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# Dissecting Basilar Artery Aneurysm in Marfan Syndrome: Case Report

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We describe a 35-year-old man with Marfan syndrome who died as a result of rupture of a basilar artery dissecting aneurysm. To the best of our knowledge, this association has not been presented before.

## Case Report

A 35-year-old man with Marfan syndrome presented to the emergency room with a severe occipital headache of several hours duration. The headache differed from his usual migraine headaches both in quality and in unresponsiveness to Cafergot.

The patient's medical history included repair of an ascending aortic aneurysm and replacement of a bicuspid aortic valve 2½ years previously. The patient was receiving chronic anticoagulant therapy.

A noncontrast enhanced CT of the brain obtained on admission was unremarkable. Concurrent lumbar puncture yielded CSF with 628 RBC/mm<sup>3</sup>, 2 WBC/mm<sup>3</sup>, and subsequently negative Gram stain and cultures. Although a traumatic lumbar puncture was a consideration, a cerebral arteriogram was performed because of clinical suspicion of a vascular lesion. The arteriogram showed a fusiform aneurysm of the basilar artery extending into the left superior cerebellar artery and the left posterior cerebral artery (Figs. 1A and 1B).

The patient became briefly unresponsive the day after admission. A repeat lumbar puncture revealed grossly bloody CSF with 2,730,000 RBC/mm<sup>3</sup> and 10,700 WBC/mm<sup>3</sup>.

Anticoagulant therapy was discontinued. The subsequent unenhanced CT scan of the brain demonstrated subarachnoid blood in the basal cisterns. The patient died on the 14th day of hospitalization.

Postmortem examination of the brain confirmed a basilar artery and contiguous vessel aneurysm. Subarachnoid hemorrhage was present around the brainstem and rostral spinal cord. Histologic sections showed an intimal tear in the basilar artery with dissection and transmural rupture. Most of the dissection plane was within the media of the basilar artery (Fig. 1C). Sections of the basilar (Fig. 1D) and systemic arteries showed multiple foci of cystic medial necrosis, consistent with Marfan syndrome.

## Discussion

Marfan syndrome is an autosomal dominant inherited disorder of connective tissue, probably due to an error in protein

metabolism, possibly in collagen or elastin. In its classic form, the syndrome is associated with abnormalities of the eye (ectopia lentis), aorta, and skeleton. The expressivity of the classic Marfan gene varies widely. The rate of occurrence of de novo mutations is estimated at 15% of all cases [1].

The cardiovascular abnormalities in Marfan syndrome include dilatation of the aortic ring, dilatation of the ascending aorta, and dissection of the aorta. Coarctation of the aorta has also been described. Extraaortic manifestations include dilatation and/or dissecting aneurysms of the pulmonary arteries and cardiac valvular and septal defects [1].

Other extraaortic vascular involvement is rare. Austin and Schaefer [2] reported a case of innominate artery and bilateral carotid artery dissection without aortic dissection in a patient with Marfan syndrome. Tortuosity and elongation of the basilar artery have been previously noted in Marfan syndrome but are rare [3, 4]. To the best of our knowledge basilar artery dissection and rupture have not been reported before in this syndrome.

Dissecting aneurysms of the intracranial arteries are uncommon. Intracranial dissecting aneurysms have a high morbidity and mortality rate [5] in contrast to the usual benign course of cervical carotid artery dissections [6]. Dissections involving the intracranial portion of the carotid artery and middle cerebral artery typically cause infarction, but posterior intracranial artery dissections have less predictable sequelae. Vertebrobasilar dissections have two major clinical presentations, depending on the arterial plane of dissection: ischemia and subarachnoid hemorrhage [7]. If the dissection plane is between the intima and media, vessel occlusion and infarction occur. In our case, the dissection extended transmurally, with the greatest length of dissection within the vessel media. Preexistent changes of collagen necrosis were present in this layer.

Intracranial arterial dissections occur most often in young adults (late 20s to early 40s) [5], with no predisposing condition identified in most cases [7]. Isolated cases related to fibromuscular dysplasia, cystic medial necrosis, arteriosclerosis, syphilis, migraine, homocystinuria, and trauma have been reported [5].

