CYSTIC LYMPHANGIOMA OF THE ORBIT*

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CYSTIC lymphangiomata are occasionally found in the neck, axilla, groin, sacral region, floor of the mouth, and retro-peritoneal space. A few cases have been described in the orbit by Gradle (1920), Weldige-Cremer (1920), Niosi (1921), and Meisner (1926). Among 120 primary intra-orbital tumours diagnosed histologically, I have seen three cystic lymphangiomata. The rarity of these lesions and the varying presenting clinical features make these three cases of general interest.

Case Reports

Case 1, a 17-year-old girl (Fig. 1), complained of gradually developing painless proptosis and swelling of the left upper eye lid for the past year. She had had no pregnancies.

The right eye was normal, with a normal fundus, and a visual acuity of 6/9.

FIG. 1.—Case 1, orbital cystic lymphangioma in a girl aged 17, showing proptosis and swelling of upper lid.

FIG. 2.—Case 1, translucent orbital cyst pushing forward the medial canthus conjunctiva.

The left upper lid was swollen, especially near the root of the nose, with ptosis, the caruncle and conjunctiva of the medial canthus being pushed forwards by a glistening translucent orbital cystic swelling (Fig. 2). There was a proptosis of 21 mm. (right 15 mm.); and limitation of ocular movements inwards. Otherwise the eye and fundus were normal; the visual acuity was 6/12.

There were no other apparent or palpable swellings or enlarged lymph glands in the body. The general and rhinological examinations showed no abnormality. The blood count was normal. The Wassermann reaction and tuberculin tests were negative.

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An x ray of the orbits showed nothing abnormal, but orbital exploration revealed a large cyst containing clear fluid in the medial side of the orbit, outside the muscle cone.

A non-encapsulated translucent tumour, measuring 0.5×0.5 cm., was excised from the conjunctival surface of the upper lid.

Histological Examination.—The large orbital cyst was lined by flat endothelial cells, and was in fact a cystic lymphangioma, with numerous Meibomian glands, accessory lacrimal glands, and cavernous lymphangiomatous spaces (Fig. 3).

FIG. 3.—Case 1, tumour removed from palpebral conjunctiva, showing Meibomian glands in tarsus, accessory lacrimal glands, and cavernous lymphangiomatous spaces. $\times 120$.

Case 2, a male infant aged 18 months, had right proptosis and a cystic orbital swelling which had rapidly pushed the right upper lid forwards in 2 days (Fig. 4).

FIG. 4.—Case 2, orbital cystic lymphangioma in a male infant aged 18 months, showing the orbital cyst pushing the upper eyelid forwards.

There was no history of trauma. The left eye was normal, with a normal fundus.

The right lower lid was normal, but the upper lid showed ecchymosis, ptosis, and a large non-pulsating cystic swelling, partially compressible and not attached to the skin, which prevented the examination of the right eye.

The child's general condition was good. There were no other apparent or palpable swellings in the body, and no allergic manifestations. The Wassermann reaction and tuberculin tests were negative. The blood count and faeces and urine examinations were normal. The clotting time was 2.5 min. and the bleeding time 2 min. A postero-anterior x ray of the skull showed a wider right orbit (Fig. 5, opposite).

After the aspiration of dark blood, the cyst diminished in size, and the eye could be examined. There was a right proptosis of 20 mm. (left 15 mm.) and limitation of ocular movements upwards. Otherwise the right eye was normal with a normal fundus.

FIG. 5.—Case 2, Postero-anterior skull x ray, showing wider right orbit.

Orbital exploration revealed a large cyst, 1.5×2 cm. in size, which was full of blood and occupied the upper part of the orbit, above the globe. The medial part of the upper lid showed a small yellowish non-encapsulated tumour, measuring 0.5×1 cm., under the palpebral and fornix conjunctivae.

Histological Examination.—The conjunctival tumour showed clear spaces lined by endothelial cells separated by connective tissue trabeculae, surrounding Krause's glands (Fig. 6).

FIG. 6.—Case 2, tumour removed from palpebral conjunctiva, showing cavernous lymphangiomatous spaces around Krause's glands. $\times 120$.

Some spaces were full of lymphocytes. The fibrous trabeculae between the spaces showed areas of chronic inflammation rich in eosinophils.

