"POLYCYTHEMIA" ASSOCIATED WITH CEREBELLAR HEMANGIOBLASTOMA

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IN 1943, Carpenter and co-workers\(^3\) reported 2 cases of cerebellar hemangioblastoma with associated "polycythemia vera." They added 3 cases from the literature. Since then, about 22 such cases have been reported in the American literature.\(^3,4,8,12,21,25\) Hemangioblastomas of the posterior fossa constitute only 2 per cent of intracranial tumors\(^5,6,16\) and those associated with "polycythemia" less than 20 per cent of the total number of hemangioblastomas.\(^4\) Thus, the association of "polycythemia" with posterior fossa hemangioblastoma is, to paraphrase Carpenter, an event of the greatest rarity.

Polycythemia rubra vera has been defined as a disease of unknown etiology, characterized by an excessive production of all marrow elements with resultant increase in red blood cell count, total red blood cell volume, white blood cell and platelet counts, and accompanied by increased blood viscosity and decreased velocity of blood flow.\(^19,23\) Since quantitative blood studies have been done in only 1 of the reported cases of cerebellar hemangioblastoma,\(^3\) it is difficult to determine whether polycythemia vera (as defined above) did, in fact, exist in the remainder. Possibly erythrocytosis, in which the red blood cell count is elevated with no increase in white blood cells or platelets, might be a more appropriate term to use.

We have recently studied 2 patients who had posterior fossa hemangioblastomas with associated erythrocytosis. Both patients were treated surgically, one with subtotal and the other with total removal of the tumor. We believe that the hematologic studies made in these 2 cases are more nearly complete than those reported heretofore.

It has not yet been proved that the relationship between the altered blood picture and the tumor is more than fortuitous. Although the basic physiopathology is unknown, the reduction in the degree of erythrocytosis in some cases following removal of the tumor suggests a cause-effect relationship. We hope that the cases here reported will yield some clue to the mechanisms involved.

CLINICAL DATA

Case 1. W.D., a 45-year-old white male, was admitted to the neurosurgery service on Nov. 3, 1952.

While in the Army in 1941, the development of signs of increased intracranial
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pressure and cerebellar dysfunction led to a diagnosis of cerebellar tumor. Ventriculo
ulography revealed an "internal hydrocephalus" and was followed by suboccipital
craniectomy. No gross tumor was found. Following operation, the patient was re-
lieved of his symptoms for 3 years. Symptoms recurred in 1946 and he then received
2000 r of x-ray therapy to the posterior fossa.

He was next seen in another hospital in 1947 with no change in his clinical pic-
ture but with a red blood cell count of 5.38 million. One year later he was re-ad-
mitted to that hospital with progression of his signs of cerebellar dysfunction and
with a red blood cell count of 6.5 million. He was seen intermittently in the out-
patient department until 1949. His discharge note read, "The Tumor Board feels
that there is a question of this man ever having had a tumor."

A rapid increase in his neurological deficits prompted admission to this hospital
on Nov. 3, 1952.

Examination. The physical findings supported a diagnosis of cerebellar neo-
plasm. Lumbar puncture yielded clear, colorless fluid under a pressure of 264 mm.
of cerebrospinal fluid. There were no cells; protein was 88 mg. per cent. Vertebral
angiography revealed a diffuse vascularity of the posterior fossa suggestive of a
vascular neoplasm. (Hematologic studies are reported in a later section.)

Operation. On Nov. 19, 1952, a posterior fossa exploration was carried out. An
extensive meningocle was encountered on the right and there were three smaller
separate ones on the left. Upon exposure of the cerebellum, a large extremely vas-
cular tumor was occupying most of the posterior fossa and involving virtually the en-
tire right cerebellar hemisphere. The neoplasm was gray and had numerous large
tortuous venous and arterial channels whose major supply was not identified. Ap-
proximately one-third of the tumor was excised from the inferior and lateral por-
tions of the exposed mass.

Postoperative Course. The patient exhibited no over-all improvement in his
neurological deficits. He was discharged to a nursing home on Feb. 19, 1953.

On April 2, 1953, he was returned to the hospital because of sudden coma. De-
spite frequent periods of apnea, he responded to therapeutic efforts and was awake
within 2 hours. However, from this point he deteriorated rapidly and died on the
following morning. Cause of death was reported as aspiration pneumonia.

Autopsy. The fresh brain showed a moderate amount of thin bloody subtentorial
fluid. There was a firm irregular mass occupying the right cerebellar lobe and a num-
ber of large tortuous veins along the right side of the brain stem and pons emerged
from the mass. The remainder of the brain was grossly normal except for slight
flattening of gyri and narrowing of sulci.

After fixation, the neoplasm was found to occupy almost the entire right cere-
bellar hemisphere. Some entering veins measured 4 to 5 mm. in diameter. On cut
section, the most striking feature was the number of large vascular channels. The
tumor was an admixture of grayish-white and reddish-brown tissue with scattered
yellow foci, these latter presumably being xanthomas of "foam cells."

The microscopic picture was likewise characterized by striking vascularity. There
were many thin-walled vessels of irregular shape with numerous sinuosoids
and venous lakes. The stroma showed two dominant characteristics—an amorphous
eosinophilic matrix and packed masses of foam cells. Pathologic diagnosis: heman-
gioblastoma (Fig. 1).

Case 2. P.V., a 32-year-old white male, was admitted to the medical service on
April 10, 1952.