CEREBELLAR HEMANGIOMA (HEMANGIOBLASTOMA)

A CLINICOPATHOLOGICAL REVIEW OF 40 CASES*

MAURICE L. SILVER, M.D.,† AND GORDON HENNIGAR, M.D.;‡

Division of Neurological Surgery and Department of Pathology,
Johns Hopkins Hospital, Baltimore, Maryland

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In the classical monograph Tumors Arising from the Blood-Vessels of the Brain, Cushing and Bailey⁴ reported 11 cases of "hemangioblastoma" of the cerebellum. Recent analysis of this cerebellar neoplasm was made by Cramer and Kimsey⁵ who presented the clinical findings in a larger series of cases, in 37 of which the tumor was histologically verified. The present paper describes the clinical and pathological findings in 40 cases of this neoplasm, which we prefer to designate "hemangioma" of the cerebellum. We use this term, as others have in the past, in place of the name "hemangioblastoma" proposed by Cushing and Bailey, for reasons that will be presented below.

CLINICAL ASPECTS

There is no predilection for either sex. In our series, there are 24 males and 16 females. The age incidence was from 3 to 62 years of age, the highest falling in two decades, the third and sixth (Fig. 1). The average age was 36.2 years. Only one member of the group belonged to the colored race.

The commonest presenting symptom was headache. Other manifestations of an expanding lesion of the posterior fossa were noted, such as ataxia, vertigo, vomiting and blurring of vision. Choked disc was present in 85 per cent of the cases. The average duration of symptoms prior to operation was 10 months.

All of the group were patients at the Johns Hopkins Hospital. Thirty-one were operated upon by Dr. Walter E. Dandy, the remainder by other attending or resident surgeons of the hospital staff. In view of the var-

Fig. 1. Age incidence of cerebellar hemangiomas.

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† Present address: 224 Thayer Street, Providence, Rhode Island.
‡ Present address: Department of Pathology, Medical College of Virginia, Richmond, Virginia.
ation in surgical technique, a detailed analysis of mortality is not warranted.

The operative mortality for the group was 20 per cent, 2 patients dying shortly after admission before operation could be performed. Of the 30 patients surviving surgery, 7 are now dead, having had an average survival period of 10.7 years; the life expectancy in this group, as computed from actuarial tables, is 37 years. At least 2 of the patients died from causes unrelated to the tumor. Twenty-three patients are still alive and their average survival to date is 6.2 years. The 5-year survival rate, commonly applied in the analysis of tumor surgery, is 50 per cent.

Nine of the 40 patients were subjected to more than one operation, 2 patients having had four operations. It is not clear whether these reoperations were performed because of independent isolated foci manifesting themselves at a later date, or because of continued growth of incompletely removed tumor tissue.

The late appearance of hemangiomas of the central nervous system in Lindau's complex is well known. In our series, 9 patients had proven Lindau's disease, 4 of them requiring multiple operations. This figure of slightly more than 20 per cent in our series of cerebellar hemangiomas agrees with that of Lindau, who found that 20 per cent of the patients with retinal hemangioma (so-called von Hippel's disease) subsequently were shown to have a cerebellar neoplasm or other evidence of Lindau's complex. It raises the serious possibility that all patients with a cerebellar hemangioma are suffering from Lindau's disease in which other members of the pathological complex have not yet appeared.

In the diagnosis of the cerebellar hemangioma, clinical laboratory examinations are of limited value. The cerebrospinal fluid protein seldom is elevated and then only minimally. This is in contrast to the spinal hemangiomas, with which it is exceedingly high. The explanation for this probably lies in the fact that the former are seldom in contact with the subarachnoid space, whereas the spinal lesions almost invariably involve this space.

In examination of the peripheral blood, we found that 40 per cent of these patients had hemoglobin values greater than 15 gm., and in 3 cases values as high as 18.5 gm., with a red blood cell count of 6.3 million, were noted. In none of the cases were blood and plasma volume determinations, arterial oxygen saturations, or bone marrow studies performed, yet in these 3 cases, the clinical diagnosis of polycythemia rubra vera was made and treated by venesection.

PATHOLOGICAL ASPECTS

Hemangioma of the cerebellum may be cystic or solid, single or multiple, and situated either in the midline or in the hemispheres. Forty per cent of the tumors showed attachment to the meninges, half of these (8 cases) to the dura, usually to the ventral aspect of the tentorium. Thirty tumors consisted of a nodule with a single large cyst, 4 had a grossly multiloculated cystic appearance, and 6 were solid.