The large orbital cyst was lined by flat endothelial cells and contained blood.

The picture is consistent with that of a cavernous lymphangioma of the lid and conjunctiva, with haemorrhage into a cystic lymphangioma of the orbit.

Case 3, a 22-year-old woman (Fig. 7, overleaf), complained of gradual painless proptosis and diminution of vision in the right eye of 7 months' duration. She had one healthy child and no other pregnancies. The left eye had been shrunken since childhood.

The right eyelids were externally normal and the visual acuity was $1/60$. A small translucent mass measuring 0.5×0.5 cm. was present in the lower conjunctival fornix.

The fundus showed post-papilloedemic optic atrophy. There was a proptosis of 21 mm. and a slight limitation of ocular movements downwards. A deep orbital mass was felt between the globe and the lower orbital margin.

FIG. 7.—Case 3, orbital cystic lymphangioma in a woman aged 22, showing proptosis.

The patient's general condition was good. There were no other apparent or palpable swellings in the body. The blood count was normal. The Wassermann and tuberculin tests were negative. X rays of both orbits showed no abnormality.

The lower conjunctival fornix tumour was removed. The orbital mass proved to be a translucent cyst in the muscle cone space which contained clear fluid and measured 1×1.5 cm.

Histological Examination.—The fornix conjunctival mass was a cavernous lymphangioma (Fig. 8).

FIG. 8.—Case 3, tumour removed from lower conjunctival fornix, showing cavernous lymphangiomatous spaces. $\times 120$.

The orbital cyst was a cystic lymphangioma lined by flat endothelial cells.

Discussion

Few orbital cystic lymphangiomata have been recorded in the literature. Even orbital cavernous lymphangiomata are rare—only some forty cases have been reported since the original observations of Forster (1886) and Wiesner (1886). As the orbit is free of lymphatic vessels, orbital lymphangiomata originate either from displaced foetal cells of the lymphatic channels (Watson and McCarthy, 1940) or from lymphangiomata extending from the

lid and conjunctiva (Werncke, 1904). The tumour has usually a congenital basis and proptosis may be evident at birth (Cabannes, 1903), but it usually grows so very slowly that advice is not sought until the patient has reached adult life (Ayres, 1895). Orbital lymphangiomas may be associated with lymphangioma of cheek (Israel, 1895), or lips and palate (Waldstein, 1910), or with hypertrophy of the face (Jess, 1936).

Wintersteiner (1898), Franklin and Cordes (1924), and Wolff (1932) observed that recurrent attacks of inflammation or haemorrhage in cases of orbital lymphangiomas manifest clinically as intermittent proptosis. Visual damage due to pressure on the optic nerve by an orbital cavernous lymphangioma was noted by Kahn (1906) and Smith (1925).

Orbital cystic lymphangiomas have to be differentiated from other orbital cysts, especially the serous cysts classically described as hygromata, which are usually instances of serous tendonitis or dilatation of fascial elements associated with the muscle tendons (Duke-Elder, 1952).

The safest treatment for cystic orbital lymphangioma is by surgical removal. Any cavernous lymphangioma can be treated by electrolysis, 75 per cent alcohol injection (Wray, 1915), or radiation (Mackay, 1915), but the best treatment is by excision.

Summary

Three cases of cystic orbital lymphangiomas were found among 120 histopathologically diagnosed primary orbital tumours. All three cases showed small cavernous lymphangiomas on the conjunctival surface of the eye lids. As the orbit is free from lymphatic vessels, the most probable explanation of the presence of an orbital cystic lymphangioma is that it comprises an extension of dilated lymphangiomatous spaces from the lid conjunctival tumour.

The presenting clinical feature differed in the three cases:

- (a) Gradual proptosis and swelling of the upper lid.
- (b) Rapid proptosis and cystic swelling of the upper lid after haemorrhage in the tumour. The condition simulated an orbital haemorrhage after trauma. Histologically the tumour trabeculae showed chronic inflammatory cells rich in eosinophils.
- (c) Gradual slight proptosis and marked diminution of vision due to the pressure of the tumour on the optic nerve causing post-papilloedemic optic atrophy.

The three cases were all treated successfully by excision of both the cystic orbital lymphangioma and the cavernous lid conjunctival lymphangioma. These are the first three orbital cystic lymphangiomas to be reported from Egypt.

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