SPINAL HEMANGIOMA (HEMANGIOBLASTOMA) IN LINDAU’S DISEASE

REPORT OF SIX CASES IN A SINGLE FAMILY*

FRANK J. OTENASEK, M.D., AND MAURICE L. SILVER, M.D.†

Division of Neurological Surgery, Johns Hopkins University Medical School and Hospital, Baltimore, Maryland

(Received for publication June 22, 1960)

It is now more than thirty years since Lindau first described the complex of multiple hemangiomas of the nervous system and viscera, which carries his name.

He noted multiple angiomata of the central nervous system, especially retinal angiomata, cerebellar cyst and spinal cord angiomata. These were associated frequently with cysts of the pancreas, and lung, and sometimes with so-called “hypernephroid” tumors of the kidney, and cavernous angiomata of the liver. Lindau placed greatest emphasis on the retina and cerebellum as sites of election for the occurrence of angiomata (called “hemangioblastoma” by Cushing and Bailey and re-named “hemangioma” by Silver and Hennigar) but in 1936 Levin showed that the spinal cord was involved in every proven case of the disease in which postmortem examination of the spinal cord was made. Levin emphasized the hereditary nature of the disease; Lindau in 1931 already had stated that the condition was familial in 90 per cent of the cases studied.

As late as 1944, Wyburn-Mason, in his excellent monograph The Vascular Abnormalities and Tumours of the Spinal Cord and its Membranes, stated that (page 81) no case had shown familial or hereditary influx, except that of Brandt, which was associated with cerebellar lesions, and his own Case 54 in the monograph. (Wyburn-Mason reported 33 cases of spinal hemangioblastoma from the literature, and added 14 of his own.) In 1947 Kinney and Fitzgerald added 3 spinal cases to the literature, and more recently Craig and Horrax reported the inheritance of the spinal form of Lindau’s disease in 2 of 3 cases in one family.

CASE HISTORIES

The opportunity to investigate the familial tendency in Lindau’s disease was provided by the admission to The Johns Hopkins Hospital in August 1948 of a 28-year-old male, member of a family with the hereditary trait, described in the Journal of the American Medical Association (page 81 on genealogy chart, Fig. 1).

His complaint on admission was weakness and numbness of the lower extremities. Two years before, he had been seen at Duke University Hospital because of poor vision in his left eye. This
was found to be caused by an angioma of the retina. Visual acuity was 20/100 in this eye. No special ophthalmologic treatment was given and vision was the same on his admission to The Johns Hopkins Hospital.

Following his visit to Duke University Hospital, slow onset of weakness developed in both legs. Examination on admission showed a band of total anesthesia corresponding roughly to T7 and T8 on the right side. There were spasticity and weakness of both legs and hypesthesia from T8 down, with considerable sparing of the sacral segments. Lumbar puncture showed a block, with xanthochromic fluid containing a total protein of 1,000 mg. per cent. A cisternal Pantopaque injection was carried out (Fig. 2). The radiographs show clearly the presence of a great mass of tortuous vessels in the dorsal region of the spine, and also indicate that the lesion extends into the lower cervical region.

Operation was carried out in the mid-dorsal region, with the finding of a mass of vascular tumor. There was great tortuosity of the blood vessels over the cord, as predicted from the myelogram. The tumor itself could not be exposed adequately or separated from the spinal cord. Decompression was performed.

Four months later, after some slight increase in signs and symptoms, roentgen-ray therapy was instituted. There has been further change over the succeeding 10-year period, with progression to complete paraplegia, and involvement of the upper extremities.

This patient led us to the study of other members of the family known to us, as well as other members of the family examined and treated elsewhere (Fig. 1). The patient referred to is #4 in this portion of the family record. The case histories of the other members of the family follow:

#1 C.D. This patient died in 1921 at the age of 32. He apparently had been in good health until the age of 24, when he began to have severe headaches. Shortly thereafter he complained of visual defects, cataracts developed and he underwent ophthalmologic surgery in Cincinnati, without improvement of his vision. Subsequently there developed signs of intracranial involvement, and he was sent to the late Dr. Charles Frazier at the University of Pennsylvania Hospital. We are able to ascertain from a letter of Dr. Frazier to the family doctor that he suspected a lesion of the right parietal lobe. A right parietal craniotomy was performed, but no tumor was found.

The patient died 4 days after operation. At autopsy, a so-called “endothelioma of the brain stem” and another tumor of the cauda equina were found. We have not been able to obtain the pathologic specimens of this patient, but the finding of a spinal lesion is quite clear.

#2 L.D.P. This is the case of a female who was paralyzed on the left side and blind in the left eye at the time of her death at the age of 41.

Visual loss in this eye developed gradually at 22 years of age and was complete at 34. She had no other incapacitating symptoms and was an active member of the community. She had five children and was able to care for her household without difficulty.

Apparently she commented occasionally about symptoms of numbness of her left arm, but it was only when weakness of that arm developed that her complaints were serious enough to require medical attention. She had bizarre attacks involving “shaking” of the left arm (which were later considered to be Jacksonian seizures). Severe headaches developed, sometimes occipital, but more often frontal in location. Then, episodes of nausea and vomiting occurred and she came...