Acta Neurochirurgica 52, 219–224 (1980)

ACTA
NEUROCHIRURGICA

Ankara University Medical Faculty, Department of Neurosurgery,
Ankara, Turkey

Venous Malformation of the Aqueduct of Sylvius Treated by Interventriculostomy: 15 Years Follow-Up

By

N. Avman and **C. Dinçer**

With 4 Figures

Summary

A single venous shunt obstructing the aqueductus of Sylvius was treated by interventriculostomy.

A passage for CSF was obtained between the third and fourth ventricles.

The catheter which was left in the aqueduct collapsed the malformation, possibly preventing a future haemorrhage.

A Simple rubber catheter in the aqueduct of Sylvius has been well tolerated by the patient for 15 years.

Vascular malformations of the periaqueductal region may cause stenosis of the aqueduct [6, 8, 12, 13]. There are only six reported cases of A-venous malformation within the aqueduct in the literature [1, 3–5, 19]. We present a case of venous malformation in the aqueduct causing obstructive hydrocephalus.

Interventriculostomy was performed 15 years ago. The patient is enjoying a normal life. To our knowledge this case is the first one treated by this surgical technique to be reported.

Case Report

G. D. 64/730 I. On 13 March 1964 this 35 years old woman was admitted with a seven years history of headache. This headache was chiefly localized at the vertex, and there was no nausea or vomiting. Her vision had been indistinct for one year.

Physical examination was normal. On neurological examination she was alert and well orientated. She had bilateral papilledema, left central facial paresis, and left hemihypoesthesia. She had dysmetria and dysdiadochokinesia on the left, and ataxic gait disturbances. There was no motor or reflex abnormality.

0001-6268/80/0052/0219/$ 01.20

Skull X-rays revealed mild intracranial hypertension and posterior shift of the calcified pineal body. EEG was normal. Peripheral constriction of visual fields was present. On right brachial angiography hydrocephalus was found. Pneumoventriculography was performed. CSF examination revealed no abnormality. Pressure was 220 mm of water. Ventricles were dilated, and narrowing of the aqueduct (tapered type) could be seen on the films.

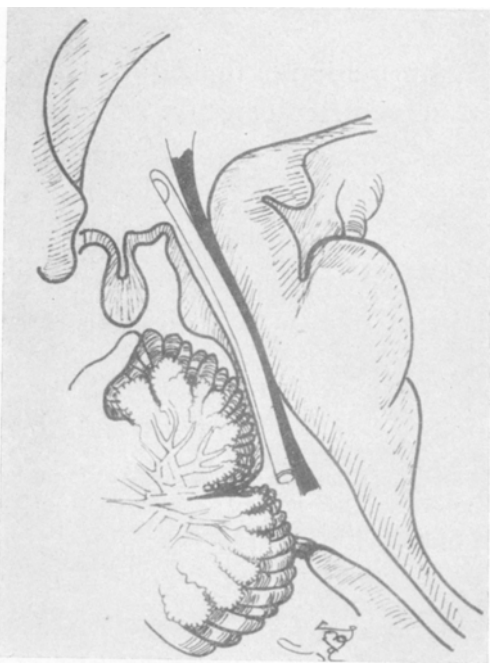


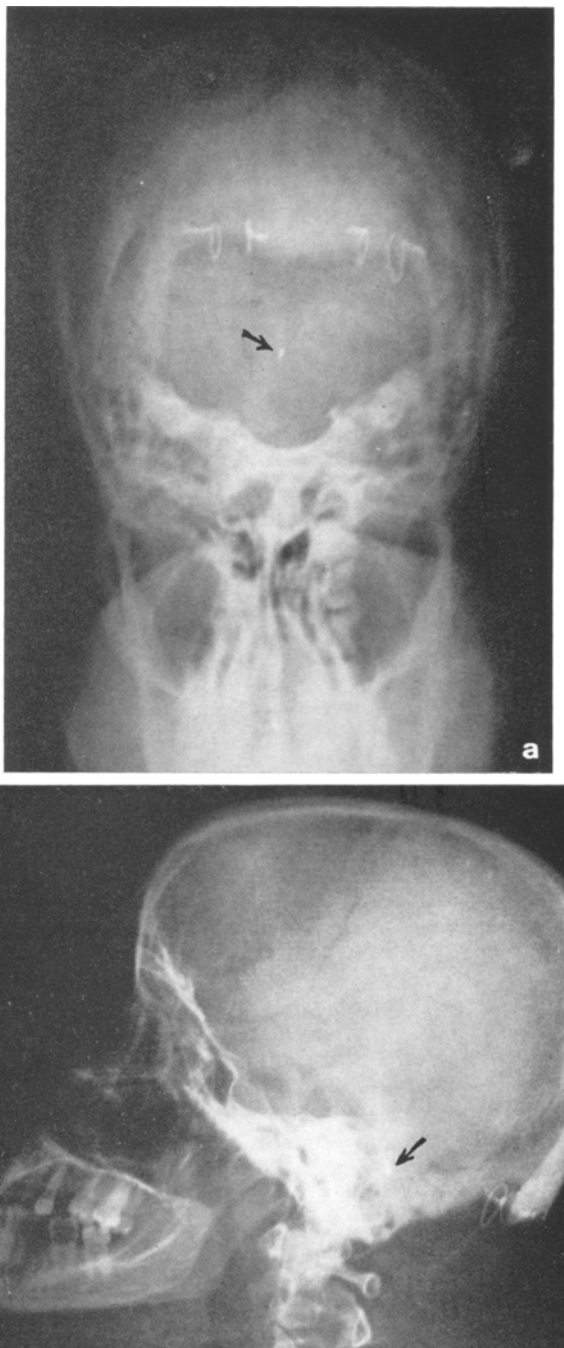
Fig. 1. Schematic diagram of the operation

