Right subclavian artery aneurysms of fibrodysplastic origin: Two case reports and review of literature

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Right subclavian aneurysms involving the intrathoracic portion of the artery are rare and those of fibrodysplastic origin are mentioned in literature only as sporadic cases. In this article, we present two cases of this uncommon pathologic condition and discuss problems concerning diagnostic tools and technical choices. The two patients underwent a successful vascular graft substitution; an echo-Doppler scan revealed that they had no disease 1 and 2 years after the operation. (J Vasc Surg 2001;33:174-7.)

Subclavian artery aneurysms (SAAs), more commonly found in the extrathoracic district, may be occasionally intrathoracic and should always be considered in the differential diagnosis of expansive lesions in the superior mediastinum, like neoplasms, lymphadenopathies, and substernal goiters. They require surgical treatment because of the known risk of rupture, embolization, and thrombosis.[1] The intrathoracic portion of the artery can be affected by atherosclerosis, Marfan syndrome, trauma, or infection, but only a few cases have been ascribed to fibromuscular dysplasia (FMD). We present two recent cases of dysplastic aneurysms affecting the subclavian artery in the same proximal tract; different invasive treatments were used in both cases.

CASE REPORTS

Case 1. A 27-year-old man who had a strong constitution, had a short neck, was asymptomatic, and had no history of traumatic lesions or infective disease came to us for an occasional chest radiograph diagnosis of a mediastinal widening with dislocation of the trachea, which resembled a substernal goiter. An iodine-131 scan did not demonstrate any alteration of the thyroid gland, whereas computed tomographic (CT) scan (without contrast medium) confirmed an enlarged right thyroidal lobe, which was extended to the paratracheal area through the thoracic inlet. Serum-free triiodothyronine, free thyroxine, and thyroid-stimulating hormone levels were normal. The operation was started with a transverse cervicotomy, but no relevant disease of the thyroid gland was ascertained. A right substernal pulsating mass was revealed and studied intraoperatively with angio-CT scan and arteriography, which revealed a proximal aneurysm of the right SAA exceeding 5 cm in diameter and the occlusion of the right vertebral artery (Fig 1). Then, through median sternotomy, a subclavian replacement with a polytetrafluoroethylene (PTFE) graft was performed (a study of somatosensory evoked potentials [SEPs] excluded cerebral ischemia after clamp occlusion of innominate and carotid arteries). The postoperative course was optimal, and the patient was discharged on the 12th postoperative day. The histologic characteristics were those of medial fibrodysplasia, with thinning, hypotrophy, and disorganization of muscular fibers, which were replaced by collagenous tissue, and intimal discontinuity, with some tracts of duplication of elastic layer and focal thickenings (Fig 2). Color Doppler ultrasound mapping of the major vessels did not demonstrate any other dysplastic lesion. Currently, the patient is well at about 1 year from the operation, and there is a normal Doppler function of the graft.

Case 2. A 27-year-old woman affected by chronic renal failure and asymptomatic for cardiovascular diseases was noted to have an abnormal mediastinal silhouette on routine chest x-ray film. Results of a physical examination revealed a reduction of the right arm blood pressure. Color Doppler ultrasound, CT scan, and magnetic resonance of the brachiocephalic vessels demonstrated the presence of a right SAA involving the intrathoracic portion of the vessel associated to a preaneurysmatic stenosis and vertebral steal syndrome. Arteriography confirmed strict stenosis of the subclavian artery right after the vertebral emergency, with a poststenotic posterior aneurysm; selective injection of the vertebral artery showed immediate contrast of the deep cervical artery, giving collaterals for the upper arm. There was no contrast evidence in the internal mammary artery or in the thyrocervical trunk. Isolation of the aneurysm was obtained through a supraclavicular transversal incision with preservation of the nervous and vascular structures. SAA replacement with a PTFE graft was performed with maintenance of the blood flow in the anonymous-carotid axis and occlusion of the subclavian and vertebral arteries, after the sectioning of the mammary and thoracicoacromial arteries. We reestablished vertebral blood flow by performing proximal anastomosis right after the origin of the vertebral artery. Histologic examination of the aneurysm wall revealed areas of fibrous degeneration and scleroialynosis, typical of medial fibrodysplasia. Two years after the operation, the patient does not show any abnormality in upper arm vascularization or in graft Doppler flow. Even in this patient, the color Doppler ultrasound mapping was normal.

DISCUSSION

Right SAAs may rarely be of fibrodysplastic origin, but more frequently their cause is attributed to a thoracic outlet syndrome or to a penetrating trauma when they are

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Competition of interest: nil.

