Case Reports

Cerebral Hemangioblastoma Occurring in a Patient with von Hippel-Lindau Disease*

Case Report

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Since Pyc-Smith’s first report in 1885,14 a variety of pathological entities commonly found in patients with cerebellar hemangioblastomas have been described. Spinal cord hemangioblastoma, angiomatosis retinae, polycystic kidneys, pancreatic cysts, hypernephroma, and erythrocytosis are among the associated lesions reported.5–7,11,17,21 The retinal lesions were described by von Hippel in 1904.9 Later Lindau described the syndrome which included retinal and cerebellar neoplasms and noted the frequent occurrence of the disease in families.12 Subsequently, the familial pattern of the syndrome was confirmed by others.

The occurrence of cerebral hemangioblastoma in a patient with von Hippel-Lindau disease is very rare. In 1931, Rochat described a cerebral hemangioblastoma un-associated with a cyst occurring in a 20-year-old man with multiple cerebellar hemangioblastomas and a family history of angiomatosis.17 Since then, occasional angioblastic tumors arising in the cerebrum have been documented, but none associated with angiomas elsewhere in the body.2–4,10,16,20

We are reporting a case in which a cerebral hemangioblastoma occurred in a patient with von Hippel-Lindau disease.

Case Report

A 24-year-old man had an asymptomatic retinal hemangioma adjacent to the right optic disc; it was first noted in September, 1956. Right occipital headache began in February, 1958, associated with pain in the right eye, nausea, and vomiting.

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First Admission. The patient was admitted to the New York Hospital-Cornell Medical Center in February, 1958, where he was found to have bilateral papilledema, a hemangioma of the right retina, and bilateral horizontal nystagmus. The hematocrit was 55%, hemoglobin 19.0 gm%, and a red blood cell count of 7.1 million per cu mm. Vertebral arteriography demonstrated a poorly localized cerebellar mass.

First Operation. At craniotomy, a left cerebellar hemangioblastoma in the wall of a cyst was excised. The hemogram was normal 3 weeks after operation. Except for a gradually enlarging scotoma of the right eye, the patient remained well for 3½ years, when occipital headache, nausea, and vomiting recurred, associated with ataxia of the right arm.

Second Admission. In September, 1961, examination showed a large right retinal hemangioma, dysarthria, and dysmetria of the right arm. The hematocrit was 49%, hemoglobin 16.5 gm%, and the red blood cell count 6.0 million per cu mm. Ventriculography demonstrated a block and anterior dislocation of the aqueduct with moderate dilatation of the ventricular system.

Second Operation. At craniotomy, a right cerebellar hemangioblastoma and cyst were identified and the tumor excised. Exploration of the left cerebellar fossa showed no evidence of recurrent tumor. The hemogram returned to normal thereafter. The patient remained well for 5 years except for mild residual ataxia of the right arm and a right visual scotoma; then he again developed nausea, vomiting, and occipital headache.

Third Admission. In July, 1966, examination showed a tense suboccipital decompression, blindness on the right due to a completely detached retina, a superior nasal defect in the
bellar lesion and a separate left parietal lesion (Fig. 1).

Third Operation. On exploration of the posterior fossa, a new left cerebellar neoplasm in the wall of a cyst was found and excised. The lesion was remote from the site of the left cerebellar hemangioblastoma excised 10 years previously. Microscopic sections of the tumor revealed abundant capillaries supported by a delicate reticulin stroma in which occasional "pseudoxanthoma cells" could be seen. The histopathologic diagnosis was cerebellar hemangioblastoma (Fig. 2 left).

Fourth Operation. A left occipital craniotomy was performed 4 weeks later to remove a large left occipitoparietal neoplasm. There was no evident attachment of the cerebral neoplasm to the meninges or to the previous tumor sites. A 4 cm cyst was found adjacent to the tumor, and the tumor itself had a small cystic area within it. The histopathologic appearance of the cerebral lesion was strikingly similar to that of the resected cerebellar hemangioblastoma (Fig. 2 right).

The postoperative course was uneventful although there was a partial expressive aphasia which was steadily improving upon the patient's discharge from the hospital.

Family History. Of 26 family members, four males and one female have had angiomasis. Cerebellar hemangioblastomas occurred in all four men. Three of the men had multiple cerebellar tumors. Our patient was the only member of the family with angiomasis retinae, associated cerebellar hemangioblastomas, erythrocytosis, and a solitary cerebral hemangioblastoma. Hypernephroma occurred in two of the men, one of whom had metastases to the other kidney and to the lungs. Polycystic kidneys were found in the same two men as were cysts in the pancreas of each. A port wine stain of the neck is the only known sign of angiomasis in the one female.

Comment

Cerebellar hemangioblastoma, while infrequently encountered, is not rare. From individual and collective experience, it is recognized that these lesions may be multiple in the cerebellum.15,18,20

Cerebral hemangioblastoma is decidedly rare, and the diagnosis is frequently disputed. Luschka's first report of a case in 1854 was later discounted by Virchow.19 Reports thereafter were few, and in retrospect the lesions
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Fig. 2. Photomicrographs of the cerebellar hemangioblastoma, third operation (left) and the cerebral hemangioblastoma, fourth operation (right). H. & E., ×400.

were probably cavernous angiomas or angioblastic meningiomas. Despite the reclassification of angiomatous neoplasms by Cushing and Bailey, the differentiation between angioblastic meningiomas and hemangioblastomas has remained difficult.

In the past, diagnosis has depended upon the topographical location of the tumor in relation to the tentorium cerebelli. The tumors arising above the tentorium and often attached to it have been termed angioblastic meningiomas, while tumors occurring beneath the tentorium and often within the cerebellum have been called hemangioblastomas; exceptions have occasionally been recorded.

The microscopic differentiation between the tumors is less well-defined. While angioblastic meningiomas are usually solid tumors and hemangioblastomas are often accompanied by cysts, the histopathologic appearance is usually indistinguishable. Abundant capillaries, reticulin stroma, and "pseudo-xanthoma cells" are characteristic of both tumors. Consequently, a diagnosis of cerebral hemangioblastoma is often made with difficulty.

The cerebral angioblastic neoplasm in our patient has been termed a hemangioblastoma. The gross and microscopic appearance of the tumor and its association with three cerebellar hemangioblastomas in the same patient seem to justify the diagnosis.

Summary

We have reported a patient in whom a cerebral hemangioblastoma had the same gross and microscopic appearance as the several cerebellar lesions removed in preceding years. He also had retinal angiomatosis and a family history of hemangioblastic tumors.

References

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