Operation: Under general anaesthesia posterior fossa exploration was done. There was no pathology in the cerebellum or fourth ventricle. An abnormal venous vascular element that partially blocked the aqueduct was detected. It entered the left lateral recess. The narrowing of the aqueduct was dilated with small plastic catheters, and one of these was left in the aqueduct (Figs. 1, 2, 3, and 4) with a small metallic clip at the end of it.

The patient improved postoperatively, and was discharged two weeks after operation.

Discussion

Angiomas of the posterior fossa often cause increased intracranial pressure. Subarachnoid haemorrhages does not play a dominant role [17]. AVMs within or adjacent to the brain stem mimic other vascular diseases, tumours, and multiple sclerosis, and may produce acute, remitting, and slowly progressive neurological syndromes [11].



Figs. 2 a–b. Postoperative A–P and lateral direct craniograms. Metallic clip (arrow) can be seen

The sex ratio is higher in females [17]. The average age in 64 patients with brain stem angiomas is 49 years [13].

Brain stem angiomas may show the following clinical presentations:

1. Subarachnoid haemorrhage with or without focal brain stem signs. The incidence of SAH in brain stem malformations is 20%.
2. Tix douloureux or hemifacial spasm in a young adult. Tics are seen in 11% of brain stem malformations [7, 10, 17].

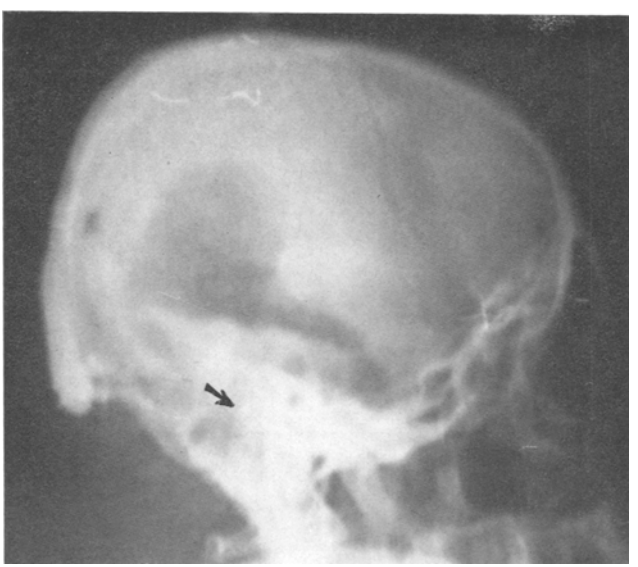


Fig. 3. Postoperative lateral ventriculogram. The arrow indicates the clip

3. Hydrocephalus with or without signs of elevated intracranial pressure. In most cases it has been obstructive but it may occur in the absence of demonstrable obstruction.
4. Progressive posterior fossa signs.
5. Recurrent occipital or hemicranial vascular headache [15, 16, 18].

The headaches of AVMs tend to occur consistently on one side, while those of migraine alternate. The visual accompaniment of AVMs is brief photism during the headache. Photism may also arise from midbrain involvement [14].

6. Remitting multifocal brain stem signs and symptoms.

Lid retraction from periaqueductal involvement (Colliers' sign) may also be misinterpreted as proptosis [2]. Occasionally there is retraction nystagmus also (Sylvian Aqueduct syndrome).

Arteriovenous malformations in the aqueduct are unusual. Alvord [1] reported a patient with hydrocephalus who became symptomatic in his sixth decade. Autopsy demonstrated a large vein coursing through the aqueduct towards the vein of Galen, pinching off an aqueductal diverticulum, which ended blindly [6].

