CASE REPORTS AND TECHNICAL NOTES

HEMANGIOBLASTOMA OF THE POSTERIOR FOSSA (LINDAU'S DISEASE)

REPORT OF TWO CASES WITH FAMILIAL HISTORY

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Posterior fossa hemangioblastoma is of especial interest because of its frequent occurrence as a true familial disease. Lindau† noted hemangioblastoma of the retina (von Hippel's disease) in 1 of his cases. In reviewing the literature on von Hippel's disease, he collected several additional examples of retinal hemangioblastoma associated with cystic hemangio-

blastomas of the cerebellum. In all, Lindau studied 16 cases of his own and 24 additional ones from the literature. Most of this material was from postmortem examinations and gave Lindau the opportunity of observing angioblastic lesions of the spinal cord, cystic kidney, hypernephroma, tumor of the epididymis and angiomatosis of the liver which he described as occurring in hemangioblastomatosis.

Cushing and Bailey‡ reported 11 hemangioblastomas of the cerebellum, in 4 of which the cysts comprised the bulk of the lesion. Cysts occurred in 4 additional cases, but in these the main mass was solid. Their 1 case with a definite family history and with both cerebellar and retinal hemangioblastomas was reported separately. It is probably the first example in which histologic verification was accomplished in the life of a patient. The work of von Hippel, Koch, Seidel and others, which led to Lindau's important contribution, has been covered in the monograph by Cushing and Bailey and will not be discussed here.

MacDonald§ reported 4 cases of hemangioblastoma of the cerebellum, surgically verified in living patients, bringing the total of patients successfully operated upon to 10. Two of the reported patients were sisters whose mother (unreported case) was known to have serious eye trouble (suggestive of von Hippel's disease) and who died of an intracranial lesion. The other two patients were a brother and a sister whose father (unreported case) had serious eye trouble and who died of an intracranial lesion. In none of these 4 patients was hemangioblastoma of the retina found at examination.

Craig, Wagener and Kernohan∥ reported 4 patients with combined retinal and cerebellar hemangioblastoma, 3 of whom survived operation with follow-up periods of 7, 1½ and 3½ years. An extensive family history of similar disorders was present in their Case 4.

Grossman and Kesert¶ found 4 instances of cerebellar tumor in one family. There was histological verification in the father, aged 45, who died, and in a son, aged 23, who recovered following operation. A daughter, aged 24, and a son, aged 19, died from what were thought to be cerebellar tumors. However, no tumor was verified by surgery or postmortem examination. In the last-mentioned patient the posterior fossa was opened at operation, but the dura was not incised.

This brief survey of the literature reveals the utmost importance of a detailed family history. In addition, the retinal tumors are of extreme importance when they are present and may well escape a casual ophthalmoscopic examination even if performed by an experienced clinician.

In the patient whose history and physical examination are strongly suggestive of a posterior fossa tumor, hemangioblastoma is extremely probable if there is either a familial history of posterior fossa tumor, a familial history of retinal hemangioblastoma or the actual presence of von Hippel's disease. The diagnosis is much more certain if, in the family history, there has been confirmation of a cerebellar cyst, or even better, histologic confirmation of an angioblastic tumor. Other clinical manifestations of Lindau's disease are not likely to be identified.

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during life. It appears to the author that a frank family history is clinically as important in
the individual case of posterior fossa hemangioblastoma as is the presence of von Hippel's
disease, and warrants the use of the term, Lindau's disease.

The patient with an angioblastic lesion of the posterior fossa with neither retinal tumor
nor family history cannot be considered an example of Lindau's disease without confirmation
by finding angioblastic changes in other organs.

REPORT OF CASES

Case 1. M.L.M., a 20-year-old white sailor, was admitted to the hospital on Mar. 17, 1945.
For 6 months suboccipital headaches had been present, and, during the last 3 months, nausea
and vomiting occurred additionally. Diplopia, failing vision, and unsteady gait of 1 month's
duration precipitated hospital admission.

The family history revealed that the father died of a cerebellar tumor, an hemangioblastoma,
confirmed by autopsy at a university hospital. The sister of the patient died from a
proved cerebellar tumor, and one paternal cousin also died from a proved cerebellar tumor.
There was no familial history of visual disturbance suggestive of von Hippel's disease. One
brother and one sister of the patient were in perfect health.

Examination. The general physical findings were negative throughout. Neurological
study revealed a rather dull patient with slurred speech. Papilledema of 2D, was present. A
careful examination of the optic fundi with the pupils fully dilated failed to reveal abnormal
vessels or tumors. Course horizontal nystagmus occurred on gaze to right or left. There was
moderate nuchal rigidity and moderate tenderness to pressure in the suboccipital region. The
gait was unsteady, and the right upper extremity showed a greater degree of ataxia and
dysdiadokokinesis than the left.

Routine laboratory work and roentgenograms of the skull and chest were negative. With
moderate cerebellar signs predominating on the right and with the clear-cut family history a
diagnosis of probable right cerebellar hemangioblastoma was made. It was decided that
ventriculography would be omitted if ventricular puncture at operation disclosed dilated
ventricles.

1st Operation. On May 5, 1945, under endotracheal ether anesthesia with the patient in
the prone position on the cerebellar head rest, bilateral trephine openings were made. The
ventricle was entered at a depth of approximately 5 cm, on each side, with a free flow of fluid
from each side. A mastoid-to-mastoid cerebellar incision was made and the suboccipital
region exposed. A trephine opening over the right hemisphere was made first and a cannula
introduced to a depth of 3 cm. Approximately 25 cc. of yellow, clotting fluid was obtained.
The exposure was then completed with removal of the arch of the atlas. A cortical incision
was made over the cyst, but adequate exposure could not be obtained because of swelling of
the cerebellum. At this time the patient appeared in critical condition with respiration shallow
at 70 per minute. Blood pressure was 110/60, but pulse rate was 170. The anesthetist could
not explain the change in the patient's condition. The Frazier cannula which had been placed
in the right lateral ventricle was exuding a little blood, and a free flow of fluid could not be ob-
tained on repeated puncture. Pink fluid was obtained from the left lateral ventricle. With
fear that a right ventricular hemorrhage accounted for the change in condition, the right oc-
cipital opening was enlarged and a small cortical incision was made down to the ventricle. No
significant hemorrhage was encountered. The wounds were closed rapidly, and the patient was
returned to his room with but slight hope for his survival.

Course. There was rapid improvement with continuation of supportive treatment, and on
the day after operation the patient's condition was entirely satisfactory. No further complica-
tions occurred.

2nd Operation. On June 14, 1945, under local anesthesia, the old incision was reopened and
the cerebellum exposed. The collapsed cyst wall was identified. It contained a rounded 3 cc.
yellow coagulum. In the caudal depth of the cyst a reddish-brown nodule, 1 cm. in length,
was identified (Fig. 1). Two large vessels entered each pole. These were coagulated, and the