THE OCCURRENCE OF HEMANGIOBLASTOMAS (TWO CEREBELLAR AND ONE SPINAL) IN THREE MEMBERS OF A FAMILY

W. McK. CRAIG, M.D., AND GILBERT HORrax, M.D.

Section on Neurosurgery, The Mayo Clinic, Rochester, Minnesota, and Department of Neurosurgery, The Lahey Clinic, Boston, Massachusetts

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The familial character of hemangiomatous cysts and tumors of the central nervous system has been stressed by numerous writers, this characteristic perhaps being true particularly of patients having associated angiomatosis of the retina as well as cysts of other organs such as the kidney, pancreas, liver, and so forth, the so-called Lindau-von Hippel disease. Lindau himself believed that about 20 per cent of all cases showed a family incidence.

Although hemangiomatous cysts in the cerebellum with or without similar associated lesions elsewhere are by far the most frequent form of this tumor situated in the central nervous system, nevertheless the spinal cord occasionally has been found to be the seat of hemangiomatous or angiomatous growths. Cushing and Bailey felt that spinal lesions of this nature were extremely rare. However, Sargent stated that he had operated upon 6 patients with spinal cord angiomas, and Wagener, Kernohan and one of us (W.McK.C.) reported several cases of typical hemangioblastoma of the spinal cord in which no other lesion was clinically demonstrable. One of these patients had histologically verified cerebellar and spinal cord hemangiomas, and from his history it is evident that his disease was familial, although it was not possible to verify the lesions in other members of his family except for an angioma of the retina in 1 sister. In 1932 Dorothy Russell reported a case of capillary hemangioma of the spinal cord associated with syringomyelia, noting the great rarity of the condition.

The 8 patients to be reported here present several interesting features. In the first place, the hemangiomatous cysts of the cerebellum in the mother (Case 1) and 1 daughter (Case 2) have been verified histologically. The mother likewise had a probable angioma of the retina (and kidney?) and was reported previously by one of us. One daughter (Case 2) had two separate verified hemangiomas, one in each cerebellar hemisphere. The other daughter (Case 3) has angiomatous or hemangiomatous tumors, identified by inspection, involving both the cervical and lumbar portions of her spinal cord.

Case 1. Slowly developing cerebellar symptomatology with retinal hemangioma difficult to recognize. Ventriculography with removal of hemangioma in the left cerebellar hemisphere. J.F., aged 42, was referred to the Mayo Clinic in September 1937 for examination. She had had some loss of vision in the right eye for 11 years which had
been diagnosed as chorioretinitis with scotoma. Recently intermittent headaches and vomiting had developed.

**Family history** was negative except for questionable migraine headaches in the mother.

**Present Illness.** The patient noticed difficulty in vision of the right eye which had been present for 11 years with no other symptoms and was considered to be chorioretinitis of tuberculous origin. Because of the headaches, an encephalogram was done, which was negative. She returned in June 1938, with a history that following encephalography there had been some relief of the headaches and very few neurologic symptoms. Four months before re-examination headaches, projectile vomiting, and some unsteadiness of gait had developed.

**Examination.** The patient was ambulatory, cooperative and fully oriented. There was marked incoordination of the left arm and leg with slight ataxia. Examination of the eyes revealed nystagmus on looking to the right, and papilledema of 4 D. on the right and 5 D. on the left. The left visual field was normal; the right showed a loss of the temporal field and the nasal upper quadrant, with diminution of vision which could have been explained by the retinal lesion. The retina had the appearance of proliferating retinitis with detachments of the lower portion of the retina in the right eye. In view of the development of cerebellar signs, the retinal lesion was reconsidered and the consensus was that it might be an angiomatous lesion of Lindau-von Hippel's disease.

**Operation** (W. McK.C.). On June 24, 1938, ventriculography was performed with removal of several ounces of fluid. Subsequent roentgenograms revealed dilatation of the lateral and 3rd ventricles with some elevation of the posterior horn of the left lateral ventricle.

A suboccipital craniotomy was done in the upright position under intratracheal anesthesia. When the dura was opened, the inferior tonsil of the left cerebellar lobe was found to be herniated below the level of the 1st cervical vertebra. The convolutions of the left cerebellar lobe were broadened and flattened and the entire lobe was covered with large, engorged, tortuous vessels (Fig. 1, a). A needle was inserted and 0.5 cm. below the surface a cyst was encountered from which 5 cc. of fluid were removed. The fluid was yellow and coagulated on standing. The vessels on the surface of the cerebellum were ligated and coagulated. An incision was made into the cyst and a nodule was found lying on the lateral wall (Fig. 1, b). The cyst wall, including the mural nodule, was removed completely. Convalescence was satisfactory.

**Subsequent Course.** An examination in March 1945, 7 years after operation, revealed no papilledema and no neurological disturbances. Vision was normal except for the defect due to the retinal lesion of the right eye which had not increased in size.

**Pathologic Report.** The tumor, which weighed 5 gm. and measured 3\times2\times2.5 cm., was a hemangio-endothelioma. On microscopic examination it was found to contain more large thin-walled blood spaces than are usually seen in hemangioblastomas. There were regions in which numerous small blood spaces were lined by endothelial cells, some of which were swollen and contained vacuoles that gave the cells a typical foamy appearance. There were, however, more foam cells between the blood spaces than actually lining the spaces. Staining showed the vacuoles of the foam cells to contain fat. The relationship of the xanthic cells to the walls of the blood spaces was best demonstrated with a combination of Perdrau's silver impregnation method for connective tissue and scarlet red staining for fat.