Arch. Soc. Amer. Oftal. Optom. (1970) 7 - 301.

LYMPHOID TUMOURS OF EYELIDS

BY

ALY MORTADA, M. D.

Cairo - Egypt.

Herbut [1] classified the lymphomatous or reticular tumours as: lymphoma, giant follicular hyperplasia (Brill - Symmer disease), Hodgkin's disease, lymphosarcoma, reticulum cell sarcoma and different types of leukaemias. Cells of these tumours share the same mother reticulum cell. Hogan and Zimmerman [2] wrote "There is probably no aspect of ophthalmic pathology that is more difficult for both the student and the experienced practicing pathologist than the differential diagnosis of malignant lymphoma and reactive lymphoid hyperplasia. At the risk of oversimplifying, lymphoid tumours of the orbit including those arising in the lacrimal grand, like those of conjunctiva, lids and uvea may be placed in three main groups.

At one extreme there is a very small group which is obviously malignant in which the cells are poorly differentiated. There may be cellular pleomorphism but polymorphism is absent. These are the cases of reticulum-cell sarcomas and cases of acute leukaemias.

There is a much larger group of lymphoid tumours of a reactive or chronic inflammatory rather than neoplactic nature. The lesions may be predominant'y proliferation of lymphocytes or of reticulum cells but many other cell types participate as polymorphonuclear leucocytes, eosinophils, plasma cells and macrophages.

The third group is lymphoid tumours composed of relatively pure lymphocytic proliferation. It is possible that some of these might evo!ve into generalized

lymphocytic lymphosarcomas but in our experience this must be a very unusual occurrence. Follow-up studies generally fail to produce evidence of generalized disease and the lesions typically respond to small amounts of radiation.

My series of 22 histopathologically diagnosed lymphoid tumours of eyelids included:

I. Twelve cases of aleukaemic reticulum cell sarcomas of eyelids occurred at any age. After partil excision for biopsy and X-ray treatment all these cases recurred and most of the patients died within five years of the onset of the disease.

II. Leukaemic infiltrations of eyelids with acute leukaemias occurred in 4 cases. All the cases were in children and were fatal within six months of the disease onset inspite of X-ray and supportive measures. They included:

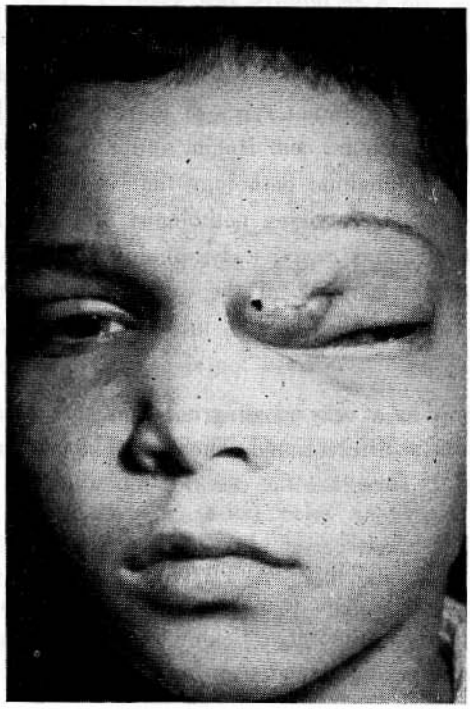

Fig. 1. (case 1) Left upper lid aleukaemic reticulosarcoma with monocytosis of one months duration in a boy aged 9 years.

