External carotid-transverse sinus fistula

Case report

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A case of congenital fistula between the posterior auricular artery and the transverse sinus is described. A comment is added on the possible embryological mechanism producing these lesions, and the usual symptomatology and treatment are discussed.

Key Words - arteriovenous fistula • posterior auricular artery • transverse sinus

Intracranial arteriovenous fistulas supplied by the external carotid circulation and not by the internal are uncommon. Some of these are acquired (traumatic), and others are spontaneous or congenital. We are reporting another spontaneous case.

Case Report

This 19-month-old baby boy was admitted to the Regional Neurosurgical Center on September 7, 1954. His development had been retarded for some time, and since the age of 12 months he had been dragging his left leg. Six months previously, when 13 months old, he had had a minor fall; his father on picking him up had noticed a pulsatile swelling in the left occipital region. Recently the child had been scratching the overlying skin in that region.

Three days before admission, he had had two epileptic episodes, one involving predominantly the right side of the body, the other the left; he was feverish at that time but no obvious cause for this was found.

Examination. On admission the patient had an upper motor neuron disturbance of the left leg and also to a lesser extent of the left arm. A pulsatile swelling was noted behind the left mastoid process, with a thrill and systolic bruit. The veins over the scalp were also dilated. Examination of the cerebrospinal fluid showed 11 red cells, 2 polymorphs, 1 lymphocyte, 225 mg% of protein, and a positive globulin; sugar and chlorides were normal. Plain skull x-ray films showed a rounded defect in the left occipital bone about \( \frac{1}{2} \) in. in diameter. Differential left percutaneous angiography was performed with and without compression of the opposite internal carotid artery. The internal carotid circulation was normal. In the external circulation, a very large posterior auricular artery was demonstrated running through the skull defect to a \( 5 \times 4 \) cm “aneurysmal sac” draining into an enormously dilated transverse sinus (Fig. 1).

Operation. On September 22, 1954, the common carotid artery was dissected at its bifurcation. It was confirmed by auscultation that occlusion of the external carotid abolished the bruit, and the artery was therefore doubly ligated. The incision was extended until finally the point of entry through the bone defect was identified. The artery was ligated flush with the bone and was then
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traced as far down as possible behind the sternomastoid muscle. It was ligated there, and the intervening segment was excised. Histologically, the excised artery showed moderately uniform hyperplasia of the intima.

Postoperative Course. The patient improved. Repeat angiography was performed. The left internal carotid angiogram was still normal. The site of the external carotid ligation was shown. There was excellent collateral circulation across the circle of Willis from left to right on right carotid compression. This demonstrated both left and right middle and anterior cerebral arteries. The anomaly was no longer filled (Fig. 2). At the time of discharge the baby was starting to walk.

Second Examination. The patient was readmitted on October 20, 1954, because of vomiting, drowsiness, and further epileptic episodes. The bruit over the anomaly had reappeared. Further left carotid angiography was normal. Right carotid and vertebral angiography demonstrated the “aneurysm” filling through branches of the right external carotid and cervical vertebral arteries, and suboccipital arterial plexus. The shape of the anomaly demonstrated on these latest angiograms was different from that on the original films. The lower half had apparently been successfully eliminated by thrombosis after the first operation and only the upper part, incorporated into the transverse sinus, now filled. The intracranial arteries did not supply the anomaly through either the carotid or vertebral systems.

Second Operation. Under anesthetic, the bruit was localized to the area over the anomaly and could be abolished by pressure in the neck. Suboccipital craniectomy was

Fig. 1. Left external carotid angiograms, arterial (left) and venous (right) phases, showing the large posterior auricular artery running through the skull defect to the aneurysmal sac.

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Fig. 2. Postoperative left internal carotid angiogram showing that the aneurysmal sac no longer filled.

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carried out. Several small arteries were found traversing the bone overlaying the lateral sinus. On opening the dura, a hugely dilated occipital sinus was encountered and bled profusely. When this had been brought under control, it was not felt justifiable to carry on and excise the remaining intracranial portion of the anomaly embedded in the transverse sinus.

Second Postoperative Course. The patient's immediate postoperative condition was unchanged but then he gradually started to deteriorate. The posterior fossa decompression became increasingly tense and bulging. A lumbar air encephalogram was performed which showed a communicating hydrocephalus. It was felt that this was secondary to a lateral sinus thrombosis. In an attempt to tide him over, a thecoperitoneal shunt was carried out but did not change his course. The patient died in January, 1955.

Postmortem Examination. The thecoperitoneal tube seemed patent. The immediate cause of death was bronchopneumonia; the underlying cause was hydrocephalus due to lateral sinus thrombosis secondary to the arteriovenous fistula. The anomaly, both transverse sinuses, and the torcular and occipital sinuses were completely blocked by massive antemortem thrombus.

Discussion

This case seems of special interest for several reasons. The age of the baby together with the well-established bone defect and the abnormality of the posterior fossa venous sinuses make a congenital etiology virtually certain. No other patient as young as this has been reported. This suggests the likelihood of an embryological etiology in other spontaneous cases reported at a later age. In the development of the venous sinuses, the primary head vein, which develops from the anterior cardinal vein, is the only structure visible up to the 12 mm stage. The increase in size of the otic capsule results in the formation of a secondary venous channel at 15 mm which becomes the transverse sinus. The inner ear is supplied by the stylomastoid branch of the posterior auricular artery; therefore, close topographical relationships are established between the transverse sinus and external carotid artery at an early period in development.

Another possible explanation is that the vasa vasorum of the transverse sinus, supplied by the meningeal branch of the occipital artery, might develop anomalously. If the vasa vasorum of all the venous sinuses derive from the external carotid artery, this could explain why fistulas apparently do not occur in the region of the inner ear.

The clinical picture in this baby was frankly malignant. Only one other case has been reported in which the neurological findings were alarming. In the other patients the symptoms were chiefly headache and bruit.

The poor response to treatment was rather surprising since, at the first operation, the normally satisfactory criteria both of ligating the feeding vessel and of excising the fistulous communication were fulfilled.

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References


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