

## Extracranial Vertebral Aneurysm with Neurofibromatosis

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**Summary.** With neurofibromatosis neuromas of the cranial and cervical nerves are common findings and meningiomas and ependymomas appear more often than in the average population. Vascular manifestations of the disease are also commonly known in the renal and gastrointestinal vessels but rarely in the large cerebral arteries. The case of a 50 year old man with neurofibromatosis and a vertebral aneurysm is reported. The plain X-ray of the cervical spine and the Pantopaque myelogram were compatible with a cervical neuroma and the patient was operated but a large aneurysm of the vertebral artery was found. The operation was discontinued and vertebral angiography was performed revealing a large sacular aneurysm which was excised. This aneurysm is most probably a manifestation of vascular disease with neurofibromatosis.

**Key words:** Neurofibromatosis — Vertebral aneurysm.

A man aged 50 years suffered from neurofibromatosis with multiple fibromatous lesions and café-au-lait spots of the skin for 20 years. No neurological deficit were noted until February 1976 when he felt sudden pain in the left shoulder and paresthesias in the left thumb and forefinger. Paresis of the left shoulder and arm with a sensory deficit developed in May 1976. Roentgenograms of the cervical spine showed a marked dilatation of all intervertebral foramina on the left side, especially at C<sub>4</sub> and C<sub>5</sub> (Fig. 1). A Pantopaque myelogram revealed a left medially convex impression of the contrast column, displacing the spinal cord to the right and producing a subtotal block (Fig. 2). No pulsation was noted under fluoroscopic

control. In consideration of the known neurofibromatosis which was histologically verified by biopsy of a cutaneous nodule, the diagnosis of a cervical neurofibroma was made although the myelographic findings were not typical. Operation was performed



**Fig. 1.** Plain X-ray of the cervical spine shows widening of all intervertebral foramina. The pedicles are thinned at C<sub>4</sub> and C<sub>5</sub>. A small erosion of the vertebral body C<sub>5</sub> is suggested

