ORBITAL BENIGN HAEMANGIO-ENDOTHELIOMATA*

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ORBITAL HAEMANGIO-ENDOTHELIOMATA, tumours of angioblastic endothelium, are very rare. Birch-Hirschfeld (1930) and Peters (1931) questioned their occurrence, but they have been described by Adson and Benedict (1934), Röttgen (1950), and other authors. In a series of 222 intra-orbital tumours, Forrest (1949) found one angio-endothelioma, and Iliff (1957) found three cases in a series of 88 primary orbital tumours. Out of 44 angiomas, Reese (1951) described six angioblastic haemangiomas and one haemangio-endothelioma.

Case Reports

Case 1, a boy aged 8 months, had shown left proptosis since childhood (Fig. 1).

FIG. 1.—Case 1. Left proptosis due to a benign haemangio-endothelioma in a male child aged 8 months.

Examination.—The right eye was normal. The left eye showed slight proptosis and a normal fundus. A soft mass was present in the medial part of the upper lid extending to the medial part of the orbit giving limitation of ocular movements inwards. General examination revealed no abnormality. There were no enlarged lymph glands or abdominal mass. X ray of both orbits was normal and the blood Wassermann reaction was negative. The blood count showed: red blood corpuscles 5,200,000, white blood corpuscles 7,100, basophils 0 per cent., eosinophils 4 per cent., staff nucleated 6 per cent., segmented 43 per cent. (total polymorphs 49 per cent.), lymphocytes 42 per cent., and monocytes 5 per cent.

Operation.—When the orbital mass was excised, it was found to be soft, red, and not encapsulated.

Histopathological examination showed clefts (Fig. 2, opposite) and neoplastic capillary vessels surrounded by proliferating endothelial cells. In some areas there was dense endothelial cell proliferation. Wilder reticulin stain showed endothelial cell proliferation inside the capillary lumina (Fig. 3, opposite).

Result.—The child has been seen regularly for the last 2 years; there has been no orbital or lid tumour recurrence, no glandular enlargement, and no distant metastasis.

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FIG. 2.—Case 1. Proliferating endothelial cells surrounding clefts and blood spaces. $\times 120$.

FIG. 3.—Case 1. Endothelial cell proliferation inside neoplastic capillaries. Wilder reticulin stain. $\times 540$.

Case 2, a 35-year-old woman, complained of left proptosis of 17 years' duration which had increased during the last 2 years (Fig. 4).

FIG. 4.—Case 2. Left proptosis due to a benign haemangio-endothelioma in a woman aged 35 years.

Examination.—The right eye was normal, with normal fundus, and visual acuity 6/9. The left eye showed forward and upward proptosis of 30 mm. Hertel (right side 16 mm.) and limitation of ocular movements in all directions. The fundus showed papilloedema. The visual acuity was 6/36 but with no error of refraction. A hard mass was felt between the lower orbital margin and the eye. General examination revealed no abnormality. There were no enlarged lymph glands or distant metastasis.

The blood Wassermann reaction and tuberculin tests were negative. The blood count showed haemoglobin 95 per cent., red blood corpuscles 4,880,000, white blood corpuscles 5,800, basophils 0 per cent., eosinophils 2 per cent., staff nucleated 6 per cent.,

segmented 54 per cent. (total polymorphs 60 per cent.), lymphocytes 34 per cent., monocytes 4 per cent.

A postero-anterior x ray of the skull (Fig. 5) showed a dilated left orbit. Oblique x rays showed normal optic canals.

FIG. 5.—Case 2. Postero-anterior x-ray of skull, showing wide left orbit.

FIG. 6.—Case 2. Tumour removed.

Operation.—Through a lower fornix conjunctival incision the tumour was found below and to the inner side of the eye. It was not attached to the optic nerve and was removed easily by blunt little-finger dissection. The tumour was encapsulated and measured 4×2 cm. (Fig. 6); it was soft, nodular, and pinkish grey in colour.

Histopathological examination showed an encapsulated tumour with a concentric arrangement of proliferating endothelial cells mostly around clefts and blood spaces (Fig. 7, opposite) and around neoplastic capillary vessels (Fig. 8, opposite). Endothelial cell metaplasia to fibroblasts was seen. In some areas the tumour was cellular.

Result.—The patient was seen 3 years after the operation. Her general condition was good. There was no orbital tumour recurrence, no glandular involvement, and no distant metastasis.

Discussion

Angioblastic haemangioma, as described by Reese (1951), is a tumour of primitive and anaplastic embryonic endothelial cells before the development of true vascular channels. These tumours usually occur in infants. They may appear locally invasive but show no metastasis.

Stout (1943) suggested the term haemangio-endothelioma to designate a tumour characterized by a proliferation of endothelial cells in the vessel lumen outlined by the reticulum sheath. Stout and Murray (1942) applied the term haemangio-pericytoma to tumours characterized by proliferation of the contractile pericytes outside the reticulum sheath of the capillary wall. By haematoxylin and eosin stain it is impossible to distinguish pericytes from endothelial cells. It is necessary to impregnate the reticulin network with silver to determine whether the cells are outside or inside the capillary network. Both types of tumour occur in any sex and at any age. They usually have a benign course, but a malignant orbital haemangio-endothelioma (angio-sarcoma) was described by Forrest (1949).

FIG. 7.—Case 2. Concentric arrangement of proliferating endothelial cells surrounding clefts and blood spaces. $\times 120$.

FIG. 8.—Case 2. Proliferating endothelial cells surrounding neoplastic capillaries. $\times 540$.

In young infants, as in our first case, benign haemangio-endotheliomata may appear invasive and cellular, and yet behave clinically as benign tumours (Hogan and Zimmerman, 1962). The two orbital haemangio-endotheliomata described above, though showing dense endothelial cell proliferation in some areas, proved to be clinically benign, as were two other orbital haemangio-endotheliomata previously described (Mortada, 1961). Careful post-operative observation of these four cases for several years supports the view that most orbital haemangio-endotheliomata are benign in nature.

Summary

(1) Out of 130 primary orbital tumours seen personally, there have been only four haemangio-endotheliomata. Two of them were encapsulated (in women aged 30 and 35 years), and the other two were non-encapsulated (in male infants aged 5 and 8 months). The four cases support the view that most orbital haemangio-endotheliomata are benign in nature.

(2) These are the first four cases of orbital benign haemangio-endotheliomata to be reported from Egypt.

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