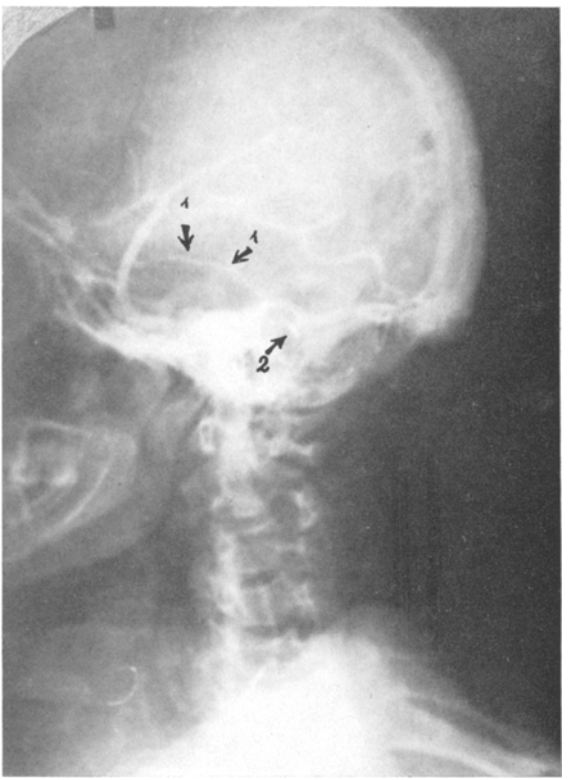


Fig. 4. Right carotid angiogram after operation. The arrows (*1*) show the Vein of Labbe, and (*2*) the metallic clip

Small arteriovenous malformations situated within the aqueduct and fourth ventricle causing obstructive hydrocephalus have been described [3, 5, 9, 19].

In our case there is an abnormal vein in the aqueduct draining towards the left lateral recess of the fourth ventricle.

In these cases of equaductal stenosis correct diagnosis can be obtained by vertebral angiography. In our case a single venous shunt could not be shown by this method.

A normal brain scan does not eliminate the possibility of an arteriovenous malformation in this region [6].

References

1. Alvord, E., Jr., The pathology of hydrocephalus. In: Disorders of the Developing Nervous Systems (Fields, W. S., Desmond, M. M., eds.). Springfield, Ill.: Ch. C Thomas. 1961.
2. Askenasy, H., Wijsenbeck, Herzberger, H., Retraction of eyelids due to arteriovenous aneurysm of midbrain. Arch. Neurol. Psychiatr. *69* (1953), 236—241.
3. Bailey, O. T., The pathology of subarachnoid hemorrhage. In: Pathogenesis and treatment of cerebrovascular disease (Fields, W. S., ed.). Springfield, Ill.: Ch. C Thomas. 1961.
4. Bruce, R. J., Two cases where abnormal vascular elements obstructed outflow from the third ventricle. J. Neurol. Neurosurg. Psychiat. *38* (1975), 824.
5. Courville, C. B., Obstructive internal hydrocephalus incident to a small vascular anomally of the midbrain. Bulletin of Los Ang. Neurol. Soc. *26* (1961), 41—45.
6. De Feo, *et al.*, Aqueductal occlusion by midline arteriovenous malformation. Surg. Neurol. *5* (1976), 59—62.
7. Gardner, W. J., Sara, G. A., Hemifacial spasm. A reversible pathophysiologic state. J. Neurosurg. *19* (1962), 240—247.
8. Graf, C., Angiomatous malformation of the Sylvian aqueduct with remarks on the management of aqueductal obstructions. J. Neuropath. Exp. Neurol. *5* (1946), 45—53.
9. Hoare, R. D., Arteriovenous aneurysm of the posterior fossa. Acta Radiol. *40* (1953), 96—102.
10. Koo, A. H., Ferris, E. J., Shapiro, J. H., Cerebral straight sinus "Jet phenomenon" in arteriovenous malformations. Radiology *94* (1970), 577—579.
11. Lessel, S., *et al.*, Brain stem arteriovenous malformations. Arch. Ophthalmol. *86* (1971), 255—259.
12. Logue, V., Monckton, G., Posterior fossa angiomas. A clinical presentation of nine cases. Brain *77* (1954), 252—273.
13. McCormick, W. F., Hardman, J. K., Boulter, T. R., Vascular malformations (Angiomas) of the brain with specific reference to those occuring in the posterior fossa. J. Neurosurg. *28* (1968), 241—251.
14. Nashold, B. S., Jr., Phosphenes resulting from stimulation of the midbrain in man. Arch. Ophthalmol. *84* (1970), 433—435.
15. Newton, T. H., Weidner, W., Graits, T., Dural arteriovenous malformations in the posterior fossa. Radiology *90* (1968), 27—35.
16. Paterson, J. H., McKissock, W., A clinical survey of intracranial angiomas with special reference to their mode of progression and surgical treatment. A report of 110 cases. Brain *77* (1954), 252—273.
17. Pia, H. N., Gleave, J. R., Grote, E., Cerebral angiomas. Advances in diagnosis and therapy. New York: Springer. 1975.
18. Reuter, S. R., Newton, T. H., Greitz, T., Arteriovenous malformations of the posterior fossa. Radiology *87* (1966), 1080—1088.
19. Rosenheck, C., Venous angioma of the Sylvian aqueduct and fourth ventricle associated with internal hydrocephalus. Arch. Neurol. Psychiat. *38* (1937), 427.

Authors' addresses: Dr. N. Avman, Professor and Chairman of Neurosurgery Department, Ankara University, and Dr. C. Dinçer, Resident in Neurosurgery Department, Ankara University, Ankara, Turkey.