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Intracranial optic nerve angioblastoma

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Angiomatous tumours causing visual disturbances are well known (Reese, 1963). The tumour may be isolated (mainly in the orbit and choroid), or it may be a manifestation of von Hippel's disease (in the retina and on the optic disc), Sturge-Weber's disease (in the choroid and intracranial), or Coats's disease (in the retina). Angiomatous tumours of the optic nerve are extremely rare. This communication presents an intracranial optic nerve angioblastoma found by chance at necropsy 23 years after the onset of visual disturbances which led to unilateral optic atrophy.

Case report

A 43-year-old man with a history of bronchial asthma, maxillary sinusitis, and osteomyelitis of the femoral bone entered the University Eye Clinic in 1949 for investigation of a swollen right optic disc associated with decreasing visual acuity. Headaches and a feeling of pressure in the right eye had been his main complaints in the last 6 months before admission. The visual acuity of this eye had never been as good as that of the left.

Ocular examination

Visual acuity was 5/35 in the right eye and 5/7·5 in the left. Ophthalmoscopic examination of the right eye showed a swollen disc with an elevation of about 2 D. The visual field in this eye revealed a concentric narrowing without a central scotoma or an enlarged blind spot. The other eye was in all respects within normal limits.

General examination

The patient had a dental cyst and bronchitis. There were no signs of maxillary sinusitis.

Laboratory data

The cerebrospinal fluid was normal. A cerebral angiogram was not performed at that time.

Diagnosis

Suspected retrobulbar neuritis.

Clinical course

For the next few months there were no changes in the ophthalmoscopic or visual field findings, but relatives later reported that the patient had gone blind in the right eye.

Termination

In 1972, 23 years after the onset of the visual disturbances, the patient died in cardiac failure. During his last illness an ophthalmoscopic examination showed total atrophy of the right optic nerve while the left eye was normal.

Necropsy*

A tumour of the right optic nerve was noted about 4 mm. in front of the optic chiasm measuring 11 × 9 mm. There was marked optic atrophy of the right optic nerve. The surface of the tumour showed tortuous blood vessels (Fig. 1, overleaf). No other tumour was found. The right eye showed

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* This was done at the Institute of Pathology, Stadt. Krankenhaus, Munchen-Harlaching, from where the brain and globe were kindly sent to us for examination.

no further pathological changes. The main diagnosis was emphysematous changes of the lungs and hypertrophy of the heart.

Neuropathological examination (E.R.) of the brain revealed no gross changes except for the optic nerve tumour.

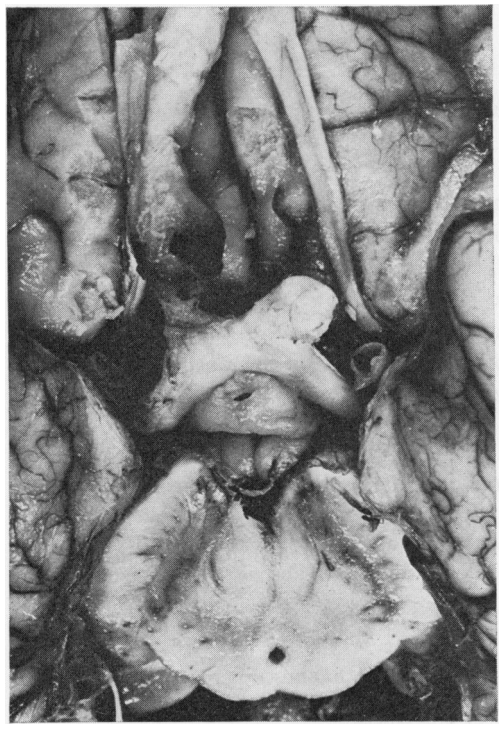

FIG. 1 *Base of the brain, showing tumour in right optic nerve*

Microscopical appearance of the tumour (SN 345/72)

Histologically the tumour appeared unencapsulated and to be located mainly within the optic nerve; it occupied almost the whole diameter of the nerve, without extending into the thickened leptomeninges. There was total atrophy of the right optic nerve (Figs 2 and 3). The highly vascularized tumour contained clusters of endothelial cells forming small spaces (Fig. 4). Silver stains revealed

