ORBITAL RETICULUM-CELL SARCOMA*

REPORT OF NINE CASES

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RETICULUM-CELL sarcomata, together with leukaemia, Hodgkin's disease, giant follicular lymphoblastoma (Brill Symmer's disease), lymphosarcoma, and lymphoma, constitutes a group of lymphomatous or reticular tumours. These different lymphoblastomata appear to represent pleomorphism of the reticular system to unknown causal stimuli (Ginsburg, 1934; Gall and Mallory, 1942; Herbut, Miller, and Erf, 1945). The nomenclature of these tumours in the literature is confused, and identical lesions have been described by various writers as lymphomata, lymphoblastomata, lymphocytomata, malignant lymphomata, lymphosarcomata, reticular-cell type of lymphosarcomata, reticulo-sarcomata, and round-cell sarcomata. Duke-Elder (1952) states that: "reticulosarcoma of the orbit is not commonly noted in the literature, but it is probable that several cases have been described as lymphosarcomata. Its occasional occurrence is, however, undoubted (Wilson, 1940; McGavic, 1943)."

Histological differentiation between lymphoid tumours and reticulum-cell tumours is often difficult, and both cell elements may participate in the formation of the same tumour (Pfanzen, 1957). The histological structure may change in the course of the disease when a previously benign tumour becomes malignant. However, the various types of lymphomatous tumours may be differentiated by their clinical behaviour and by the nature of the cells and intercellular fibres constituting the tumour.

Lymphomata are benign tumours composed of differentiated lymphocytes, but lymphosarcomata are highly invasive tumours. Their component cells are similar to lymphocytes, consisting of a mere rim of faintly stained cytoplasm and a relatively large nucleus with a dense chromatin network. The cells are closely and uniformly packed. Numerous mitotic figures are present.

Lymphomata and lymphosarcomata may both be associated with leukaemia. Giant follicular lymphadenopathy is essentially a lymphocytic hyperplasia and may arise from several different causes.

Reticulosarcoma cells are fairly large, and of varied shapes, round, polyhedral, oval, or irregular. Their cytoplasm is feebly stained, with a relatively

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large vesicular nucleus, a sharply defined nuclear membrane, and a delicate network of chromatin with granules at the intersections. Mitotic figures are present. The cells are not uniformly or closely packed. Many reticular argentophil fibres extend between the tumour cells, and may be seen clearly with Wilder's silver or Foot's stain.

As the orbit is free of lymphoid tissue, Axenfeld (1891) and Meller (1905) suggested that orbital lymphoblastomata were expressions of either leukaemia or Hodgkin's disease. Goldzieher (1907) and Coats (1915) ascribed their presence to pre-existing foetal lymphoid orbital remnants. It seems that they spread to the orbit from lymphatic elements in the lids, conjunctiva, lacrimal gland, or nasopharynx. About 5 per cent. of primary tumours of the lacrimal gland arise from its lymphoid tissue.

In sixteen cases of ocular lymphomatous disease, Heath (1949) found that 20 per cent. involved the lids and 20 per cent. the orbit. It is curious that Attiah (1933) reported 44 cases, Schreck (1939) 259 cases, Godfredsen (1947) 78 cases, Forrest (1949) 222 cases; Handousa (1951) sixteen cases, and Reese (1951) 335 cases of orbital tumour without any mention of reticulum-cell sarcoma. In 25 orbital tumours of the haemopoietic system Forrest (1949) found two cases of Hodgkin's disease and one plasmacytoma; the classification of the other 23 cases was difficult and included lymphomata and lymphosarcomata. Among 88 orbital tumours, Iliff (1957) found ten lymphosarcomata but no reticulosarcoma. Out of forty orbital tumours, Foster (1951) found three lymphomata. In 36 orbital lymphomatous tumours Reese (1951) found 25 lymphosarcomata, eight lymphomata, and three cases of Hodgkin's disease.

Reticulosarcoma of the orbit has been recorded at the extreme ages of 10 years (Kamel, 1948) and 83 years (Bürki, 1943).

McGavic (1955) found the survival time in eight patients with lymphocytic tumours to be 13 to 6 years, in three with giant follicular tumours 13 years, in eight with reticulum-cell lesions 7.2 years, and in one with Hodgkin's disease 4 years.

The attempted treatment of malignant lymphoblastomata is usually distressing. Most are radiosensitive (Crocì, 1934), but recurrence is usual (McGavic 1943). Radioactive phosphorus (P32) may occasionally prolong life (Lawrence, Dobson, Low-Beer, and Brown, 1948). Intravenous administration of nitrogen mustard may be useful when radio-resistance develops (Groen, Godfried, Kromsigt, Reisel, and Tillema, 1949; Chambers, 1950), and cortisone may have a temporary beneficial effect.

