Ruptured aneurysm of the anterior spinal artery associated with pseudoxanthoma elasticum

Case report

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A ruptured aneurysm of the anterior spinal artery associated with pseudoxanthoma elasticum is reported. This aneurysm was demonstrated by selective spinal angiography.

KEY WORDS • aneurysm • anterior spinal artery • pseudoxanthoma elasticum

The occurrence of an aneurysm of the anterior spinal artery is extremely rare; no more than a few cases have been documented. Only two of these cases were demonstrated by angiography. We present a patient with a ruptured aneurysm of the anterior spinal artery associated with pseudoxanthoma elasticum (PXE). The aneurysm was revealed by selective spinal angiography.

Case Report

This 37-year-old woman was admitted to Shimodate Municipal Hospital because of the sudden onset of a severe headache and low-back ache. She had previously been in excellent health. Her past history was negative except that she had had a skin eruption since 20 years old. Her family history revealed that her sister had similar eruptions in the neck and hypertension but received no medical treatment.

Examination. Blood pressure was 240/140 mm Hg. Yellowish papules or reticulated plaques were noted symmetrically in the neck and axillae. Neurological examination revealed hypesthesia to pain and light touch in the right lower extremity. There was urinary retention. In addition, signs of meningeal irritation were present. Examination of the optic fundi showed angioid streaks in both eyes, more marked on the right side. A lumbar puncture revealed bloody cerebrospinal fluid with an opening pressure of 90 mm H₂O. A chest x-ray film showed slight cardiomegaly.

Skull x-ray films were normal, and bilateral carotid angiograms showed no intracranial aneurysm or arteriovenous malformation. Bilateral retrograde brachial angiograms revealed elongation of the basilar artery, but no vascular anomaly. Routine laboratory studies including tests for syphilis were negative.

Since persistence of the low-back ache, urinary retention, and hypesthesia of the right lower limb suggested a vascular disorder of the spinal cord, spinal angiography via the right femoral artery was performed. Abdominal aortography revealed marked stenosis of the abdominal aorta and femoral arteries. Development of an anastomotic circulation in the paravertebral region was demonstrated at T10–L2 (Fig. 1). Dilatation of both internal thoracic arteries was also noted. In addition, an aneurysm of the anterior spinal artery was disclosed at T-10 in selective left T-9 intercostal angiography (Fig. 2).

Course. Biopsy specimens of the skin lesions were diagnosed at the Department of Dermatology of Tokyo Women's Medical College as PXE. High blood pressure persisted in spite of various kinds of antihypertensive drugs. A radioisotopic renogram showed hypofunction of the left kidney, and the level of plasma renin activity was slightly increased. The patient's neurological condition gradually recovered. No surgical therapy for the aneurysm was performed, but she was transferred to Tokyo Women's Medical College for treatment of stenosis of the abdominal aorta.
Anterior spinal artery aneurysm

FIG. 1. Aortography showing marked stenosis of the abdominal aorta (arrows) and collateral circulation in the paravertebral area.

Discussion

Aneurysms arising from the anterior spinal artery are extremely rare. Only a few cases have been documented in the literature. It is well known that in cases of coarctation of the aorta the anterior spinal artery is animportant collateral pathway, and this artery becomes elongated and dilated. Thus, an aneurysm may arise from the tortuous anterior spinal artery in association with coarctation of the aorta.

Pseudoxanthoma elasticum is a rare hereditary disease characterized by systemic degeneration of elastic fibers, and often accompanies various vascular lesions and premature arteriosclerosis. Vascular complications of the central nervous system in cases of PXE include cerebral aneurysm or aneurysmal dilatation with or without subarachnoid hemorrhage, occlusion or stenosis of the internal carotid artery (ICA) or vertebral artery, and carotid rete mirabile with carotid-cavernous fistula. Of these complications, cerebral aneurysms are the most common. Nearly all of these aneurysms arise from the ICA, and are often bilateral or calcified. Ischemic cervical myelopathy associated with PXE has also been reported, but an anterior spinal artery aneurysm has not previously been documented. This is the first report of such a case.

The marked stenosis of the abdominal aorta seen in this case is a rare vascular lesion, since the aorta is uncommonly involved in cases of PXE. Stenosis of the abdominal aorta probably caused the anterior spinal artery to become markedly dilated because of its importance as a collateral pathway, as in cases of coarctation of the aorta. Presumably, the aneurysm developed in combination with degeneration of elastic fibers of the artery, hemodynamic changes that occurred naturally at the hair-pin bend of the tortuous anterior spinal artery, and severe hypertension.

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