Multiple arteriovenous malformations involving the scalp, dura, retina, cerebrum, and posterior fossa

Case report

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A case is presented of multiple arteriovenous malformations involving the scalp, dura, retina, cerebrum, and posterior fossa in one patient who also had an internal carotid aneurysm. Cobalt irradiation was followed by symptomatic improvement.

Key Words arteriovenous malformations, multiple aneurysm

Multiple intracranial arteriovenous malformations occurring in one patient are extremely rare. We are reporting a case of arteriovenous malformations involving the scalp, dura, retina, cerebrum and posterior fossa in one patient.

Case Report

A 19-year-old woman was admitted to our clinic on April 15, 1968, because of staggering gait and blindness in the right eye. It had been noted at birth that she had a dilated vein in the right cheek and chin. At 11 years of age she began to complain of visual disturbance in her right eye, accompanied at age 13 by difficulty in writing and walking due to tremor and ataxia, and developed complete loss of sight by 14 years of age. During the 3 years before admission she noticed a slow progression of these symptoms and on admission was unable to walk without help. She had suffered no epileptic seizures or loss of hearing during childhood, although occipital headache sometimes occurred. The family history was without significance.

Examination. The patient was a small, mentally retarded, 19-year-old female. She had a mild right exophthalmos and an angiomatous right palpebral conjunctiva. A markedly dilated vein was present on the right side of her face and neck (Fig. 1). Auscultation of the skull disclosed a loud bruit over all parts of the head, synchronous with the heart beat, with maximum intensity over the neck and occipital region. Neurological examination revealed complete loss of sight in the right eye, and anisocoria. The right pupil was larger than the left and did not react to the light. Strabismus, divergence, and ophthalmoparesis of the internal rectal muscle of the right eye were noted. Other cranial nerves were normal. There were bilateral cerebellar signs (Romberg sign, adiadochokinesia, abnormal finger-to-finger test, and muscular hypotonia), which were more marked on the right side. Lumbar puncture revealed a pressure of 130 mm of water with no cells, and normal Queckenstedt's sign. The hematocrit was 37.7%, hemoglobin 12.0 gm%, and a red blood cell
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Fig. 1. Right epe shows a mild exophthalmos due to arteriovenous malformation of the palpebral conjunctiva. Note the markedly dilated vein on the right side of the face and neck.

Fig. 2. Lateral projection of right selective internal carotid arteriogram, showing the dilated internal carotid artery, pea-sized aneurysm (arrow) and arteriovenous malformation.

Fig. 3. Anteroposterior projection, showing a large arteriovenous malformation and the dilated middle cerebral artery.

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count was 4.2 million per cu mm. Other routine tests were within normal limits. An electroencephalogram disclosed right-sided cerebral dysfunction. Plain x-ray films of the skull revealed the widening of the groove of the middle meningeal artery which extended to the parietal and the occipital region, and bony bulging in the parietal region.

Right selective internal carotid arteriogram in the lateral projection (Fig. 2) showed a dilated internal carotid artery, a pea-sized aneurysm just before the artery entered the cranium, a large arteriovenous malformation fed by the lenticulostriate arteries, and a markedly dilated and tortuous posterior cerebral artery extending from the syphon to the temporal occipital and parietal region which drains into the great vein of Galen. The arteriogram in the anteroposterior projection (Fig. 3) revealed a large arteriovenous malformation in the region of the basal ganglia and the dilated middle cerebral artery; the anterior cerebral artery could not be visualized. Selective external carotid arteriograms (Fig. 4) demonstrated
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The following findings. The internal and external maxillary arteries whose peripheral branches formed the arteriovenous malformations in the orbit and its surroundings were dilated and tortuous. The posterior branches of the hypertrophied middle meningeal artery formed the arteriovenous malformation, and consequently the malformation situated in the deep cerebrum was visualized poorly. In the fundus of the right eye (Fig. 5) there was a markedly dilated and tortuous arteriovenous malformation obliterating the optic disc and extending to the peripheral area of the retina. Selective vertebral arteriogram (Fig. 6) demonstrated the abnormally meandering course of the dilated vertebral artery and an arteriovenous malformation which occupied almost the entire posterior cranial fossa.

Subsequent Course. A cerebellar ataxia developed gradually after admission and resulted in inability to walk 3 months after admission. Cobalt irradiation was started on July 29, 1968. A total of 3450 R to the supratentorial region, and 2300 R to the arteriovenous malformation of the posterior fossa was followed by symptomatic improvement as manifested by improved cerebellar ataxia and amelioration of headache.

Comments

The simultaneous occurrence of the cerebral aneurysms and arteriovenous malformations is relatively rare, although they may have a common congenital basis. Multiple arteriovenous malformations are also extremely rare in one patient. Only a few cases with multiple malformations have been reported. It is generally agreed that arteriovenous malformations arise from a develop-
mental defect, a failure of persistence of the primitive capillary bed, and that intracranial aneurysms develop on the basis of a congenital vascular deficiency. According to Paterson and McKissock, the increased blood flow due to an arteriovenous malformation might throw abnormal stress on the cerebral vessels concerned and predispose them to the formation of aneurysms. In our case, however, the aneurysm developed on a different arterial system.

There has been little in the literature as to the genesis of multiple arteriovenous malformations; maybe they arise from multiple developmental defects or a multiple failure of persistence of the primitive capillary beds. It is now widely agreed that excision is the most effective treatment for arteriovenous malformations, since only a few patients have shown significant improvement after radiotherapy or medication. However, Potter pleaded a case for radiotherapy, pointing out that several of the patients with the longest survivals in his series had been treated with radium, although the efficacy of such treatment has been denied by Olivecrona and Riives. According to Pool, 44% of 220 patients who received medical and x-ray treatment showed good results; the comparable figure for 187 patients whose lesions were excised was 74%. X-ray therapy seems advisable whenever surgery cannot be done with reasonable safety.

References

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