Cerebral Hemangioblastomas
Review of Literature and Report of Two Personal Cases

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Cerebellar hemangioblastomas, not a rare occurrence in neurosurgical practice, are now well known in their anatomicopathological aspects and, because of their peculiar angiographic appearance, often can be identified preoperatively by vertebralography. Very little is known, on the other hand, about tumors of the same kind occurring elsewhere in the central nervous system, viz. cerebral and spinal hemangioblastomas, since there have been extremely few proven cases. Cushing and Bailey\(^5\) wrote in 1928 that most of the cases published up to that date as cerebral hemangioblastomas had been labelled erroneously or substantiated insufficiently and that only 4 of them (those of Borchers,\(^4\) Bielschowsky,\(^3\) Berger and Guleke,\(^2\) Schley\(^17\) could more or less withstand criticism. In 1956 Zülch\(^20\) rejected even these and regarded as proven only 2, those reported by Rochat\(^15\) and by Kautzky and Vierdt\(^10\) since Cushing and Bailey’s monograph.

We were therefore sceptical of the 2 cases of supratentorial hemangioblastomas that we had encountered in the past few years and doubted the histological diagnosis very much. However, repeated examination of the slides and postoperative rechecking of the patients led us to the conclusion that we were on firm ground. Moreover, as there are some published cases that Zülch\(^20\) apparently had not noticed, we felt that the whole subject deserved review. A point of particular interest in this connexion is that carotidography was performed in our cases and in some of the previous cases and the angiographic features are, in our view, worth considering as a possible aid to preoperative diagnosis.

All the cases published to date, whether in our opinion acceptable or not, are summarised here in some detail whilst our cases are reported fully.

Our 2 cases were found in a series of 1,483 primary supratentorial growths verified at operation at the Neurological Institute of Milan over a period of 16 years (September 1946–August 1962). In the same period of time 34 cerebellar hemangioblastomas were operated upon.

Case Reports

Case 1. Serial No. 4509B (1958). G.D., a boy aged 10, 1 month before admission to the Neurological Institute had a sudden attack of frontal headache and vomiting immediately followed by coma. Lumbar puncture, performed at another hospital, had yielded bloody fluid. On regaining consciousness some hours later, the patient displayed right hemiparesis and left ophthalmoplegia, which did not clear up completely.

Examination. Right pupil was larger than left. There was palsy of the left abducens with limitation of upward and downward rotation of the left bulb. Visual acuity was 10/10 for both eyes; right homonymous hemianopia and mild blurring of optic disks were present. He had right hemiparesis with hyperreflexia and Babinski’s sign.

Electroencephalogram showed slow arrhythmic monophasic waves (1–2 c./sec.) in the left temporo-occipital region. Left carotidography: in the posterior temporal region, both in arterial and venous phases, there was a roundish, pea-sized, densely contrasted nodule, fed by a branch of the middle cerebral artery and surrounded partially by a small tangle of pathological vessels. Normal vessels surrounding the nodule were displaced peripherally around it (Figs. 1 and 2).

Preoperative Diagnosis. Vascular tumor, possibly hemangioblastoma with surrounding hematoma.

Operation (Oct. 9, 1958). Under intratracheal anesthesia with N\(_2\)O and ether, a left temporo-occipital flap was made. In the lower part of the exposed region, immediately underlying the cortex, there was a tangerine-sized collection of darkish blood clots. After emptying, in the wall of the resulting cavity a round, reddish nodule, about 7 mm. in diameter, fed by two small arteries

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(1–1 1/2 mm. in diameter) was observed and removed completely without difficulty.

Postoperative course was uneventful.

Microscopical Examination. The neoplastic tissue was clearly demarcated by the surrounding cerebral parenchyma and was composed uniformly of vesicular cells with very clear cytoplasm and not very intensely stained central nucleus. The tissue was crossed by blood vessels, in the main very fine and composed of endothelial lining only (Figs. 3 and 4). Fine fibres of reticulin surrounding the blood vessels and spreading through the intervening tissue were stained by the silver-carbonate method (Fig. 5).

Subsequent Course. At follow-up in May 1962 (3 1/2 years after operation), the patient was in good health. There were no abnormal findings excepting right inferior homonymous quadrantanopsia.

Case 2. Serial No. 29742 (1960). D.M.R., a man aged 27, had a 10-year history of episodic dazzling fits in the left eye accompanied by palpitation of the heart and psychic anguish, twice followed by brief loss of consciousness. He had transient bouts of pain in right fronto-orbital region for 3 years; transient blurring for 4 months and finally permanent impairment of vision.

Examination. Positive neurological findings were: mild left motor impairment; deviation to the left when walking with eyes closed; nystagmus