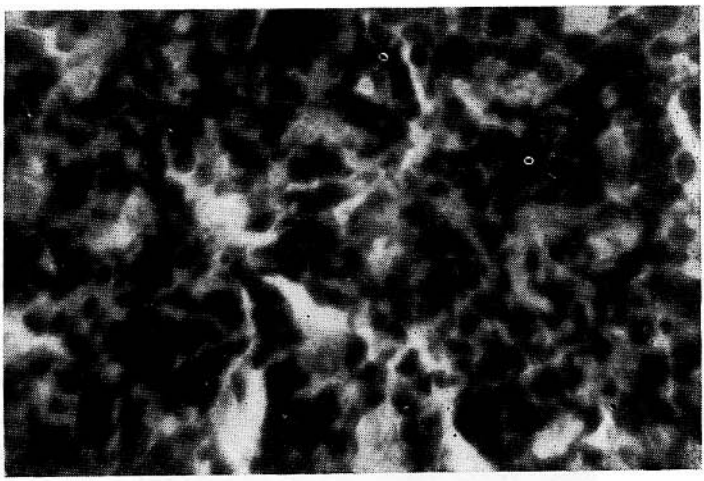

Fig. 2. (case 1) Eyelid reticulosarcoma showing malignant reticulum Cells. Note irregularity in size, shape and staining properties of the cellas and their nuclei (H. & E. 540).

a) Three cases of eyelids myeloblastic infiltrations with myeloblastic leukaemia.

b) One case of right upper lid monoblastic infiltration with monoblastic leukaemia.

III. 6 cases of aleukaemic eyelid lymphoma of a reactive lymphocytic hyperplasia nature occurred in adult patients. All the cases were cured after partial tumor excision and mild anti-inflammatory doses of X-rays.

Case reports.

The lid lymphoid tumours usually extend also to the conjunctiva, lacrimal gland or orbit. The following 3 cases are chosen to describe the main lymphoid tumours of the eyelids.

Case 1:

9 years old boy (Fig. 1) complained of swelling of his left upper lid of one month duration. His general condition was poor. Both eyes were normal, fundi normal, visual acuity in each eye 6/12.

A soft tumour occupied the left upper lid. There were no enlarged lymph glands, liver or spleen. The blood Wassermann reaction test was negative. The differential blood count showed monocytosis. Red blood corpuscles 4'600.000:

white blood corpuscles 7,100. Basophils 1, Eosinophils 6, staff nucleated 7, segmented 54, lymphocytes 25, monocytes 7. X-ray examination of orbits was normal.

The tumour which was very soft, vascular and friable came out in pieces and was found to involve the upper part of the orbit. Histopathological examination of the removed tumour pieces stained with Haematoxylin and Eosin showed large cells of irregular size and shape held in a reticular stroma. The cells nuclei were large of various size, shape and staining properties (Fig. 2). Foots silver reticulin stain showed the cells to lie on argyrophilic reticular fibres. The picture was consistent with a reticulum cell sarcoma of eyelid. Fresh tumour cells smear showed malignant reticulum cells. The cells were fairly large and of varied shapes, round, polyhedral, oval or irregular. Their cytoplasm was feebly stained. Their nuclei were relatively arge varied in shape, size and staining properties. The

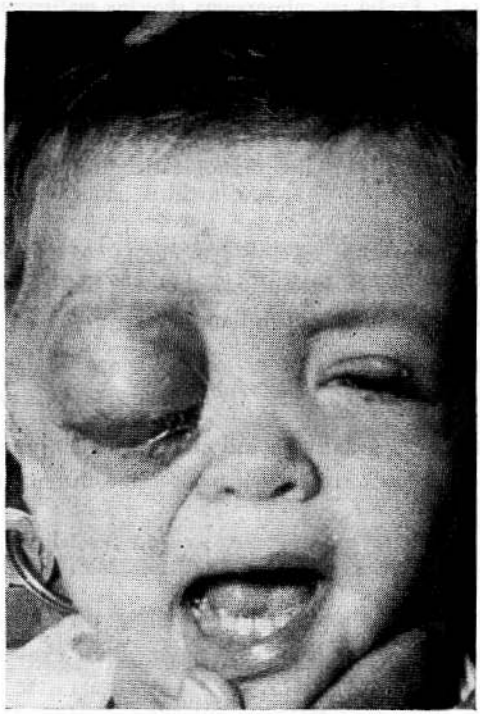

Fig. 3. (aces 2) Right upper eyelid myeloblastic leukaemic infiltrations with myeloblastic leukaemia of one month durations in a male child aged 3 years.

