FAMILIAL HEMANGIOBLASTOMA OF THE CEREBELLUM

PEDIGREE OF TWO FAMILIES

JOHN E. ADAMS, M.D.

Department of Neurological Surgery, University of California School of Medicine, and the San Francisco City and County Hospital, San Francisco, California

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The hereditary basis for the occurrence of some hemangioblastomas of the central nervous system has been well established. However, the reports of such families are sufficiently scarce to warrant recording two additional families having members of several generations afflicted with this lesion.

Case 1. M.V., a 22-year-old married woman of Hawaiian extraction, was first admitted to the San Francisco Hospital on Aug. 1, 1948, complaining of occipital and frontal headaches accompanied by nausea and vomiting for the previous 3 months. Physical and neurological examinations were negative. The family history obtained from the patient indicated only that her father had died of a brain tumor of unknown type. Subsequently, it was revealed that several members of the paternal branch of the family in the preceding two generations had had brain tumors (Fig. 1).

Fig. 1

The patient was discharged, but was admitted to another hospital in January 1949, for the same persisting complaints. A laparotomy and appendectomy were performed without relief. Irrational and confused behavior followed this operation, and after falling out of bed on her 6th postoperative day, she was readmitted to the San Francisco Hospital.

Examination. At this time, papilledema was noted, accompanied by bilateral extensor plantar responses and bilateral external rectus palsies. Roentgenograms of the skull were normal. Lumbar puncture demonstrated a CSF pressure of 290 mm. of water, xanthochromic fluid, 232 RBC/c.mm., and a protein content of 450 mg. per cent. Because of the fall, a diagnosis of subdural hematoma was entertained but trephination was negative. Subsequent ventriculography revealed a uniformly dilated ventricular system without deformity although the 4th ventricle was poorly filled with air.

Course. Following ventriculography the patient improved. The headaches and diplopia disappeared and the papilledema became less marked.

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She was next seen 10 months later, again complaining of headaches and difficulty with speech. Papilledema was marked (6 D. bilaterally), and examination disclosed truncal ataxia and palsies of the 5th, 6th, 7th, 9th, 10th, and 12th cranial nerves on the left.

Operation. An exploration of the posterior fossa was performed on Oct. 27, 1949, and a large cystic hemangioblastoma of the left cerebellar hemisphere was removed. A similar non-cystic tumor involved the upper cervical cord and lower brain stem. A biopsy was made of this.

Course. Roentgen therapy was given following a stormy postoperative course. She remained well until August 1951, when she suddenly expired at home. Permission for an autopsy was not granted.

In the following two cases the patients were brother and sister and were both hospitalized at the University of California Hospital at the same time with almost identical histories and tumors.

Case 2. E.M., aged 40, brother of Case 3, was admitted in April 1950. He had suffered from increasingly severe occipital headaches for the previous 4 months. He had noted staggering gait for 2 months.

Examination. Positive neurological findings were 2 D. of papilledema bilaterally, slight truncal ataxia, and fine nystagmus on right lateral gaze. Ventriculography demonstrated a symmetrical hydrocephalus and obstruction of the 4th ventricle.

1st Operation. An immediate suboccipital craniectomy was performed, and a large midline cyst of the cerebellar vermis with a nubbin of tumor at its superior pole was encountered and removed.

Microscopic examination of the tumor disclosed the typical picture of hemangioblastoma. Course. He remained well for 2½ years, when he reentered the hospital with signs of recurrent tumor.

2nd Operation. Re-exploration disclosed a large solid tumor involving the left cerebellar hemisphere. Successful removal was accomplished and postoperative irradiation was instituted.

Course. At present, the patient is well.

Case 3. V.N., aged 35, sister of Case 2, was admitted 2 weeks after her brother's first admission. Her complaints were similar to his, namely, occipital headaches, staggering gait, morning vomiting, and vertigo.