Case Reports

Case 1, a man aged 38 years, complained of oedema of the right upper eye lid and proptosis of 2 years' duration (Fig. 1, opposite).

The left eye was normal, with visual acuity 6/12.

The right eye showed a solid oedema of the upper lid and dilated conjunctival vessels. The fundus showed papilloedema. The visual acuity was 6/18. The eye was proptosed

FIG. 1.—Case 1, reticulo-sarcoma of right upper lid, lacrimal gland, and orbit of 2 years' duration in a man aged 38 years.

25 mm. downwards, with limitation of ocular movements in all directions. The lacrimal gland was palpable.

The general health was good, with no enlarged lymph glands, liver, or spleen. The blood Wasserman reaction and differential count were normal. An x-ray of the orbits and examination of the nose and nasopharynx did not reveal any abnormality.

The tumour was red in colour, 1.5×1 cm. in volume, and partly encapsulated. Histopathological examination showed it to be a reticulosarcoma of the orbit extending from the lacrimal gland (Fig. 2).

FIG. 2.—Case 1, microphotograph, showing reticulosarcoma cells involving the lacrimal gland and extending to the orbital tissues. $\times 690$.

Case 2, a girl aged 4 years, had left proptosis (Fig. 3) of one month's duration. The right eye was normal.

FIG. 3.—Case 2, left orbital reticulosarcoma with involvement of the lacrimal gland in a girl aged 4 years.

The left eye was proptosed 23 mm. downwards with limitation of ocular movements upwards. The fundus was normal. A tumour could be palpated in the upper part of the orbit.

The general health was good. The blood Wassermann reaction, blood count, x-ray of the orbits, and ear, nose, and throat examination were all normal.

The tumour lay between the lateral and superior rectus muscles extending anteriorly to involve the orbital part of the lacrimal gland. It was white, partly encapsulated, 2 × 3 cm. in volume, and soft in consistency.

Histopathological examination showed a reticulosarcoma (Fig. 4). The lacrimal gland was also found at biopsy to be infiltrated by reticulosarcoma cells.

FIG. 4.—Case 2, microphotograph, showing pleomorphism of cells, and presence of reticular fibres. × 690.

Case 3, a girl aged 5 years, had left proptosis (Fig. 5) of 4 months' duration.

FIG. 5.—Case 3, left orbital reticulosarcoma with involvement of the lacrimal gland in a girl aged 5 years.

The right eye was normal, the left being proptosed 28 mm. downwards with conjunctival chemosis and corneal ulceration due to lagophthalmos. A soft tumour occupied the upper part of the orbit. The blood Wassermann reaction, differential count, and ear, nose, and throat examination were normal. The general condition was poor, but there was no abdominal masses or enlargement of the lymph glands, liver, or spleen. X-ray examination of the left orbit showed destruction of the lesser wing of the sphenoid (Fig. 6, opposite), but the orbital cavity was not widened. An oblique skull x ray showed normal optic canals. One week later the proptosis was so severe that the eye was dislocated on to the cheek (Fig. 7, opposite).

FIG. 6.—Case 3, x ray of orbits, showing destruction of left lesser wing of sphenoid bone.

FIG. 7.—Case 3, in one week the proptosis increased so that the eye was dislocated on to the cheek.

The tumour, which was very soft, vascular, and friable, came away in pieces, and was found to involve the lacrimal gland. Histopathological examination showed a reticulo-sarcoma of the orbit extending from the lacrimal gland. Two weeks later the child died of cachexia.

Case 4, a girl aged 6 years, had a swelling of the left upper eyelid with proptosis of 2 months' duration. The right eye was normal. The left eye showed a firm lobulated swelling of the upper eyelid extending backwards in the upper part of the orbit. The globe was proptosed 24 mm. downwards. The fundus showed post-papilloedemic optic atrophy. The visual acuity was 1/60. The blood Wassermann reaction and differential count were normal. X-ray examination showed widening of the left orbit. One week later the proptosis was severer (Fig. 8), and biopsy showed reticulosarcoma of the orbit and lacrimal gland.

FIG. 8.—Case 4, showing rapid progress of orbital reticulosarcoma in one week.

Case 5, a man aged 64 years, complained of right proptosis (Fig. 9, opposite) of 3 years' duration. The left eye was normal, and the visual acuity 6/9. The right eye was proptosed 25 mm. downwards and outwards with limitation of ocular movements. The fundus showed post-papilloedemic optic atrophy. The visual acuity was 1/60. A soft mass was felt between the globe and the orbital roof. It was not attached to the skin and extended backwards in the orbit.

The general health was good. The blood Wassermann reaction and blood count were normal. X-ray examination of the orbits were normal.