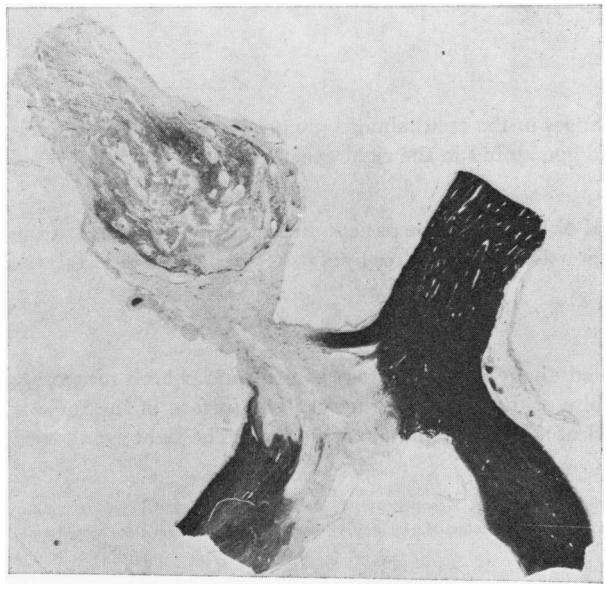

FIG. 2 *Section of optic chiasm (Woelcke myelin stain), showing total demyelination of right optic nerve*

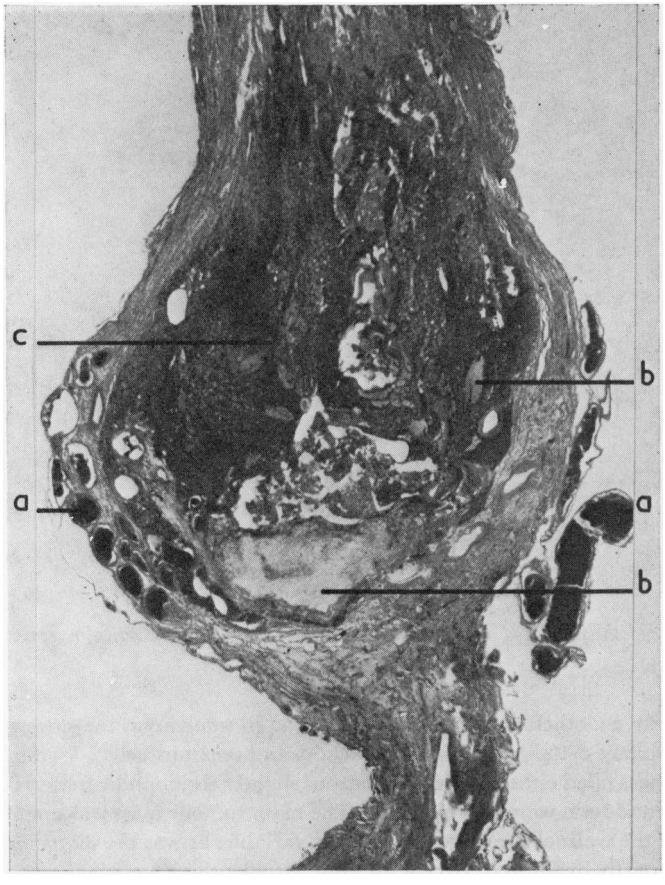

FIG. 3 *Low-power view of tumour (Goldner stain), showing enlarged vessels on surface (a), cysts (b), and cell-rich main tumour mass (c)*

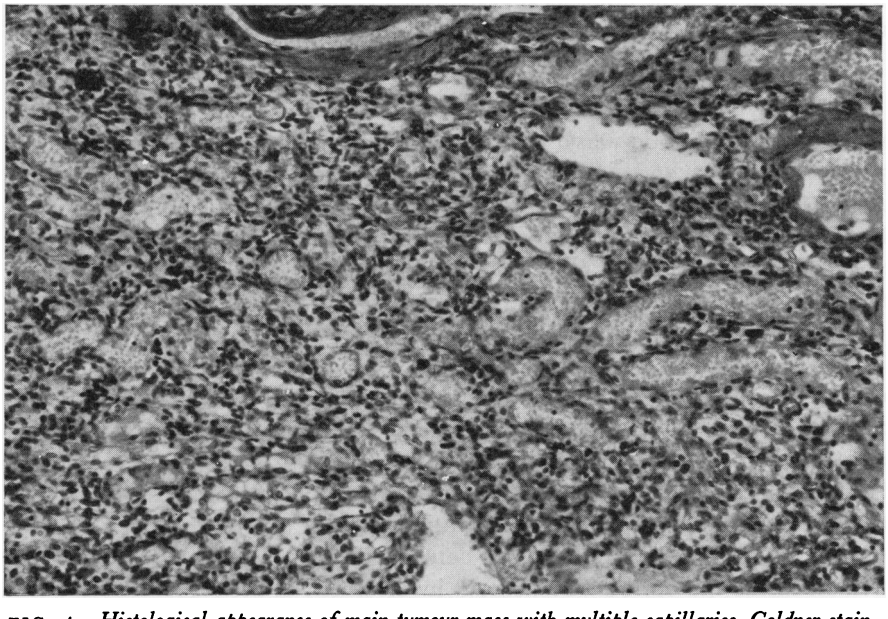

FIG. 4 *Histological appearance of main tumour mass with multiple capillaries. Goldner stain.* × 130