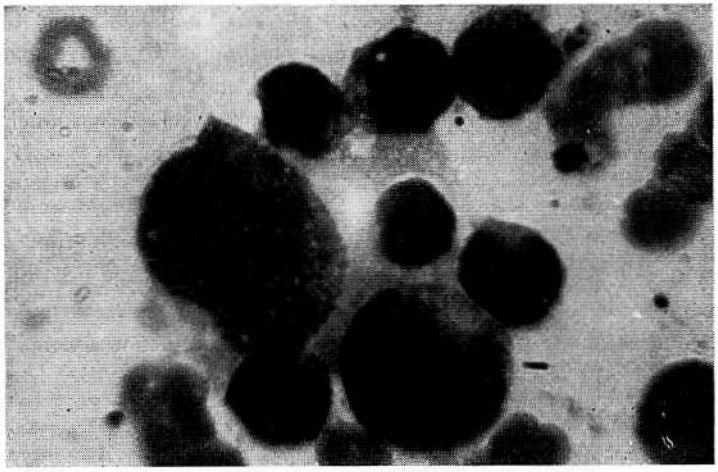

Fig. 4. (case 2) Blood smear showing myeloblasts. (X 1200).

nucleus was vesicular with a sharply defined nuclear membrane and a delicate network of chromatin with granules at the intersections. Many reticular fibres extended between the cells. All the cells present were primitive reticulum cells. There were no differentiated cells.

Inspide of X-ray treatment to the lid tumour and supportive measures the tumour recurred after four months and the girl died within one year of the disease onset.

Case 2:

3 years old boy (Fig. 3) had right upper lid swelling and proptosis of one month duration. The child was anaemic, showing dyspnea on exertion and painful knee joints. Temperature was 37.9 c. and pulse 90. Gums were swollen and bled easily. The liver was not enlarged but the spleen was enlarged one finger below the costal margin. Lymph glands were not enlarged. There were no septic foci.

The right upper lid was swallen due to cellular infiltration. There were right proptosis of 26 mm, limitation of ocular movements and chemosis of conjunctiva. The right fundus showed papilloedema, dilated tortuous retinal veins and fine retinal haemorrhages. The left eyelids and eye were normal.

Skull X-rays, faeces and urine were normal. Blood Wassermann and tuberculin tests were negative .Blood count showed the picture of myeloblastic leukaemia

(Fig. 4), Haemoglobin 40%, red blood corpuscles 3'330.000, white blood corpuscles 22.700; platelets 67.400 basophils 0% eosinophils 3%, myeloblasts 83%, premyelocytes 2%, myelocytes 2%, juneviles 0%, staff nucleated 1%, segmented 2%, lymphocytes 0%, monoblasts 0%, promonocytes 0%, monocytes 0%, macroblasts 0%, normoblasts 6%, and reticulum cells 1%.

The right upper lid tumour was hard grey infiltrating the lid and extending in the upper part of the right orbit. Histopathological examination of a part of the tumour stained with haematoxylin and eosin showed large cells of irregular shape held in a fine reticular stroma. The cells nuclei were large, of varied shape, size and staining properties (Fig. 5). The picture was consistent with myeloblastic infiltrations.

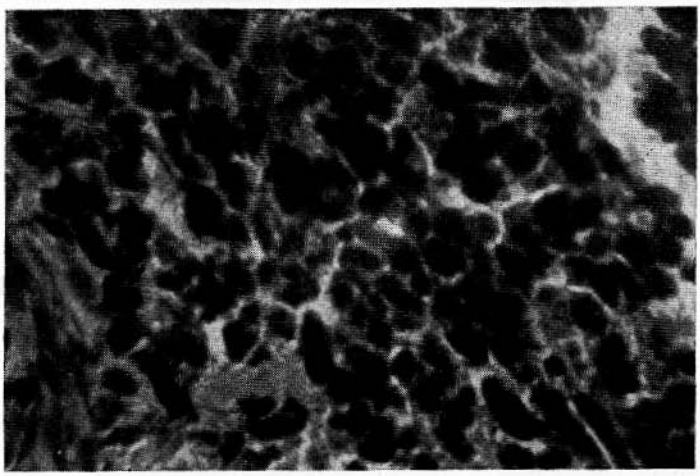

Fig. 5. (case 2) Myeloblastic infiltrations of eyelid, showing cells of different size, shape and staining properties held in a reticular stroma (H. & E. X 540).