The tumour, which involved the lacrimal gland, measured $3\frac{1}{2} \times 3 \times 2$ cm. and was pink, soft, and partly encapsulated. Histopathological examination showed it to be a reticulosarcoma of the orbit and lacrimal gland. Fig. 10 shows the patient after removal of the tumour and x-ray treatment.

FIG. 9.—Case 5, orbital reticulosarcoma with involvement of the lacrimal gland of 3 years' duration in a man aged 60 years.

FIG. 10.—Case 5, after removal of tumour and x-ray treatment.

Case 6, a man aged 28, complained of left proptosis of one month's duration. The right eye was normal, and the visual acuity 6/12.

The left eye showed proptosis 22 mm. with limitation of ocular movements, optic atrophy, and no perception of light. A soft mass was felt in the lower inner part of the orbit extending backwards. The patient refused treatment, but was seen one month later with severe proptosis simulating orbital cellulitis (Fig. 11).

FIG. 11.—Case 6, orbital reticulosarcoma simulating orbital cellulitis.

The general condition was bad; the blood Wassermann reaction and differential count were normal. An ear, nose, and throat examination showed a necrotic tumour in the left lateral nasal wall; the nasal septum was pushed to the right, there was a downwards bulging of the hard palate. A tumour was palpable at the left side of the nasopharynx roof. No metastasis were felt in the neck or elsewhere. An x-ray examination showed: widening of the left orbit; opacity of both maxillary antra with evidence of destruction of the left antrum; destruction of the anterior and posterior ethmoidal air cells; and invasion of the sphenoid sinus.

A nasopharyngeal mass was discovered (Fig. 12, opposite), and the pathological report was a reticulosarcoma.

Wilder's stain showed many reticulum argentophil fibres extending between the tumour cells.

FIG. 12.—Case 6, x ray showing nasopharyngeal mass.

Case 7, a Sudanese girl aged 16, complained of a right orbital mass felt through the right lower lid (Fig. 13) of 2 months' duration. The mass had been incompletely removed a month previously through a lower lid skin incision, and a painless proptosis had quickly developed.

FIG. 13.—Case 7, orbital reticulosarcoma, of one month's duration, extending from the lower conjunctival fornix in a girl aged 16 years.

The general health was good. The lymph glands, spleen, and liver were normal. The blood Wassermann reaction and the differential count were negative. The ear, nose, and throat examination and x rays of the orbits were normal.

The left eye was normal, with visual acuity 6/6.

The right eye was proptosed 22 mm. upwards with limitation of ocular movements in all directions. A soft mass was felt through the lower lid extending backwards in the orbit below the globe. The fundus showed a lower exudative detachment. The visual acuity was 6/12.

The tumour was removed by blunt finger dissection through a lower fornix conjunctival incision. It was irregular, soft, pink, not encapsulated, and measured $1 \times 4 \times 5$ cm. Histopathological examination showed a reticulosarcoma. Treatment was continued by radiation.

The patient returned 2 years later with a recurrence in the right orbit and the neck scar of a block dissection for malignant cervical lymph glands (Fig. 14, overleaf). The right eye showed +10 D hypermetropia, and the fundus showed posterior retinal striae. The visual acuity was 6/60.

FIG. 14.—Case 7, recurrence after 2 years. Note neck scar of block dissection.

A hard irregular mass was felt in the lower outer part of the orbit. There were no enlarged lymph glands. The blood count and *x* rays of the orbit were normal. After one month of radiation treatment, the hypermetropia and retinal striae disappeared, and the vision improved to 6/12, but a small hard mass could still be felt in the lower part of the right orbit.

Case 8, a woman aged 50 years, complained of left proptosis of one year's duration (Fig. 15). The right eye was normal, and the visual acuity 6/12.

The left eye showed oedema of the lids, proptosis 26 mm. upwards, and limitation of ocular movements. The fundus showed post-papilloedemic optic atrophy. The visual acuity was 1/60. A soft palpable mass was present in lower outer part of the orbit.

The general health was good. The blood Wassermann reaction and differential count were normal. A postero-anterior skull *x*-ray showed destruction of the left lateral orbital margin (Fig. 16) by the invading tumour. The tumour, which was not encapsulated, measured $3 \times 2 \times 2$ cm., pink and lobulated. The histopathological report showed it to be a reticulosarcoma.

FIG. 15.—Case 8, orbital reticulosarcoma of one year's duration extending from the lower conjunctival fornix in a woman aged 50 years.

FIG. 16.—Case 8, *x* ray showing destruction of left lateral orbital margin by invading tumour.

Fig. 17 shows the patient after removal of the tumour and x-ray treatment.

FIG. 17.—Case 8, after removal of tumour and x-ray treatment.

FIG. 18.—Case 9, orbital reticulosarcoma of one month's duration extending from lower conjunctival fornix in a man aged 30 years. There was generalized enlargement of the lymph glands and blood eosinophilia.

