Case Reports and Technical Note

Coarctation of the Aorta and Cerebral Aneurysm
Report of Two Cases

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Eppinger in 1871 is credited with first describing the association of coarctation of the aorta and cerebral aneurysm. Later Hamby in 1952 said that the combination was common. Pool and Potts in 15 do not mention the matter in their book on cerebral aneurysms. We are reporting two cases.

Case Reports

Case 1. This 35-year-old man was transferred to the Neurosurgical Unit on February 25, 1966, from another hospital to which he had been admitted on February 22. While on the lavatory he had developed severe headache and collapsed to the floor without losing consciousness. A lumbar puncture had shown a blood-stained cerebrospinal fluid. There had been no previous episodes, but in the last 2 months he had had frequent headaches and had felt tired and generally irritable.

Originally he denied any previous illness, but his wife remembered that at the ages of 20 and 21 years he had been examined for military service and had been declined on account of high blood pressure. Two years later he had been examined for a life insurance policy and had been passed as fully fit.

Examination. He was a small wiry man who did not have any abnormal neurological signs. There was mild arteriovenous nipping of the retinal vessels. The blood pressure in the arms was 200/100 mm Hg, in the thigh, 150/100 mm Hg. The femoral pulses were feeble and delayed.Bounding pulsations could be felt from the periscapular arteries. A systolic bruit could be heard all over the precordium.

Radiographs of the chest showed rib notching (Fig. 1). The heart shadow was not enlarged. An aortogram done through the right femoral artery demonstrated a coarctation of the aorta 3 cm distal to the left subclavian artery. The catheter passed through the coarctation but it was impossible to get enough contrast medium into the cerebral arteries for diagnostic purposes. Tracings taken from the aortic catheter gave a pressure of 180/70 mm Hg above the lesion and 100/75 mm Hg below it.

Three days later bilateral carotid arteriograms were done. The right external carotid artery was absent, and its branches arose from the internal carotid artery. The right cerebral artery did not fill from the right internal carotid artery. The left carotid arteriogram demonstrated a lobulated aneurysm 1 cm in diameter arising from the...
posterior aspect of the anterior communicating artery (Fig. 2). Both anterior cerebral arteries filled from the left side.

The aneurysm was considered to be an unsatisfactory lesion to treat. It was uncertain whether the defect in the circle of Willis was due to spasm or to arterial anomaly. On April 4, the coarctation was resected. The immediate postoperative course was normal and he was discharged home on April 14.

**Course.** He was readmitted to Dunedin on May 17 for reassessment. Observations of the blood pressure in the arms gave readings between 170 to 140 mm Hg systolic and 100 to 90 mm Hg diastolic. The carotid arteriograms were repeated and showed no change from those done in March. It was concluded that the deficiency of the anterior cerebral circulation was developmental and not due to spasm. It was decided to continue the conservative management of the cerebral aneurysm.

He was seen on August 2, 1966. He had returned to work and was doing a full day’s work without complaint. He was taking reserpine 5 mg twice a day because the blood pressure was still elevated. A casual blood pressure taken from the right arm was 170/90 mm Hg.

**Comment.** This patient had a common developmental anomaly of the circle of Willis and another known anomaly of the internal carotid artery in the neck. As might be expected in a patient his age, resection of the coarctation of the aorta did not completely control the hypertension.

**Case 2.** This 19-year-old bricklayer was admitted to the Neurosurgical Unit on June 15, 1966, from Grey Hospital, Greymouth, by air ambulance. He had been admitted to that hospital on June 10 after having developed a severe headache while sitting in a motor car. Until this illness he had done hard physical labor and had indulged in strenuous sports with ease. He was found to have an elevated blood pressure and signs of coarctation of the aorta. A lumbar puncture produced heavily blood-stained cerebrospinal fluid.

**Examination.** There were no abnormal neurological signs, and the retinal vessels were normal. The blood pressure in the right arm was 150/100 mm Hg and in the left thigh was 130/100 mm Hg. The right femoral pulse was absent, and the left was feeble and delayed. There was marked pulsation from the arteries around the scapula.

Radiographs of the chest showed notching of the ribs. The heart size was normal. Bilateral carotid arteriograms were normal on the right side, but on the left showed a large aneurysm at the bifurcation of the internal carotid artery. There was a second aneurysm of medium size arising from the junction of the left anterior cerebral artery and the anterior communicating artery (Fig. 3).

**Course.** It was considered that the cerebral aneurysms were not suitable for surgical treatment. Suddenly on the evening of June 22 he had another subarachnoid hemorrhage and died. Autopsy permission was refused.

Although coarctation of the aorta was not proved by surgery or autopsy, the physical signs were so characteristic as to make any other diagnosis unlikely.

