Giant intracranial aneurysm associated with Marfan's syndrome

Case report

Divisions of Neurosurgery and Neuroradiology, University of Utah Medical Center, Salt Lake City, Utah

The authors describe a giant intracranial internal carotid aneurysm in a patient with Marfan's syndrome. Treatment consisted of internal carotid ligation with subsequent thrombosis of the lesion. The patient's course was complicated by carotid arterial dissection and possible laminar thrombosis within the aneurysm increasing the size of the lesion. Three clinical syndromes, Marfan's, Ehlers-Danlos, and pseudo-xanthoma elasticum, are discussed. The literature is reviewed with reference to the hazards of treatment of vascular pathology in patients with these connective tissue disorders.

Key Words • carotid artery • carotid artery thrombosis • Marfan's syndrome • aneurysm, cerebral • cerebral angiography • connective tissue disease

The treatment of giant intracranial aneurysms is difficult, and the problem is compounded by friability of the vessel wall in patients with connective tissue disorders. We are reporting a case of Marfan's syndrome in which an indirect approach for treatment of intracranial aneurysm produced near-fatal complications.

Case Report

This 38-year-old woman was admitted on April 18, 1974, with a chief complaint of progressive headache for 2 months. The headache was dull, continuous, and located behind the left ear; it was worsened by movement of her head. Occasionally it became burning in character and radiated to the top of her head. When the pain was severe, she experienced much nausea and vomiting, and noticed increasing numbness of the left side of her face. Her past history included a diagnosis of Marfan's syndrome made on the basis of her stature, arachnodactyly, pectus excavatum, and chronically dislocating elbow and knee. In 1956, an ophthalmologist performed recession and resection on the right eye for congenital exotropia. Because of persistence of the defect, in 1970 a marginal tenotomy of the lateral rectus was performed with 5-mm resection of the medial rectus of the right eye and a medial and lateral rectus resection of the left eye.
Aneurysm in Marfan's syndrome

Examination. Neurological examination was unremarkable, except for an intermittent left pupillary enlargement noted by one observer. Interpretation of the ocular muscle examination was difficult because of the previous surgery, but there was no evidence of third, fourth, or sixth cranial nerve paresis. A brain scan showed a highly vascular, solitary lesion in the vicinity of the left sphenoid wing. Cerebral angiography on April 24, 1974, revealed a 3-cm aneurysm involving the cavernous portion of the left internal carotid artery (Fig. 1). There was good cross-filling of the left anterior and middle cerebral arteries from the right carotid circulation and forward collateral flow via the circle of Willis was demonstrated. The vertebral arteries were dilated and tortuous, and there was aneurysmal dilation of the right internal mammary artery (Fig. 2). The arch and proximal great vessels were normal in appearance. The patient was discharged to be readmitted to the Neurosurgery Service on May 13, 1974.

Operation. On May 14, a Salibi clamp was placed on the left internal carotid artery just above the common carotid bifurcation. Occlusion was begun on May 15 and completed on May 17, during which time the patient was monitored with frequent neurological examinations as well as ophthalmodynamometry as an indicator of the left internal carotid thrombosis. The patient tolerated the procedure well until 4 hours after complete occlusion when she suddenly developed a fixed, dilated left pupil, left lid lag, and a left-sided headache, but without alteration of mental function. Shortly thereafter, she complained severely of chest and back pain and...
began vomiting. Lumbar puncture showed a pressure of 130 mm H₂O and clear, colorless fluid. A chest film, electrocardiogram, and serum enzymes were normal. Angiography was not performed. Her symptoms abated over the next 48 hours and she was discharged on May 23, with only mild occasional left-sided headaches and a persistent dilated left pupil. The reason for her chest and back pain was obscure and the pain resolved over a few days time. The headache and third nerve dysfunction were considered to be due to further enlargement of the cerebral aneurysm secondary to laminar thrombosis, a phenomenon in aneurysms with a large volume orifice. 2,3,12,16,17,20,22–24

Postoperative Course. On June 4, 1974, the patient was readmitted for repeat angiography to determine the effect of the carotid clamping. Arch injection showed complete occlusion of the left subclavian artery 3 cm distal to the takeoff of the left vertebral artery (Fig. 3). The left carotid clamp totally occluded flow into the head from the left carotid artery, but below the clamp there was dissection of the left carotid extending down to the arch. There was also dissection involving the innominate, right subclavian, and right carotid artery. The right vertebral artery filled in a retrograde fashion and flowed into the right subclavian artery. The major flow to the head was through the grossly enlarged left vertebral artery. Right carotid injection revealed filling of the left middle cerebral artery via the anterior communicating artery via the anterior communicating artery (Fig. 4). No filling of the aneurysm occurred. The patient was seen frequently thereafter, and by June, 1975, she was asymptomatic except for a mild residual left third nerve paresis.
Aneurysm in Marfan’s syndrome

Fig. 4. Postoperative angiography by right common carotid injection reveals dense opacification of the left middle cerebral artery, indicating that the principal collateral pathway to the left middle cerebral artery following left carotid occlusion is the anterior communicating artery from the right anterior circulation.

Discussion

Marfan’s syndrome was first described in 1896. It is an undefined fundamental defect of connective tissue, most likely related to the elastic fiber. A simple mendelian autosomal dominant with a relatively high grade of penetrance, it is found throughout the world in equal sex distribution. Hallmarks of the disorder are aortic dilatation, dissecting aortic aneurysm, ectopia lentis, arachnodactyly, loose jointedness, and pectus excavatum. Vascular pathology consists of early Erdheim’s cystic medial necrosis followed by fragmentation and sparsity of elastic fibers and increased collagenous tissue in vascular cystic spaces. The media is the only pathological area of the vessels. McKusick stresses the frequency of many undiagnosed cases of this disorder because of its variable penetrance.