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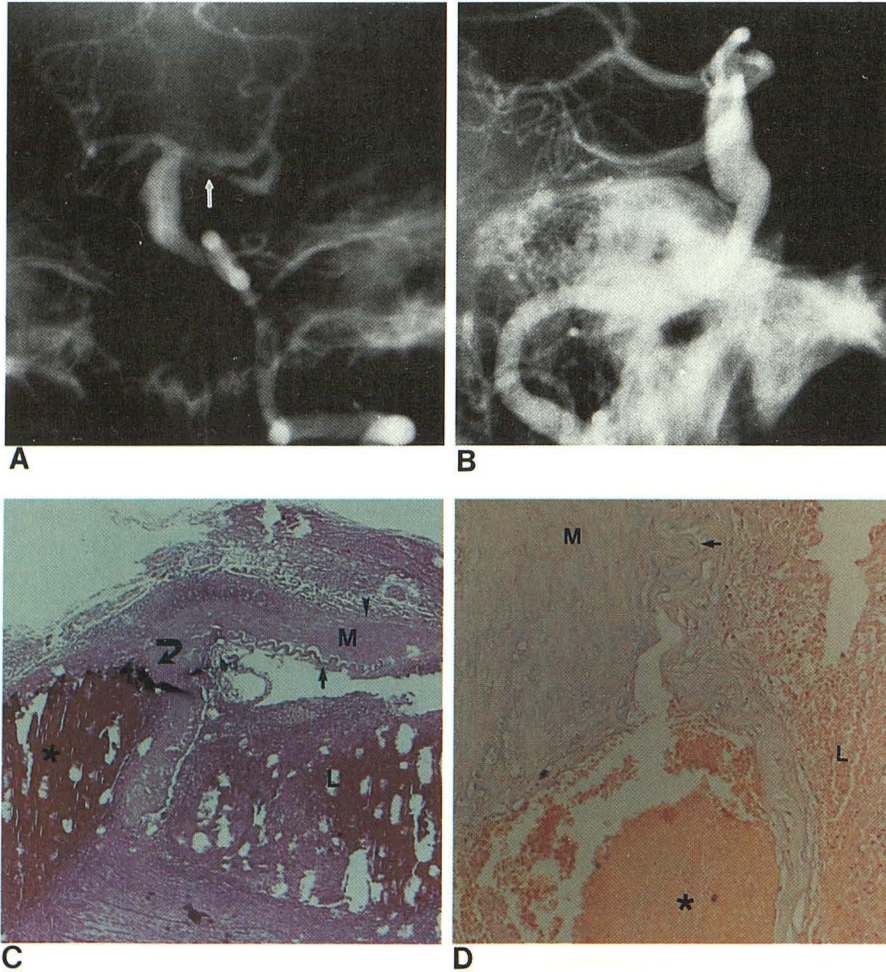


Fig. 1.—A, Anteroposterior view from left vertebral arteriogram shows dilatation of basilar, left superior cerebellar, and left posterior cerebral arteries. Note intervening narrowing in left superior cerebellar artery (arrow), which suggests dissection.

B, Lateral view from left vertebral arteriogram shows same findings.

C, Histologic section of basilar artery shows intramural thrombus from dissection (asterisk). The lumen (L), internal elastic membrane (straight arrow), and media (M) are marked. Note that the thrombus splits the media (curved arrow) and extends to the adventitial layer (arrow-head). (H and E stain)

D, Histologic section of basilar artery shows small vacuoles and blue-staining mucosubstance within media layer (M), indicating degeneration. L = lumen, arrow = internal elastic membrane, asterisk = dissection thrombus. (Alcian blue stain)

Angiographically, no single feature has been identified in all cases of intracranial arterial dissection. A narrowed, tapered, or irregular segment ("string sign") or total occlusion distal to an irregular narrowed segment is often observed. Fusiform dilatation proximal to the dissection and/or poststenotic dilatation ("string and pearl sign") also have been described [8]. A double lumen is pathognomonic but is rarely seen. In our patient, the fusiform basilar artery aneurysm with luminal narrowing of the proximal left superior cerebellar artery and dilatation of the more distal left superior cerebellar artery corresponded with the dissection that was present at autopsy.

In summary, a 35-year-old man with Marfan syndrome and previous aortic repair died following rupture of a basilar artery dissecting aneurysm. The presence of basilar artery dissection represents a rare association in this syndrome.

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