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**Father**
Died. Age 50
Cerebellar tumor
Type unknown

**Mother**
Alive and well

<table>
<thead>
<tr>
<th>Father</th>
<th>Mother</th>
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<tbody>
<tr>
<td>☣—Sister</td>
<td>☣ (E.M.) UC HOSP.</td>
</tr>
<tr>
<td>☣—Brother</td>
<td>☣ (V.N.)</td>
</tr>
<tr>
<td>☣—Sister</td>
<td>☣—Sister</td>
</tr>
<tr>
<td>☣—Brother</td>
<td>☣—Brother</td>
</tr>
<tr>
<td>Died. Age 40</td>
<td>Age 28</td>
</tr>
<tr>
<td>Died following removal of right cerebellar tumor, type unknown 1947</td>
<td>Died during removal of right cerebellar hemispheric hemangioblastoma 1930.</td>
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**SISTERS**

<table>
<thead>
<tr>
<th>1st Paternal cousin</th>
<th>1st Paternal cousin</th>
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<tr>
<td>☣ — died at age 46 after removal of cerebellar cystic hemangioblastoma.</td>
<td>☣ — died at age 38 after removal of cerebellar cystic hemangioblastoma.</td>
</tr>
<tr>
<td>Grandparents died of natural causes.</td>
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Examination. Two D. of papilledema were present bilaterally and moderate truncal ataxia was noted. Ventriculography demonstrated a block in the posterior fossa.

Operation. Immediate suboccipital craniectomy disclosed a cystic and solid tumor involving both the cerebellar vermis and the right cerebellar hemisphere. The patient was operated upon in the sitting position and unfortunately expired during the procedure.

Autopsy disclosed air embolism as the cause of death. The cerebellar tumor was a cystic hemangioblastoma. In addition, there were present cystic adenomata of the pancreas.

The family pedigree of Cases 2 and 3 is shown in Fig. 2. In none of the 3 cases was any angiomatous involvement of the retina demonstrated. It is of interest that in each of these families, symptoms referable to the tumor appeared at a younger age in successive generations. This tendency has been commented upon in previous reports of such families.1

REFERENCE


BRAIN NECROSIS FOLLOWING X-RAY THERAPY

ELDON L. FOLTZ, M.D.,* JOHN B. HOLYOKE, M.D., AND HENRY L. HEYL, M.D.

Department of Neurosurgery and Department of Pathology, Dartmouth Medical School, Mary Hitchcock Memorial Hospital, and Hitchcock Clinic, Hanover, New Hampshire

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Brain and nerve tissue are usually resistant to direct damage by x-ray radiations, even with high dosages.1,4,11 There is increasing evidence, however, that in rare instances the vascular system of the brain can be severely damaged by irradiation.2,7,8,10,12,14,15 We have recently studied a case that we feel demonstrates this phenomenon. The patient, originally treated for a basal cell epithelioma of the scalp with x-ray therapy, subsequently showed evidence of an underlying, expanding, intracranial lesion. At operation a non-tumor mass was removed from the temporal lobe; microscopic study of this mass revealed startling blood-vessel changes.

CASE REPORT

Past History. L.C., a 45-year-old white female, was first seen in November, 1945, when a biopsy diagnosis of a basal cell epithelioma of the right temporal scalp was made. Superficial x-ray therapy (3500 r) was given to the lesion on Nov. 12, 1945. During the next 2 years, similar lesions appeared over the neck, shoulder, chest, and abdomen. These were treated as they appeared, either by local excision or by electrofulguration. The tumor in the right temporal area reappeared in 1948. Further x-ray therapy was felt unwise because of the marked scarring, atrophy, and depilation of the skin in that area. The lesion was excised and again a diagnosis of basal cell epithelioma was made. She had no complaints suggesting intracranial disease at that time.

Present Illness. On July 30, 1948, the patient was brought to the hospital because of lethargy, confusion, incontinence, and headache of 4 weeks' duration. The onset had been marked by disabling pain and weakness in both legs and was followed by urinary and fecal incontinence. Daily convulsive seizures, lasting about 10 minutes, started 2 weeks before admission; there were no recognized aura or localizing manifestations.

Examination. She was a well-developed, obese, white woman with marked facial hirsutism. Her pulse was 80, respiratory rate 18, and B.P. 145/85. She was apathetic, lethargic, and

* Present address: University of Washington Medical School, Seattle 5, Washington.