Case 9, a man aged 30 years, complained of loss of vision and right proptosis (Fig. 18) of one month's duration. He was emaciated and anaemic. The left eye was normal, and the visual acuity 6/9.

The right eye showed proptosis of 25 mm., chemosis of the conjunctiva, corneal ulcer, limitation of movement, and optic atrophy. There was no perception of light. A mass was felt between the globe and lower orbital margin near the lower conjunctival fornix.

The axillary and inguinal lymph glands were enlarged. X-ray examination of the orbits and the blood Wassermann reaction were normal; the differential count and bone marrow showed eosinophilia with no allergic or parasitic affection.

The tumour measured 3×1.5 cm.; it was irregular, partly encapsulated, soft, and whitish. An inguinal lymph gland was also removed. Histopathological examination of the tumour and the lymph gland showed reticulosarcoma.

Summary

Out of 100 orbital tumours removed surgically, nine proved on histopathological examination to be reticulosarcomata. They occurred at the ages of 4, 5, 6, 16, 28, 30, 38, 50, and 64 years, and the numbers of males and females were about equal. The patients, were all free from septic foci, allergic reactions, and parasitic diseases.

Reticulosarcoma of the orbit causes a painless proptosis of one month to 3 years' duration. The less the duration the softer, more vascular, and more malignant is the tumour. In the nine cases described there was a palpable orbital mass. The clinical details are analysed in the Table.

Orbital reticulo-sarcomata (characterized histopathologically by large pleomorphic cells with pale cytoplasm, vesicular nuclei, and argentophil reticular fibres), though rarely recorded in the literature, are the commonest malignant tumours of the orbit in Egypt. The orbit is free of lymphoid tissue, and the cases described showed that orbital reticulosarcomata arose from the lymphoid tissue of the lacrimal gland in four cases, from the conjunctiva in three cases, lids one case, and nasopharynx one case.

Many of the cases described in the literature as orbital lymphomata, lymphosarcomata, and round-cell sarcomata are probably reticulosarcomata.

TABLE
CLINICAL DETAILS IN NINE

Case No.	Age (yrs)	Sex	Duration (mths)	Side	Degree of Proptosis
1 (Fig. 1)	38	Male	24	R	25 mm. downwards
2 (Fig. 3)	4	Female	1	L	23 mm. downwards
3 (Fig. 5) (Fig. 7)	5	Female	4	L	28 mm. downwards
4 (Fig. 8)	6	Female	2	L	24 mm. downwards
5 (Fig. 9) (Fig. 10)	64	Male	36	R	25 mm. down and outwards
6 (Fig. 11)	28	Male	1	L	22 mm.
7 (Fig. 13)	16	Female	2	R	22 mm. upwards
			2 years later	R	—
8 (Fig. 15) (Fig. 17 after operation)	50	Female	12	L	26 mm. upwards
9 (Fig. 18)	30	Male	1	R	25 mm.

CASES OF RETICULUM-CELL SARCOMA

Extent of Orbital Tumour	Lymph Glands	Blood	Condition of Eye	Tumour (cm.)
Into orbit from lacrimal gland, lid and conjunctiva	Normal	Normal	Papilloedema 6/18	Red 1.5 × 1 Partly encapsulated (Fig. 2)
To external and superior rectus from lacrimal gland	Normal	Normal	Normal fundus	2 × 3 White, partly encapsulated, soft (Fig. 4)
Upper part of orbit from lacrimal gland	Normal	Normal	Corneal ulceration	Soft, vascular, friable (Fig. 6)
From upper eyelid and lacrimal gland into orbit	Normal	Normal	Optic atrophy 1/60	—
Soft mass between globe and orbital roof from lacrimal gland	Normal	Normal	Optic atrophy 1/60	3.5 × 3 × 2 Pink, soft, and partly encapsulated
Soft mass behind inner part of orbit from nasopharynx	Normal	Normal	Optic atrophy No P.L.	Nasopharyngeal necrotic (Fig. 12, x ray)
Soft mass from conjunctiva backwards into lower part of orbit	Normal	Normal	6/12 Lower exudative retinal detachment	1 × 4 × 5 Soft, pink, not encapsulated
Hard mass in lower outer region of orbit	Malignant lymph glands dissected (Fig. 14)	Normal	6/60 Retinal striae Hypermetropia	—
Soft mass in lower outer region of orbit from conjunctiva	Normal	Normal	Optic atrophy 1/60	3 × 2 × 2 Pink lobulate, not encapsulated (Fig. 16, x ray)
Mass between globe and lower orbital margin from conjunctiva	Enlarged	Eosinophilia	Corneal ulcer Optic atrophy No P.L.	3 × 1.5 Soft, whitish, irregular, partly encapsulated

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