**Discussion**

Coarctation of the aorta of the adult type is one of the less common congenital abnormalities of the heart and great vessels. The incidence in general autopsy studies has varied from 1 in 2500 to 1 in 1176. Maude Abbott in 1928 delineated the syndrome of coarctation of the aorta. There was a 4:1 ratio of male to female, 74% died before the
age of 40 years, and a few lived to a ripe old age. Death was due to cerebral hemorrhage in 12.5% of the cases. Later Schwartz and Baronofsky\textsuperscript{17} found 31 autopsied cases of cerebral aneurysm and coarctation of the aorta. The highest incidence of aneurysm rupture occurred between the ages of 20 and 40 years; 30% of these patients had multiple cerebral aneurysms. Walton\textsuperscript{20} and Stehbens\textsuperscript{19} found an even higher incidence of multiple cerebral aneurysms associated with coarctation. The incidence of multiple cerebral aneurysms in ordinary subarachnoid hemorrhage uncomplicated by coarctation is about 10%.\textsuperscript{12}

Our experience with subarachnoid hemorrhage due to cerebral aneurysm shows that associated coarctation of the aorta is rare. From 1943 to 1966, among 545 patients with proven cerebral aneurysms, only two patients had coarctation. Walton\textsuperscript{20} had one coarctation in 312 subarachnoid hemorrhages, and Du Boulay\textsuperscript{6} had two coarctations in 197 patients with cerebral aneurysm. In view of the lethal nature of the combination of simple subarachnoid hemorrhage (mortality 54%) and coarctation of the aorta (mortality 86%) plus the relative rarity of aortic coarctation, it is not surprising that neurosurgeons are seldom involved in this problem.

Most patients have received surgical treatment for one of the lesions. Schwartz and Baronofsky\textsuperscript{17} found two patients who had had the coarctation resected after subarachnoid hemorrhage; one of these died. Another patient had both conditions successfully treated surgically but in two stages, 3 years apart. Signier, \textit{et al.},\textsuperscript{18} reviewed 10 cases where carotid arteriography had been done for cerebral hemorrhage and coarctation. Three did not have cerebral aneurysms, but one died later and was found to have an aneurysm. In this series three had no surgery, one had an intracranial clot evacuated, four had the coarctation resected without treatment of the aneurysm, and two had aneurysms operated upon without treatment of the coarctation. Haxton\textsuperscript{9} resected the coarctation of a 21-year-old female with an angiographically proven basilar aneurysm; she survived 11 years that included a preg-
nancy without incident. Matson had 14 patients with cerebral aneurysm in childhood, of whom three had coarctation of the aorta; in two of these it was possible to treat both lesions successfully.

Total cerebral angiography is essential preoperatively because of the high incidence of multiple lesions. Solitary cerebral aneurysms may be treated surgically although there must be caution in dealing with anterior communicating aneurysms associated with hypertension. Our experience is in accord with that of McKissock, et al., hence our conservative approach in Case 1. Any definitive treatment of the cerebral aneurysm should be soon followed by resection of the aortic lesion where the hazards are much less. When the cerebral lesion is considered inoperable, aortic surgery may be worthwhile, although as an alternative, antihypertensive treatment with drugs may be successful.

The outlook for coarctation of the aorta has been revolutionized by surgery, although death from cerebrovascular accidents may occur later. Rumel, et al., had one death from cerebral hemorrhage out of 1539 survivors of surgically treated coarctation. Efskind and Sanderud had one death from subarachnoid hemorrhage in 130 patients surviving up to 12 years after aortic surgery. Haxton operated on an 8-year-old patient for coarctation; the child died 5 years later of ruptured cerebral aneurysm. It is probable that the surgical correction of coarctation of the aorta at its optimum age, between 8 and 20 years, exercises a beneficial effect on the cerebral vessels, and later hemorrhages are uncommon. To have waited until the coarctation of the aorta has shown itself by cerebral hemorrhage is to have waited too long.

Subarachnoid hemorrhage may originate from a ruptured aneurysm of the spinal artery associated with coarctation of the aorta. Wyburn-Mason reviewed four such cases and Blackwood a single case.

It is often held that any patient with a congenital lesion is liable to have another. Stehbens showed that patients with cerebral aneurysms are no more likely to have other congenital lesions than are general patients. He and Walton thought that the association of coarctation and cerebral aneurysm was due to hypertension. Occurrence of cerebral aneurysm in childhood, however, makes it clear that defects in the arterial wall do occur at this age, some of which may be congenital.

Summary

We have described two cases in which coarctation of the aorta and cerebral aneurysm occurred in the same patient, and have reviewed related reports.

References

1. ABBOTT, M. E. Coarctation of the aorta of the adult type. II. A statistical study and historical retrospect of 200 recorded cases, with autopsy, of stenosis or obliteration of the descending arch in subjects above the age of two years. Am. Heart J., 1928, 3:392-421, 574-618.


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