The lid tumour was treated by X-ray. Blood transfusion, liver extract, vitamin B-complex penicillin and cortisone were given but the child died one month later from severe epistaxis.

Case 3:

44 years old woman (Fig. 6) complained of a swelling of her left lower lid of 7 months duration. Both eyes were normal with visual acuity in each eye

Fifg. 6. (case 3) Left lower lid beningn reactive lymphocytic hyperplasia of 8 months duration in a 44 years old woman.

6/12. The right eyelids were normal. The general condition was good. Only the left preauricular lymph gland was enlarged. There were no enlarged, liver or spleen. Temperature was normal. The blood total and differential counts were normal.

Histopathological examination of the diffuse lid tumour removed showed diffuse mature lymphocytic infiltration (Fig. 7).

Fresh tumour cells smear (Fig. 8) showed non-malignant reticulum cells, differentiated lymphoblasts and abundant mature lymphocytes. The lid diffuse tumour was a reactive lymphoid hyperplasia.

Treatment continued by mild anti inflamatory doses of X-ray to the affected left lower lid and enlarged left preauricular lymph gland. The left lower lid swell-

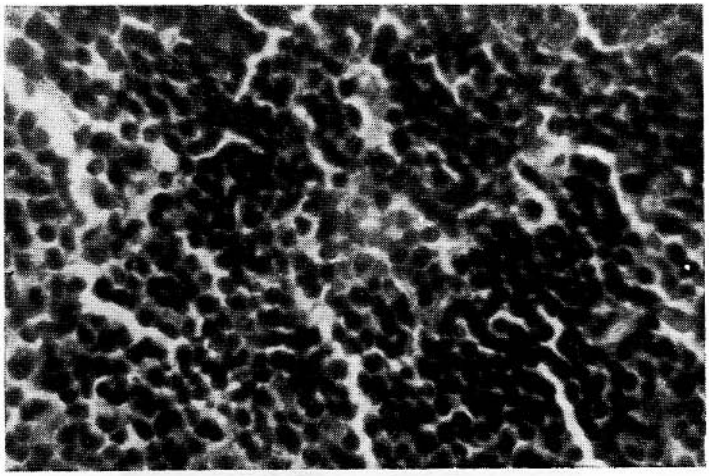

Fig. 7. (case 3) Reactive lyphocytic hyperplasia showing hyperplasia of mature lymphocytes. (H. & E. X 540).

ing and enlarged preauricular lymph gland disappeared (Fig. 9). For 6 years follow up there was no tumour recurrence, leukaemic blood changes, or enlarged lymph glands.

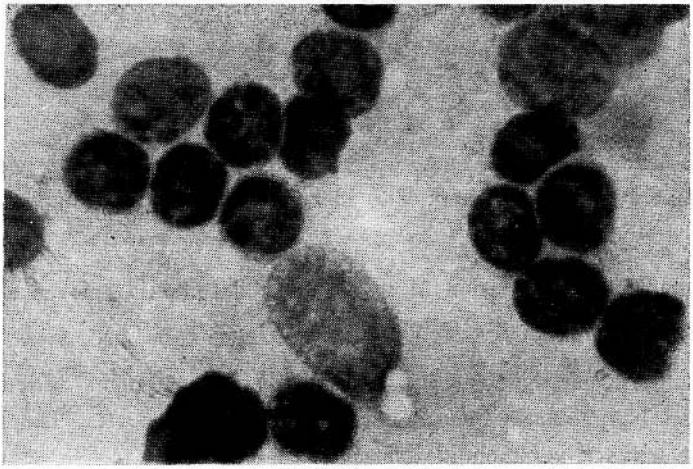

Fig. 8. (case 3) Benign reactive lymphocytic hyperplasia cells smear showing non-malignant reticulum cells, differentiated lymphocytes (Giemsa stain X 1200).

