a better prognosis.

Conjunctival MALT usually presents with mild symptoms of redness, irritation and photophobia.⁷ Examination findings are of orange-pink, salmon-pink or pale masses in the upper or lower fornices and are often bilateral. In all known cases no recurrence or dissemination after treatment has been described.⁷ The mainstay of treatment is local radiotherapy.¹⁰ However, chemotherapy is an alternative when radiation is inappropriate.

This case illustrates an unusual cause of unilateral red eye and highlights the importance of careful ocular examination and the value of a histological diagnosis.

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Acute lymphoblastic leukaemia presenting with arthritis in an adult patient

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Summary

The earliest manifestations of leukaemia often include rheumatic signs and symptoms. Arthritis is a well recognised complication of leukaemia in children, but acute and chronic leukaemia may also cause arthritis in adults. Leukaemic arthritis may occur at any time during the course of leukaemia and may be the presenting manifestation. It should therefore be considered in the differential diagnosis of both childhood and adult rheumatic disease. We present an adult patient presenting with arthritis due to acute leukaemia.

Keywords: arthritis; leukaemia; synovial fluid analysis

Figure 1 Ankle radiographs showing soft tissue swelling and joint effusion

A significant number of patients with leukaemia have bone or joint symptoms as a part of their disease.¹ However, arthritis as an initial manifestation of leukaemia is extremely rare.² Leukaemic arthritis (LA) may be defined as joint pain and swelling in association with peripheral blood or bone marrow leukaemia after other causes of arthritis have been excluded.³ Early in the course of the disease there may be difficulty in arriving at the correct diagnosis when studies of peripheral blood are not diagnostic.

We report an adult patient presenting with arthritis whose peripheral smear was normal. However, examination of synovial fluid and bone marrow revealed LA and acute lymphoblastic leukaemia (ALL).

Case report

A 22-year-old man was referred to our hospital with complaints of pain and swelling of his ankles for the last 3 months. On initial evaluation, he was conscious and fully oriented. Body temperature, blood pressure, pulse rate were 38.3°C, 130/80 mmHg, and 86 beats/min, respectively. On physical examination there was diffuse swelling and tenderness with erythema and discharge in both ankles. Other physical findings were within the normal range. Haematological studies revealed a haemoglobin level of 9.6 g/dl, leukocytes $10.3 \times 10^9/l$ (55% neutrophils, 22% lymphocytes, 19% monocytes, 4% eosinophils), platelets $310 \times 10^9/l$, with an erythrocyte sedimentation rate of 103 mm/h. Blood biochemical values were normal. Serum uric acid was 5.4 mg/dl (normal range 3.5–8.5 mg/dl). Antinuclear antibody, rheuma-

toid factor, and serological tests for hepatitis B were negative. Anti-DNA and serum complement level were within normal limits. Radiographs of the ankles showed soft tissue swelling and joint effusion (figure 1). The synovial fluid could not be aspirated during the first arthroscopy. The patient was prescribed indomethacin and colchicum for the pain, but during the following 2 weeks there was no benefit and the patient's symptoms increased, with complaints of pain and swelling of knees. Arthroscopy was repeated and cytologic examination revealed immature blastic cells (figure 2). Flow cytometric analysis of synovial fluid revealed CD19 58%, CD20 70%, and HLA-DR 91%. A bone marrow aspiration and biopsy was performed. Infiltration of leukaemic cells was seen on bone marrow aspiration (figure 3), and a diagnosis of ALL (L2, B-cell origin) was made. Mitoxantrone, vincristine and prednisolone were

Figure 2 Synovial fluid examination revealing immature blast cells

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Accepted 17 December 1998