Hemangioblastoma of the Cerebellum Associated with Erythrocytosis and an Unusual Blood Supply

Case Report

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The association of erythrocytosis with various tumors and cysts is well known. These conditions include: renal cysts, renal hypernephroma, uterine fibroids, and cerebellar hemangioblastomas. Patients with these disorders usually have an absolute increase in red cell mass without leukocytosis, thrombocytosis, abnormal blood gases, or splenomegaly. Erythropoietin activity has been demonstrated in cyst fluid aspirated from three patients with cerebellar hemangioblastomas. To our knowledge no patient has been reported who had erythropoietin activity demonstrated in a solid hemangioblastoma.

In 1956, Bernasconi and Cassinari described an artery that they felt arose from the external carotid system associated with several cases of tentorial meningiomas. Subsequently, it was demonstrated by a number of authors that this vessel arose from the cavernous portion of the internal carotid artery. Additional reports have appeared documenting its association with vascular malformations, malignant gliomas, and ependymomas in the posterior fossa; however, we could find no reports of its association with a hemangioblastoma. The present case appears to represent such an instance, and in addition, high titer of erythropoietin activity were demonstrated in a saline extract of the solid portion of the tumor.

Case Report

The patient was a 39-year-old white man with the following history. On January 14, 1965, he had had a documented subarachnoid hemorrhage associated with some dysdystaxia of the right arm and leg, which subsequently cleared. A left carotid angiogram was reported to have been normal. The hemoglobin at that time was 16.1 g/dL with a hematocrit of 45%. At the time of admission to the University of Nebraska Hospital on December 9, 1966, the patient reported increasing difficulty with balance and a staggering gait of 6 months' duration. For 2 months before admission he had had intermittent headaches and some blurring of vision.

Examination. The patient had bilateral papilledema. The reflexes were generally hyperactive, the right more than the left, without Babinski responses, and there was a lack of associated movement of the right arm when he walked. There was a slight unsteadiness on tandem walking, terminal dystaxia of the right finger-to-nose test, and dysdiadochokinesia of the upper extremities, the right greater than the left.

At admission the hemoglobin was 19 g/dL, the red blood cell count (RBC) 6.33 million, and the packed cell volume (PCV) 58% with a normal white blood cell count and differential; repeat blood counts confirmed the original reports. Blood gas studies revealed normal pO₂ and pCO₂. Bone marrow studies revealed erythroid hyperplasia consistent with secondary erythrocytosis. The total red cell volume was 40.1 cc/kg compared to a normal volume of 31.8 ± 3.5 cc/kg, while total plasma volume and total blood volume were within normal limits, providing evidence of an absolute erythrocytosis.

Brain scan revealed an increased uptake in the right side of the posterior fossa. Bilateral carotid angiograms revealed a vascular lesion in the region of the right cerebellar hemisphere receiving its blood supply from two vessels which apparently arose from the cavernous portion of the right internal carotid (Fig. 1). The left carotid was negative. A retrograde axillary angiogram revealed

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marked filling of the same vascular mass from the vertebral system. It appeared to derive a large portion of its blood supply from the right superior cerebellar artery (Fig. 2). Both studies demonstrated rapid arteriovenous shunting.

Because of the patient's erythrocytosis, a diagnosis of cerebellar hemangioblastoma was entertained; however, the extreme vascularity of the lesion raised the question whether it might be an arteriovenous malformation. Because of the vascularity it was elected to perform a subtemporal approach in order to clip the superior cerebellar artery.

Fig. 1. Right carotid angiogram. Note two vessels arising from cavernous portion of internal carotid supplying lesion in posterior fossa. Smaller inferior vessel (open arrow) arises from proximal caudal portion, and larger superior vessel arises more rostrally (closed arrow).

Fig. 2. Right vertebral angiogram. The lesion derives most of its blood supply from the vertebral system. Note large right superior cerebellar artery. Left: Lateral view. Right: Anteroposterior view.