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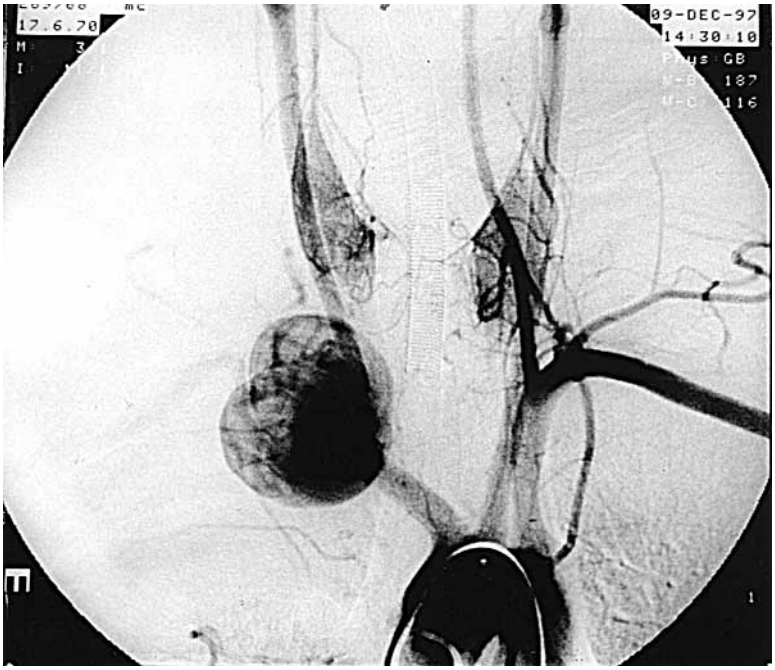

Fig 1. Intraoperative angiography reveals large aneurysm of proximal right subclavian artery.

localized in the extrathoracic portion of the artery. In the intrathoracic portion, they are mostly due to atherosclerosis or to Marfan syndrome.[2] In reviewing literature on the argument, we found only single reports or small series, which were often not satisfactory in regard to precise histologic diagnosis of dysplastic lesions, which can comprehend FMD but also medial degeneration, cystic medial necrosis, and congenital collagen disease.[1,3,4]

FMD is a rare pathologic condition that mostly affects females (90%). The effective incidence of the disease is actually unknown; it is prevalent in renal arteries, followed by carotid and vertebral arteries.[5] Heffelfinger et al[6] report an autoptic incidence of renal FMD of up to 1%, whether radiographic incidence of the typical "string of beads stenoses" aspect in carotid angiograms varies from 0.25% to 1%.[7] In some series multifocal artery involvement occurs in up to 25% of the cases.[5] Of the three histologic types of FMD (medial FMD, intimal fibroplasia, and periarterial or periadventitial fibroplasia), only the first two types have been observed in the brachiocephalic region, with an absolute prevalence of medial FMD.[5] FMD is more frequently responsible for stenotic lesions (thus the term *fibromuscular hyperplasia*),[8] whereas aneurysmatic presentation is relatively more uncommon. To our knowledge, only one case of right SAA with histologic diagnosis of FMD is reported in the English medical literature: McCready et al[9] describe a 44-year-old woman affected by two small saccular SAAs that are responsible for vertebral steal syndrome and are successfully treated with resection and primary end-to-end reanastomosis. Few sporadic reports of fibrodysplastic aneurysms are found regarding left or aberrant right subclavian, axillary, and brachial aneurysms,[10-14] whereas in a review of English medical literature, Rhee et al[15] report 21 described cases of extracranial carotid aneurysms up to 1996.

Symptoms are prevalently related either to the presence of an enlarging mass in the upper mediastinum or to the associated proximal stenosis. Pairolero et al[16] and Coselli and Crawford,[1] who have collected two of the largest series of SAAs of which none were due to FMD, report symptoms in about three fourths of the patients, including shock from rupture, sensation of pulsating supraclavicular mass, arm pain and claudication, Horner's syndrome, and hoarseness from vocal cord paralysis, whereas peripheral ischemia seems more typical of distal (extrathoracic subclavian artery, axillary or brachial) localizations. In McCready et al's case and ours, clinical appearance was more subtle, because one patient was totally asymptomatic, whereas the other two had mild symptoms due to vertebral steal syndrome.