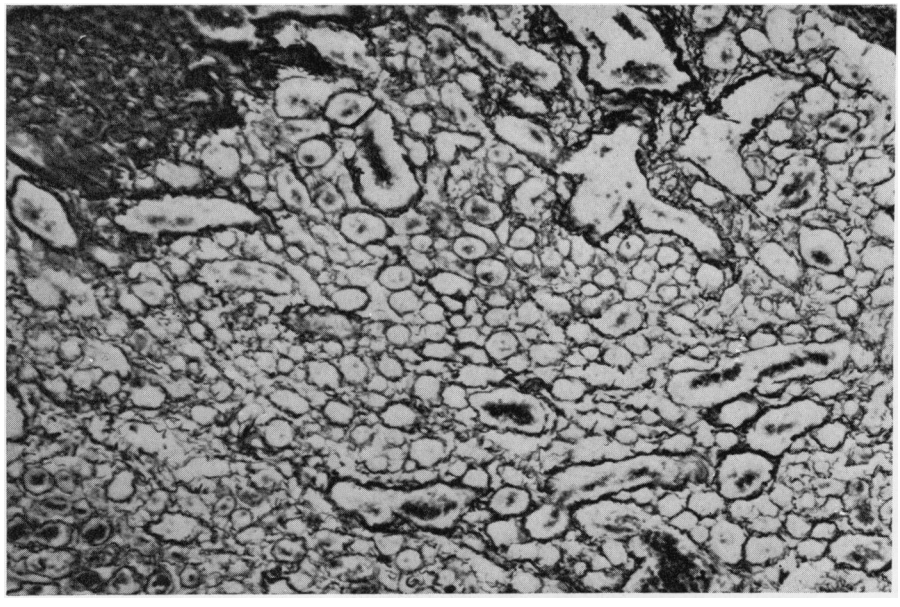

FIG. 5 *Reticulum framework around endothelium-lined blood-filled spaces. Kelemen* (1971) *reticulin stain.* × 130

a framework of reticulum around these endothelium-lined spaces (Fig. 5). In some areas there were large swollen cells with vacuolated foamy cytoplasm (so-called pseudoxanthomatous cells). Within the tumour there were some cystic spaces filled either with homogeneous slightly eosinophilic material or with blood. Small amounts of haemosiderin were also observed. The main tumour mass was made up of capillaries. The architecture of the walls of the larger vessels was variable, as was the diameter of the vessels. Some vessel walls were partly hyalinized. Except for optic atrophy there were no pathological changes in the right eye. The histological examination of the brain revealed trans-synaptic atrophy in both lateral geniculate bodies in the corresponding cell layers.

Discussion

Angiomatous tumours of the optic nerve are most frequently described in the optic disc (Vogel, 1965; Darr, Hughes, and McNair, 1966; Walsh and Hoyt, 1969; Duke-Elder, 1971), and are usually regarded as manifestation of von Hippel's disease. We have found one report of an intraorbital optic nerve angioma (Schneider, 1942) and one of an intracranial optic nerve angioma (Verga, 1930). Both were attributed to von Hippel-Lindau disease. In fact the case described by Verga (1930) is almost identical with ours. He too observed by chance at necropsy an angiomatous tumour about the same size as that in our case and also situated in the prechiasmal portion of the right optic nerve in a patient aged 57 years. According to Roussy and Oberling (1930), Verga called the tumour an angioreticuloma. Reese (1963) classified Verga's case as a capillary angioma. Our observation also suggests a capillary angioma or, according to Stout (1943), Zülch (1956), and Bailey and Ford (1942), a haemangio-endothelioma or angioblastoma. The clusters of so-called pseudoxanthomatous cells and the observation of cystic spaces within the tumour as seen in Lindau's tumour suggest a relationship between these tumours. The main growth of the tumour mass inside the optic nerve without involvement of the leptomeninges and and the absence of a capsule around the tumour (Corradini and Browder, 1948; Zülch,

1956) seem to rule out the possibility of an angioblastic meningioma which is also known to occur in the optic nerve.

Summary

A small prechiasmal angiomatous tumour of the right optic nerve was found by chance at necropsy. The tumour had caused decreasing visual acuity, narrowing of the visual field, and swelling of the optic disc 23 years before, with slowly progressive optic atrophy. Histological examination revealed an angioblastoma showing the features of Lindau's tumour.

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