CASE REPORTS

Immunoblastic lymphadenopathy – report of a case

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Summary
A case of immunoblastic lymphadenopathy has been presented which had the characteristic clinical and histological features. The patient did not respond to chemotherapy and succumbed to the disease. Instead of the usual polyclonal gammopathy associated with this condition, he showed a fall in IgM levels, IgG and IgA being within normal limits. The significance of the latter finding is not known.

Introduction
Generalized lymphadenopathy with or without fever and hepatosplenomegaly is seen in a variety of diseases. Lymph node biopsy for establishing the diagnosis may sometimes be essential but the distinction between malignant and reactive proliferation of the lymphoreticular tissue is at times difficult. The entity described as angioimmunoblastic lymphadenopathy with dysproteinæmia by Frizzera, Moran and Rappaport (1974) and as immunoblastic lymphadenopathy by Lukes and Tindle (1975) is situated on the borderline between benign and malignant immunoblastic proliferation and exercises the skill of a pathologist. It is imperative that the condition should be distinguished from Hodgkin's disease as the available evidence indicates that chemotherapy may be harmful to the patient in the former condition.

Case report
A 35-year-old man presented with a history of fever and painful enlargement of inguinal lymph nodes of a month's duration. He had taken a course of tetracycline and chloromycetin without any benefit. A week before admission to hospital he had noticed a painful swelling in the neck. He also had a marked loss of weight and appetite. There was no skin rash or itching.

Examination revealed a generalized lymphadenopathy with involvement of cervical, axillary and inguinal lymph nodes. The liver was not enlarged and the spleen was just palpable. In addition, the patient was febrile and had mild pallor.

Laboratory investigations
Haemoglobin was 11.6 g/dl, WBC 6.8 x 10^9/l (32% neutrophils, 48% mature lymphocytes, 15% monocytes, 3% eosinophils and 2% basophils) and ESR was 40 mm/first hour (Westergren). The peripheral smear showed a normocytic, normochromic picture. Serum globulins were 3.4 g/dl. The immunoglobulins as measured by Mancini's radial gel-immunodiffusion technique were as follows (normal values in parentheses): IgG, 163 i.u. (120–246); IgA, 63 i.u. (55–268); IgM, 56 i.u. (108–204). Bone marrow smear showed an increase in the reticulum cells and well differentiated plasma cells. Chest X-ray was normal. An abdominal lymphangiography showed enlargement of the internal iliac and preaortic group of lymph nodes with marked lymphangiectasis. Two lymph node biopsies were taken. Both showed the characteristic histological picture of immunoblastic lymphadenopathy. The patient was given antibiotics for one week and was discharged.

A month later he was re-admitted. The predominant complaint was lymphoedema of the left lower limb. However, fever was continuing. Weight-loss was marked and the lymph nodes had further enlarged. Therapy with cyclophosphamide (200 mg/day) was started. The patient's general condition rapidly deteriorated and he died 12 days after starting chemotherapy. Permission for a post-mortem was refused.

Pathological features
Sections from two lymph node biopsies measuring 1 x 0.8 x 0.6 cm and 2 x 1 x 1 cm were stained with haematoxylin eosin, periodic acid Schiff, methyl green pyronin and reticulin.
Both biopsies showed similar histological features. The lymph node architecture was completely effaced. There was a marked proliferation of small branching capillaries lined with plump endothelial cells (Fig. 1). The lymph node was infiltrated by a polymorphous population of cells and it showed a marked depletion of lymphocytes. The cellular infiltrate was composed of plasma cells, immunoblasts, and a few lymphocytes (Fig. 2). The immunoblasts were large polygonal cells having a moderate amount of amphophilic cytoplasm, an oval large nucleus with finely reticulated chromatin and prominent nucleoli. An occasional binucleate Sternberg–Reed-like cell and a few mitotic figures were also seen (Fig. 3). In a few areas there was an amorphous eosinophilic material separating the cellular infiltrate. Similar material was also seen deposited in the walls of the vessels.
Case reports

Discussion

The clinical manifestations of this case diagnosed histologically as immunoblastic lymphadenopathy were similar to those described by Lukes and Tindle (1975) and Frizzera et al. (1974). The disease developed rapidly, presenting with fever, lymphadenopathy and loss of weight, and was clinically diagnosed as Hodgkin's disease.

The histology of lymph node biopsy showed the characteristic features of immunoblastic lymphadenopathy, fulfilling the four essential criteria laid down by Lukes and Tindle (1975). These are:

(a) diffuse involvement of the lymph node;
(b) proliferation of immunoblasts, plasma cytoid immunoblasts and plasma cells with lymphocyte depletion;
(c) proliferation of arborizing small capillaries;
(d) deposit of an amorphous eosinophilic interstitial material.

Most of the patients with this disease show polyclonal gammmopathy in the serum. However, this patient showed normal values of IgG and IgA and a subnormal IgM level. The significance of this finding is not known. Definite hyperglobulinaemia was found in thirteen out of fourteen cases analysed by Lukes and Tindle (1975), whereas one patient showed no abnormality in the serum globulins. Deficiency of any of the immunoglobulins has not been reported so far.

The cytotoxic therapy was of no avail in this case and the condition of the patient deteriorated rapidly after starting the treatment. This was also the experience of Kalus (1976).

The aetiology of this condition is as yet unknown. Lukes and Tindle (1975) consider it as a hyperimmune proliferation of the B-cell system which may be triggered off as a result of hypersensitivity reaction to therapeutic agents. Schultz and Yunis (1975) reported a case of immunoblastic lymphadenopathy associated with a prolonged administration of liver extract, suggesting that the syndrome may be caused by chronic antigenic stimulation. Occurrence of malignant lymphoma is well known in association with disorders of the immune system such as systemic lupus erythematosus, rheumatoid arthritis, Sjögren's syndrome and alpha chain disease (as cited by Rappaport and Moran, 1975). Immunoblastic lymphadenopathy, although not considered essentially malignant, has a potential for developing into a malignant tumour of lymphoreticular tissue. In the absence of a post-mortem in this case, the possibility of a malignant transformation of the lesion could not be excluded. In the series of Lukes and Tindle (1975), three cases of immunoblastic lymphadenopathy showed a transformation to immunoblastic sarcoma.

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