Diagnosis was occasional, after the chest radiographs. In our first reported case, CT scan without contrast media interpreted the mass as a substernal goiter. If contrast media cannot be used (for example, in idiosyncratic reactions or renal failure), the use of alternative examinations such as magnetic resonance angiography or Doppler ultrasound to reveal masses of vascular origin is mandatory. We consider arteriography or magnetic resonance angiography to be of extreme utility to determine extension to other brachiocephalic trunks or the presence of possible associated lesions.[17]

The surgical approach to the right subclavian artery is often feasible simply through a supraclavicular incision, but sometimes, sternotomy is required to prevent potentially

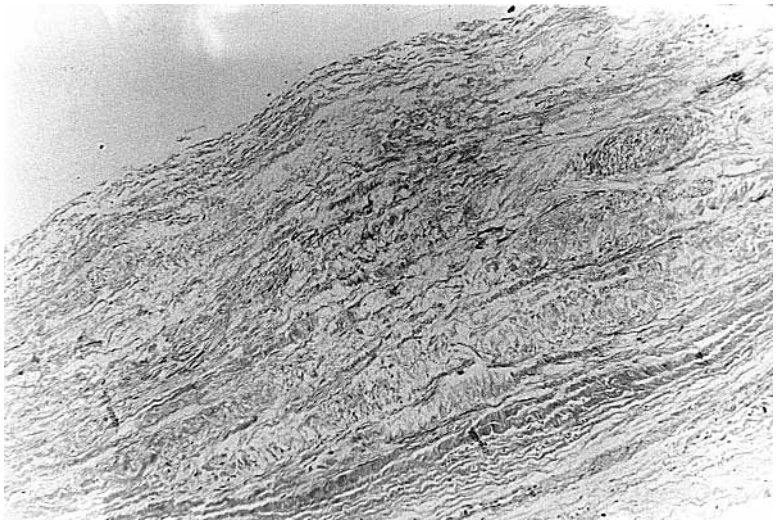

Fig 2. Histologic examination (orcein stain, original magnification ×20) demonstrates hypotrophy and disorganization of the median layer, replaced by collagen and mucoid material.

lethal bleedings due to inappropriate control of the vessel in its intrathoracic portion.[18] For this reason adequate information of the patient and preparation of the surgical field and equipment are required to perform immediate complete or split sternotomy whenever necessary.[19] Aneurysm location was similar in our two cases; in both, we obtained an adequate control of the innominate, carotid, and subclavian arteries. The more invasive approach required in the young man was conditioned because of his strong constitution. Surgical management comprises replacement of the aneurysmatic tract with synthetic or autologous graft, followed by an extra-anatomic bypass graft or resection and ligature whenever substitution is feasible. Coselli and Crawford[1] report the treatment of a subclavian false aneurysm by the use of resection and angioplasty with a polyester fiber (Dacron) patch[1]; a similar reconstruction is presented by Bower et al[2] in a case of saccular aneurysm. A patient who was recently treated with endovascular stenting is described by May et al.[20] The simple ligature of the artery, proposed by pioneers of this surgery such as Greenough and Halsted, is not performed anymore because of acute ischemia of the upper arm occurring in about a quarter of the patients.[16] In both cases we performed endoaneurysmectomy and PTFE replacement, with isolation and respect to the vagus, inferior laryngeal nerve (usually underlying the aneurysm), phrenic nerves, and right internal jugular vein. The functional results of the graft (PTFE, 7 mm) have been asserted with intraoperatory Doppler scan.

The perioperative mortality rate in the medical literature varies between 3% and 8%, and it usually includes patients who are treated in the emergency department for aneurysmatic rupture or who require treatment for associated pathologic conditions (aortocoronary bypass grafts or other vascular substitutions).[1,16,18,21] The morbidity rate reported in such studies was 6% to 8%, which was mostly due to postoperative bleedings or to motility dysfunctions due to cranial nerve injuries. We consider precise anatomic definition of the lesion (based on radiologic study) important to prevent morbidity, along with an adequate incision (to obtain an optimal proximal and distal control) and accurate dissection and isolation of nervous and vascular structures. The cerebral response to each surgical maneuver can be controlled with SEPs, especially when the carotid flow is partially or totally interrupted by the clamp. The number of long-term survivors in the medical literature is satisfactory, even if the small number of cases reported does not permit a distinction between different lesions and associated pathologic conditions, but we must consider the worse prognosis in terms of early and late mortality of SAAs in comparison with other aneurysms of the aortic arch vessels.[16,19,22] Both of our patients did well postoperatively, and they preserve an optimal graft function after more than 1 year, without any dilatation or infective event during color Doppler scan control.

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