The So-Called Solid Hemangioblastomas of the Cerebellum and Vertebral Angiography

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Despite the extensive application of cerebral angiography during the last two decades, it is our impression that this method is used relatively little in the management of the diagnostic problems of expanding lesions of the cerebellar fossa. This may be ascribable to several reasons, such as the lack of a satisfactory technique to allow a regular and adequate visualization of the vertribobasilar arterial system, the more difficult interpretation of the posterior circulation resulting most probably from insufficient familiarity with its angiographic patterns, and the fact that the yield of the angiogram in the expanding lesions of the cerebellar fossa is much less than in those located supratentorially.

Although the present techniques for filling the vertebral system still leave much to be desired, they have improved sufficiently to give data that otherwise are unobtainable by the rest of the preoperative investigations. This information has diagnostic value and is of great help in planning the surgical attack, the correctness of which will determine the success or the failure of the therapeutic procedure in a number of cases.

The so-called solid cerebellar hemangioblastomas are among such instances since they offer serious difficulties for their total excision in contrast with the so-called cystic hemangioblastomas in which the small neo-plastic nodule in the wall of a large nontumoral cavity is of easy surgical control. Their incidence is small, but they are sufficiently significant, particularly if we take into consideration the fact that they can be removed at operation, totally and successfully.

The angiographic demonstration of this type of lesion was first shown by Olivecrona. In his case the tumor was not found at the 1st operation, but was demonstrated by vertebral angiography carried out some days later. A detailed description of the radiographic appearance was made by Lindgren.1 We have considered it interesting to present 2 of our patients who, in our judgment, illustrate the usefulness of angiography in the diagnosis and treatment of these cases.

One of our patients was a 14-year-old girl, a fact which in itself represents an unusual instance since it is exceptional for cerebellar hemangioblastomas to manifest themselves in patients at this early age.

Case Reports

Case 1. #3-84-17. J.H.F., a 42-year-old white male, came to the J. Hillis Miller Health Center complaining of headache, mild vertigo, and weakness of both lower extremities. These symptoms had been progressing slowly for a period of 1 month. On the morning of admission he became aware of increased weakness of the lower extremities so that his knees buckled up under him. He also noticed an increase in the severity of his headache.

Examination revealed a horizontal nystagmus on both lateral gazes and a mildly ataxic gait with evidence of swaying anteriorly and posteriorly but without falling on the Romberg test. There was an extensor plantar response of the left big toe. The hematocrit was 49 per cent.

On the day of admission, Jan. 17, 1963, right retrograde brachial angiography was performed and a highly vascular lesion, 3 cm. in diameter, was demonstrated in the region of the cerebellar vermis (Figs. 1 and 2). This lesion retained the contrast material late into the venous phase and its main feeding vessels seemed to be the posterior inferior cerebellar artery bilaterally as well as the superior cerebellar artery. The radiographic appearance was that of a hemangioblastoma.

Operation. On the evening of the day of admission a suboccipital craniectomy was performed through a midline incision in the skin and a total extracapsular removal of the neoplasm was accomplished.

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Fig. 1. *Case 1.* Lateral views (arterial and venous phases) of angiogram.

Fig. 2. *Case 1.* Towne’s views (arterial and venous phases) of angiogram.
The histopathological diagnosis was hemangio-blastoma.

Course. This patient ran an uncomplicated postoperative course and he was discharged on Feb. 2, 1963, almost asymptomatic. On April 1, he returned to his original work and he is presently leading a normal life.

Case 2. 3-50-48. C.B., a 14-year-old colored female, was admitted for the first time to the J. Hillis Miller Health Center on Oct. 1, 1962, because of pain in the head and neck and progressive loss of vision since early August 1962. She became practically blind about 3 weeks prior to admission. Also during the 3 or 4 weeks that preceded admission she had complained of staggering and dizziness.

Examination. The outstanding finding on admission was absence of vision bilaterally with equally dilated pupils that reacted sluggishly to light. There were nystagmoid jerks on lateral gaze, and both optic discs were blurred and pale with the appearance of optic atrophy secondary to papilledema. The corneal reflex was absent on the left side. There was some dysmetria on the finger-to-nose test bilaterally and generalized hypotonicity. Percussion of the upper cervical region elicited local pain in that area. She had a hematocrit of 56 per cent with 6,450,000 red blood cells per c.mm. and 6,000 white blood cells per c.mm. in her peripheral blood. Routine roentgenograms of the skull demonstrated evidence of generalized increased intracranial pressure. A mercury203 brain scan was inconclusive and the electroencephalogram was grossly abnormal with diffuse, slow dysrhythmia and some focal alteration over the left posterior hemisphere. The rest of the laboratory data before the ventriculogram was not significant pathologically.

On October 3, ventriculography disclosed a midline space-occupying lesion of the cerebellar fossa, compressing the roof of the 4th ventricle and displacing it anteriorly. In addition there was a mass of soft tissue extending below the foramen magnum to the level of the upper margin of C2, which was interpreted as representing herniation of the cerebellar tonsils.