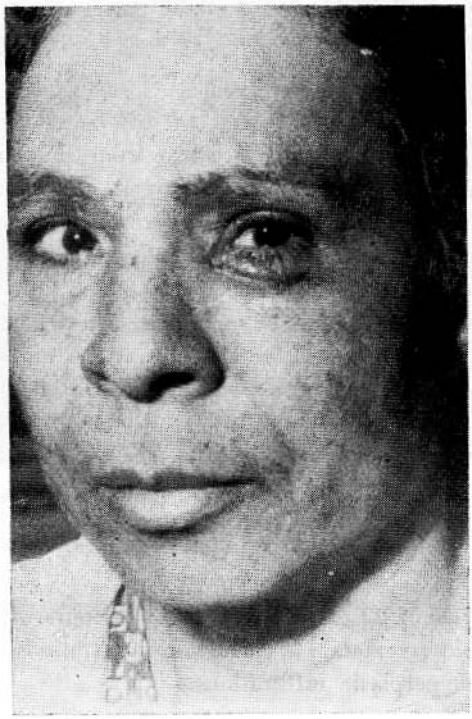

Fig. 9. (case 3) The woman after anti-inflammatory doses of x-ray to affected left lower lid. Shwoing absence of left lower lid swelling.

Comment

Mortada [3] described nine cases of orbital reticulum cell sarcomata showing that they are the commonest malignant tumours of the orbit in Egypt. In one of these cases the orbital tumour extended from the eyelids. Mortada [4] described 3 cases of acute leukaemias accompanied by orbital and lids leukaemic infiltrations. These cases included a case of myeloblastic infiltrations with myeloblastic leukaemia a case of monoblastic infiltrations with monoblastic leukaemia and a case of lymphoblastic infiltrations with lymphoblastic leukaemia, The 3 cases occurred in children and had a rapid fatal termination. Mortada [5] described 4 cases of orbital, lid, lacrimal gland and conjunctival partly encapsulated or nonencapsulated infiltrating masses composed of mature lymphocytes of a benign

reactive hyperplasia nature. Many of these cases may histopathologically erroneously diagnosed as lymphosarcomata.

Mortada [6] stresses that in a section fixed in formalin and stained with Haematoxylin and Eosin it is usually difficult to differentiate orbital lid ,lacrimal gland and conjunctival reticulum cell sarcoma from monoblastic, myeloblastic or lymphoblastic leukaemic infiltration. Also it is sometimes difficult to differentiate malignant lymphoma from benign reactive lymphoid hyperplasia. Differentiation of the different lymphoid tumours is easier and quicker after examining their tumours cells smears stained with Giemsa stain. In case of reticulosarcoma the smear shows malignant reticulum cells, reticular fibres but no differentiated cell, in monoblastic leukaemic infiltrations monoblasts; in myeloblastic leukaemic infiltrations myeloblasts and benign reactive lymphoid hyperplasia non-malignant reticulum cells, differentiated lymphob'asts and lymphocytes.

SUMMARY

1) Among my series of 22 cases of lymphoid tumours of eyelids there were: 12 aleukaemic reticulosarcomata, 4 leukaemic infiltrations and 6 aleukaemic reactive lymphocytic hyperplasia infiltrations.

2) Lymphoid tumours of eyelids are usually accompanied by the same lymphoid infiltrations of the conjunctiva, orbit or lacrimal gland.

3) In eyelids reticulosarcomas occur at any age, lymphomas usually in adults and leukaemic infiltrations occur mostly in infants and are fatal by virtue of the leukaemic proccess.

REFERENCES

1. HERBUT P. A.: Pathology. London. Kimpton P. 948. 1959.
2. HOGAN M. J. and ZIMMERMAN. L. E.: Ophthalmic pathology Philadelphia. Saunders P. 765. 1962.
3. MORTADA A.: "Orbital reticulum-cell-sarcoma report of nine cases". Brit. J. Ophthal 45, 365, 1961.
4. MORTADA A.: "Orbital lymphoblastomas and leukaemias in children". Amer. J. Ophthal 55, 327, 1963.
5. MORTADA A.: "Reactive lymphocytic hyperplasia of orbit, lids, conjunctiva and lacrimal gland". Amer J. Ophthal. 56, 1963.
6. MORTADA A.: "Orbital, lacrimal gland, lid and conjunctiva lymphoid tumours cells smears". Orient. A. Ophth. 2, 60, 1964.

18A, 26 July Street