

UVEAL LYMPHOSARCOMA*

BY

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LYMPHOMATOUS tumours involving the lids and orbit have been recognized and repeatedly reported in the literature for many years; intra-ocular involvement on the contrary is rare if the lymphomatous infiltrations associated with lymphatic leukaemia are excluded. Kukán (1935) and Triebenstein (1920) have each described a case of uveal lymphosarcoma complicated by secondary glaucoma, and other isolated cases of ocular lymphoma have been reported by Hartshorne (1922), McGavic (1943), and Cooper and Riker (1951). Further indication of the comparative rarity of the condition is that, in a series of 67 cases of orbital lymphoma examined by Heath (1948), only three showed involvement of the globe. Of the biopsies of lymphomatous tumours examined in the pathology department at the Institute of Ophthalmology during the past 10 years, only one recent specimen showed evidence of intra-ocular involvement; since this case proved to be of especial clinical and pathological interest in that both orbital and intra-ocular tissues were involved, it has been made the subject of this report.

Case Report

A female aged 59 first attended the out-patient department on May 31, 1951, complaining of redness and severe pain in the left eye of one week's duration. She gave a history of sudden loss of vision in the eye 2 years previously but had been otherwise quite free of symptoms up to the time of the present complaint.

Examination.—The left eye was intensely injected and there was a diffuse and marked corneal oedema. The intra-ocular tension was much increased to digital palpation and the vision was reduced to a bare perception of light but no details of the posterior corneal surface, iris or fundus were visible owing to the extreme corneal oedema. A tentative diagnosis of thrombotic glaucoma was made, and in view of the severe pain the eye was enucleated. On removing the globe it was found that the posterior portion of the orbit was filled with a white gelatinous mass of tissue which histologically consisted of a dense aggregation of closely packed lymphocytes, which not only closely adhered to the posterior sclera of the globe, but also extended into the subarachnoid space and optic nerve (Fig. 1). Sections of the globe itself showed that the choroid, and to a lesser extent the iris and ciliary body, were densely infiltrated with small lymphocytes among which a few immature cells were visible (Fig. 2). The lymphocytic infiltration in the posterior choroid reached massive proportions and the normal choroidal structure was completely destroyed (Fig. 3). The degenerate retina was totally detached and a subretinal exudate was present.

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FIG. 1.—Subarachnoid space invaded; optic nerve shows scattered lymphocytic infiltration. $\times 15$

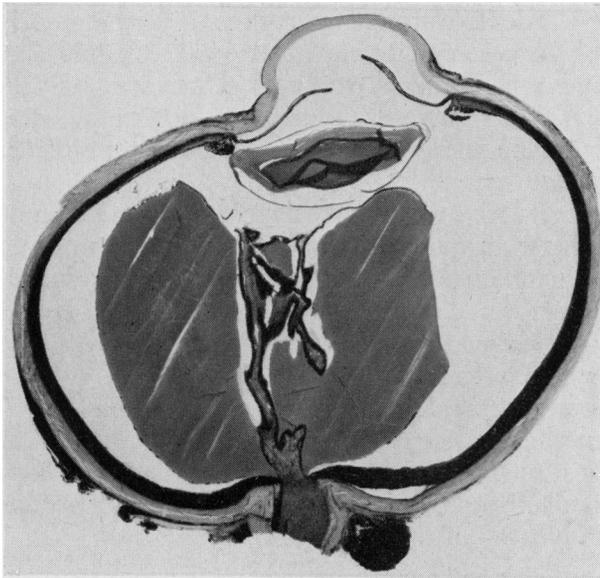
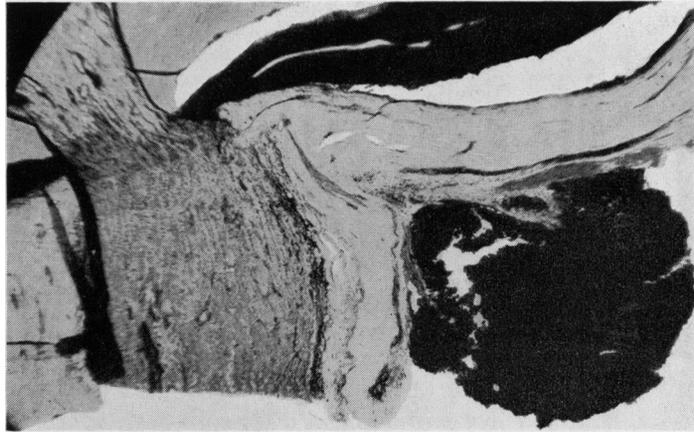
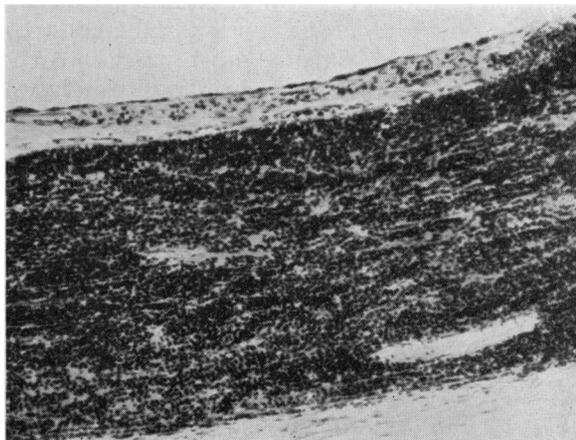