Operation. Following the ventriculography a suboccipital craniectomy was performed through a midline incision in removal of the posterior rim of the foramen magnum and the lamina of C1. A large neoplastic mass was found in the vermis of the cerebellum and extending down into the spinal canal as far as the upper level of C2. The lower portion of the tumor from the cisterna magna to C2 consisted of a cyst with poorly vascularized walls and containing yellow fluid. The upper part of the tumor, on the contrary, was highly vascular presenting large arteriovenous channels some of which had a diameter of about 5 to 7 mm. Two of these arteriovenous channels ran upward along the surface of the vermis toward the region of the confluence of the sinuses. The most causal and avascular portion of the tumor was punctured with a small needle and markedly yellowish fluid was obtained. The dorsal wall of this cystic cavity was removed with relatively little bleeding, but great care was taken to avoid extension of the resection into the highly vascularized portion of the tumor. At this stage of the operation there was evidence that the circulation of the spinal fluid through the 4th ventricle had been re-established, but nevertheless a Torkildsen’s procedure was carried out. The wound was closed without any further attempt at removing the larger vascular mass of the neoplasm since no satisfactory plane of cleavage could be established.

Histopathological diagnosis of the tissue removed was hemangio-blastoma.

Course. The patient withstood the procedure well and a course of supervoltage external irradiation was given to the mass in the cerebellar fossa. Her general physical condition and the neurological findings on the day of discharge (Oct. 21, 1962) were essentially the same as those noted before operation.

After discharge the patient continued to suffer from intermittent generalized headaches which became worse during the 3rd week of February, 1963. She was seen in the Out-Patient Department on February 26, and signs of elevated intracranial pressure were noted. The area of the suboccipital craniectomy was full and tender and there was evidence of fluid around the catheter underneath the scalp. There seemed to be some decrease in her mental ability and her gait appeared more ataxic, but otherwise general physical and neurological findings were essentially the same as previously recorded.

Readmission, Mar. 5, 1963. Her hematocrit was 46 per cent. A right retrograde brachial angiogram on March 6 revealed a highly vascular mass occupying the region of the vermis of the cerebellum and extending more toward the right than toward the left. From the rate of flow in the various vessels it would appear that the major feeding arterial supply was the posterior inferior cerebellar artery although the vascular staining of the tumor obscures this somewhat. The venous drainage was visualized and the size and boundaries of the rounded and lobulated mass were shown well on the angiographic study (Figs. 3 and 4).

2nd Operation. On March 8, the suboccipital craniectomy was re-opened and a total extra-capsular removal of the remaining neoplasm was accomplished.

Course. The patient withstood the procedure well and her condition was satisfactory until the
evening of the 2nd postoperative day. Then there developed an acute episode of a marked rise in the intracranial pressure with rigidity of decerebration and Cheyne-Stokes respirations. Ventricular taps relieved the situation only temporarily. Since all of the tumor was removed at the 2nd operation a completely free flow of spinal fluid through the aqueduct and 4th ventricle was established, and since adhesions were noted not only on the surface of the cerebellar hemisphere but also around the cerebellopontine angle, it was considered that in all probability there was an adhesive arachnoiditis blocking the circulation of the spinal fluid upward through the basal cisterns at the tentorial notch.

3rd Operation. A Pudenz ventriculo-atrial shunt was inserted.

Course. Following this shunt the intracranial pressure remained under control although she had a long and difficult postoperative course. The polycythemia disappeared but it was not until 2 months after operation that considerable improvement had taken place. She was discharged on June 8, 1963 in satisfactory condition. Arrangements were made for her admission to a school for the blind.

Comment

The angiograms of the cases described
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were obtained by means of a retrograde brachial technique through a cannula using Hypaque 50 per cent, the Taveras pressure injector, and the Sánchez-Pérez seriograph.

The angiographic demonstration permitted an accurate diagnosis of the lesion, its location, extension, and borders, and gave an adequate idea as to the situation of the arterial supply and venous drainage of the tumor. The knowledge of these details facilitated the control of the vascular pedicles and the establishment of the correct plane of cleavage for the total extracapsular removal of the neoplasm. Our Case 2 may represent an example in which if angiography had been used on the 1st admission before ventriculography, this and perhaps the 2 subsequent operations with the difficulties described could have been obviated.

We share the opinion that angiography should be used systematically in the work-up of patients, particularly adults, presenting manifestations of a possible expanding lesion of the cerebellar fossa. Besides not altering the intracranial equilibrium, angiography could obviate the necessity for pneumoencephalography and/or ventriculography and supply information not obtainable by other methods of investigation.

Summary

Two cases of so-called solid hemangio-blastomas demonstrated angiographically are presented. One of the patients was 14 years of age. The usefulness of angiography in the management of the diagnostic problems of the expanding lesions of the cerebellar fossa is emphasized.

References