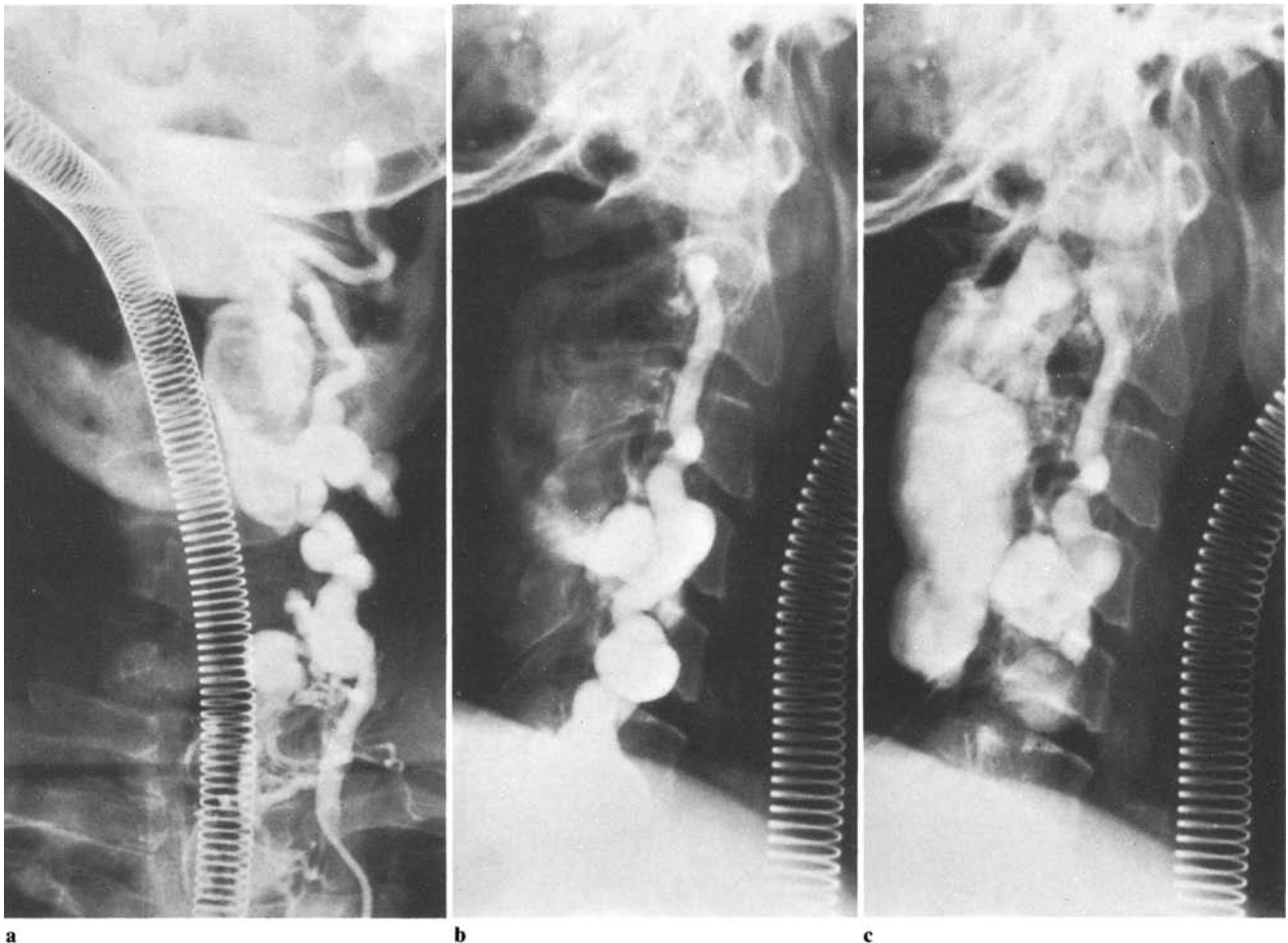


in May 1976 and a large aneurysm of the vertebral artery was found compressing the spinal canal. In default of an angiogram, the operation was interrupted. A large saccular aneurysm of the vertebral artery was later demonstrated angiographically (Fig. 3). Most of the small muscular arteries filled from the vertebral artery. At a second operation the vertebral artery was ligated at its origin, and at a third operation the aneurysm was removed. A large aneurysmal sac was found extending from C<sub>6</sub> to C<sub>2</sub>. Despite previous ligation of the vertebral artery the aneurysm filled through muscular anastomoses. The aneurysm was located extradurally and the nerves C<sub>2</sub> to C<sub>6</sub> were stretched and pushed forward over the aneurysm. At the level of C<sub>2</sub> a fingerform aneurysmatic sac compressed the spinal cord. A slow recovery of the neurological deficit in the left arm occurred after the operation and one year later the patient complained only of a minor weakness of his left shoulder.

### Discussion

Neurofibromas, especially of the eighth, but also of the other cerebral and cervical nerves are common

**Fig. 2.** Pantopaque myelography: The cervical cord is displaced to the right by an extradural, sharply delineated mass, beginning at C<sub>5-6</sub>. A total stop occurs at C<sub>3</sub>. No pulsation was noted under fluoroscopic control



**Fig. 3.** Left vertebral angiography. **a** AP view. **b** Lateral view, early phase. **c** Lateral view, late phase. There is a saccular aneurysm of the vertebral artery beginning before the artery enters the costotransverse foramen at C<sub>7</sub>. A large intraspinal aneurysmatic sac extends from C<sub>5</sub> to C<sub>2</sub>

findings with neurofibromatosis. Other tumors such as meningiomas, astrocytomas and ependymomas [1] as well as pheochromocytomas [2] are encountered with neurofibromatosis.

Neurofibromatosis is a generalized connective tissue disease and vascular participation is known. Reubi [3] classified the arterial changes of the renal arteries and Feyrter described [4] the vascular manifestations of neurofibromatosis in the gastrointestinal tract. Stenotic lesions are common, not only in the renal and intestinal arteries, but also in the abdominal aorta, producing coarctation [5]. Occlusion of the great cerebral vessels may produce Moyamoya disease [6]. Aneurysma dilatations also are encountered in the abdominal, especially renal arteries and rarely one finds aneurysms of the circle of Willis with neurofibromatosis [6, 7]. Almost all extracranial vertebral aneurysms are traumatic, commonly by penetrating injuries, traffic accidents or sport injuries, seldom after chiropractic manipulations of the cervical spine [8, 9, 10]. Our patient never suffered cervical trauma and we think, that this vertebral aneurysm is a manifestation of neurofibromatosis. Even without histological proof this assumption is justified, since it is known that the histological proof of vascular neurofibromatosis is often impossible from small bioptic pieces [5]. The changes of the neural foramina and vertebral bodies produced by anomalies of the vertebral artery are well known [11, 12]. Vascular lesions as well as neuromas may cause widening of the intervertebral foramina.

## Conclusion

In patients with neurofibromatosis vascular changes may mimic radiological signs and clinical symptoms of neuromas as in this case. Bearing this in mind, a not absolutely typical picture on the myelogram or on plain X-Rays should alert us to search for other pathology than neuroma.

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