FIG. 2.—Ciliary body and choroid show dense lymphocytic infiltration. Aggregations of closely packed lymphocytes in retro-orbital space adherent to posterior sclera. $\times 3.5$

FIG. 3.—Choroid densely infiltrated by mass of lymphocytes. $\times 90$



The histological appearance was consistent with the diagnosis of either a primary choroidal lymphosarcoma with extra-ocular extension or a combined ocular and orbital lymphosarcomatous infiltration as part of a generalized lymphosarcomatosis.

Laboratory Investigations.—Repeated blood counts and a sternal puncture showed no abnormality.

Follow-up.—After an uneventful recovery, a course of deep x-ray therapy was given to the socket and the patient was discharged. She has been examined at intervals since her discharge from hospital and up to the present time there is no evidence of recurrence in the socket or of any systemic lymphosarcomatous involvement.

Discussion

As is well known, proliferation of lymphoid tissue is a common manifestation of chronic infection and as such may occur in almost any part of the body, but the early pathological differentiation between the hyperplastic and the neoplastic lymphoid proliferations is in many cases a matter of extreme difficulty, the histological line of demarcation between them being but poorly defined in the initial stages. Thus early biopsy of a lymphoid nodule may show an apparently benign proliferation of lymphoid tissue, and only in the later stages may the occurrence of a generalized lymphadenopathy or of blood changes of a leukaemic type reveal the truly malignant nature of the condition.

In recent years many attempts have been made to classify the group of closely related tumours arising from the cellular elements of the lymphatic system, but a most unfortunate confusion of terminology has resulted.

The classification of neoplastic disease generally is founded upon cytological factors, and rests primarily upon the identification of the predominating cells and their differentiation.

It is upon this basis that Lumb (1952) has introduced a pathological classification of lymphoid tumours which, although allowing for much histological overlap, constitutes a practical scheme of subdivision conforming with both pathological and clinical characteristics. In Lumb's classification four groups are defined according to the identity of the predominant cells:

- (1) Predominant lymphocytic differentiation (lymphocytic lymphoblastic lymphoma, lymphosarcoma, and follicular lymphoma).
- (2) Predominant reticulin cell proliferation (reticulin cell sarcoma).
- (3) Mixed cell differentiation in which occur lymphocytes, reticulin cells, and other cells formed by the primitive mesenchymal cell (Hodgkin's disease and lympho-reticular lymphoma).
- (4) Anaplastic sarcomata (including all the anaplastic varieties of the above three groups).

Within each of these groups a fairly well differentiated picture may be found, and the disease may correspondingly vary in severity from a benign stage, with a survival prognosis of many years, to an intensely malignant form, with a prognosis of only a few months. It is now widely accepted that the appearance of an isolated lymphoma anywhere in the body may be

the precursor of a generalized lymphomatous dissemination; and, although such solitary lesions not uncommonly occur, it must be emphasized that these often constitute a transient phase which will eventually progress into a generalized process if the patient survives long enough.

This fact is well illustrated by a case cited by Feinstein and Krause (1952) of a conjunctival lymphoma which was kept under observation for 18 years before it produced systemic manifestations; further confirmation is provided by Heath (1948), who in his review of conjunctival lymphomata, found that, even when the lesions were small and immediately removed, 50 per cent. of the cases ultimately showed evidence of general disease.

The long-term prognosis of isolated ocular lesions would, therefore, appear to be uncertain, but lymphomata are characteristically radio-sensitive and the immediate results of early combined surgical removal and x-ray therapy are usually good. An assessment of the probable ultimate prognosis in such cases will be possible only after an extended and extensive follow-up has been carried out.

Summary

A case of uveal and retrobulbar lymphosarcoma associated with glaucoma is reported, and the pathological classification and prognosis of ocular lymphoid tumours are discussed.

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