Ehlers-Danlos syndrome was described as early as 1682 by a Dutch surgeon. His patient was a Spaniard who could pull his right pectoral skin to his left ear. In the history of medicine these are the “India rubber men” and much has been written about the extraordinary dilatability of the skin. The specific abnormality is not clear, but is most likely a defect in the normal “basket weave” pattern of collagen fibers. This is a rather rare disorder found almost exclusively in persons of eastern European ancestry. It is characterized by dissecting aneurysms of the aorta, loose jointedness, subcutaneous hemorrhages, molluscoid pseudotumors, and fragile, brittle skin. Sutures do not hold well in the skin and dehiscence of surgical wounds is a frequent problem.

Pseudoxanthoma elasticum, described in 1886, is an autosomal recessive connective tissue disorder of collagen. Its distribution is almost worldwide. Its features include thick skin with elevated yellowish areas at the normal skin creases, angioid streaks of the optic fundus, dilatation of the aorta, premature medial calcification of the peripheral arteries, and gastrointestinal hemorrhages.

Dissecting aortic aneurysms in patients with Marfan’s syndrome present a difficult surgical problem. A search of the literature reveals only three cases of aneurysms in vessels other than the thoracic or abdominal aorta. Hardin reported a case of extracranial carotid aneurysm. He successfully resected it and performed an end-to-end anastomosis without incident.

Ehlers-Danlos syndrome has been associated with multiple intracranial aneurysms, spontaneous rupture of arteries, and spontaneous carotid cavernous fistulas. Rubinstein and Cohen described a case of a 47-year-old woman with a subarachnoid hemorrhage. Attempted ligation of a posterior cerebral aneurysm was frustrated by the extremely flimsy vessel “virtually falling to pieces when touched,” and ligation of the subclavian artery had to be performed. The patient did not recover and an autopsy revealed multiple aneurysms of the cerebral circulation.

Two cases of spontaneous carotid cavernous fistula in patients with Ehlers-Danlos syndrome were described by Graf in 1965. Angiography of both patients showed dilated
H. L. Finney, T. S. Roberts and R. E. Anderson
ectatic redundant internal carotid arteries. One was treated with carotid ligation, but later died of rupture of the anterior left ventricular wall. The second case was treated effectively by common carotid ligation; the patient's sibling had been treated successfully elsewhere with carotid ligation for the same lesion.

In 1966, Schoolman and Kepes reported a case of bilateral spontaneous carotid cavernous fistula in a patient with Ehlers-Danlos syndrome. Ligations of the left internal and external carotid arteries proved the fragility of the vessels when the superior thyroid artery inadvertently separated from the external carotid artery. The patient later developed a contralateral carotid sinus fistula and died during an angiogram. Schoolman and Kepes warned against "forme fruste" cases of Ehlers-Danlos syndrome; they and others suggested that some cases of "spontaneous" carotid cavernous fistula could be abortive forms of this disorder.

Scheie and Hogan reported 10 cases of pseudoxanthoma elasticum in 1957. In a survey of the arterial system for evidence of vascular disease, they found an internal carotid aneurysm at the parasellar region in one case and bilateral common carotid aneurysm in another. All three pathological vessels showed calcification of the aneurysm wall. Clinical evidence of peripheral arterial disease was present in all patients.

A case of angiod streaks of the optic fundus in pseudoxanthoma elasticum and aneurysm of the internal carotid artery was reported by Dixon. He noted that the occurrence of intracerebral aneurysm in a patient with this disorder "should not be surprising in view of the known generalized elastic tissue defect involving the large vessels." The patient underwent internal carotid ligation in the neck followed by craniotomy and direct clipping with good results. Dixon cautioned that "The vascular components of pseudoxanthoma elasticum with angiod streaks should be kept in mind." It was pointed out by Scheie and Hogan that "it is possible for vascular lesions to be its only manifestation."

The present case of giant intracranial aneurysm in a patient with Marfan's syndrome was complicated by dissection of the left common carotid, innominate, right subclavian, and right common carotid arteries. Clinical onset of dissection began 4 hours after complete occlusion of the left internal carotid artery with a Salibi clamp and was manifested by back and chest pain, nausea and vomiting, and diaphoresis. A disruption or weakening of the wall had resulted from manipulation of the artery during surgical placement or from the catheter tip during previous angiography. Extreme care was essential during surgery to prevent damage to the fragile vessel. Excessive catheter manipulation and high-pressure injections during angiography should be avoided in such cases.

Summary

A patient with Marfan's syndrome and a giant intracranial aneurysm of the internal carotid artery was treated with extracranial carotid occlusion using a Salibi clamp. The patient developed a third nerve paralysis and severe chest and back pain 72 hours after occlusion. Follow-up angiography showed thrombosis of the aneurysm and dissection of the thoracic great vessels. The third nerve paralysis was believed to be secondary to laminar thrombotic enlargement of the aneurysm, and the vascular dissection was believed to be subsequent to either intimal tear from the clamp, manipulation during the clamp placement, or trauma from the catheter tip from the previous angiogram. Experience with vascular pathology in connective tissue disease states is reviewed with this as the first reported case of its kind.

Acknowledgment

The authors wish to thank Mrs. Pamela Foote for assistance with preparation of the manuscript.

References

1. Bettelheim H: [The practical importance of ophthalmodynamometry and ophthalmodynamography.] Ther Umsch 29:177–188, 1972 (Ger; English abstract)
Aneurysm in Marfan's syndrome


Address reprint requests to: H. Lee Finney, M.D., Division of Neurosurgery, University of Utah Medical School, Salt Lake City, Utah 84112.