

Case report

Cervical aortic arch with aneurysm formation

Bora Farsak, Mustafa Yilmaz, Sadi Kaplan, Erkmen Böke*

Department of Thoracic Cardiovascular Surgery, Hacettepe University, Faculty of Medicine, 06100 Ankara, Turkey

Received 9 February 1998; revised version received 29 June 1998; accepted 7 July 1998

Abstract

Cervical aortic arch is a very rare anomaly presented as a pulsatile mass on the neck and usually with symptoms of dysphagia, cough and hoarseness. Rarer than the cervical aortic arch, is the aneurysm formation and, despite the equal sex distribution of cervical aortic arch, aneurysm formation always occurs in young females with only nine cases reported. We report herein a 24-year-old woman, diagnosed as cervical aortic arch with aneurysm formation due to basophilic degeneration, treated successfully with surgical intervention. To our knowledge no similar case has been reported. © 1998 Elsevier Science B.V. All rights reserved

Keywords: Aorta; Cervical aortic arch; Aneurysm formation

1. Introduction

Since its first introduction by Reid [1] in 1914 very few cases of cervical aortic arch have been reported. Rarer than that is the cervical aortic arch complicated with aneurysm formation with only nine cases found in the literature [2–9].

We herein report a case of cervical aortic arch complicated with aneurysm formation caused by basophilic degeneration of the aortic wall.

2. Case report

A 24-year-old woman presented complaining of a pulsatile mass on the left side of her neck, minimal swelling on the left side of the face, intermittent attacks of headache, tinnitus and numbness in her left upper limb. On closer questioning, she noted an increase in the frequency of the last mentioned complaints and in the size of the mass on her left neck with minimal solid dysphagia and 6 kg weight loss in the last 6 months. Her medical history was otherwise unremarkable. On physical examination, blood pressure

was 120/70 mmHg and all peripheral pulses were palpable and there was no difference compared with the left upper limb. There was a pulsatile left supraclavicular and supra-sternal mass. Chest roentgenogram showed a widened superior mediastinum and a subsequent chest computed tomographic scan revealed mild dilatation of the aorta. Echocardiography demonstrated a trace mitral insufficiency. Intravenous digital subtraction angiography showed a large saccular aneurysm of the aorta, beginning just beyond the left common carotid artery, including the orifice of the left subclavian artery (Fig. 1).

Left brachiocephalic and common carotid arteries were normal but tortuosity and irregularity in the right subclavian and brachiocephalic artery was noted. With this diagnosis, it decided that the patient was to be a candidate for a surgical intervention.

At operation, with midline sternotomy, thin walled saccular aneurysm of the aorta and left subclavian artery was identified. The long aneurysmal segment of the aorta, just distal to the left common carotid artery and aneurysmal segment of the left subclavian artery was resected and reconstructed with a prosthetic interposition graft (Meadox Medicals, Oakland, NJ).

The aorta was reconstructed in an end-to-end fashion and the left subclavian artery was reconstructed to the grafted

* Corresponding author. Tel.: +90 312 3117377.

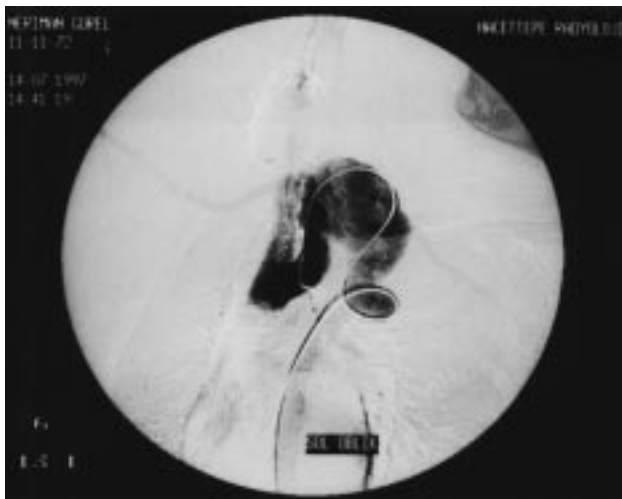


Fig. 1. Intravenous digital subtraction angiography showed a large saccular aneurysm of the aorta, beginning just beyond the left common carotid artery, including the orifice of the left subclavian artery.

segment of the aorta by using a prosthetic interposition graft (Meadox Medicals, Oakland, NJ). Throughout the operation the distal aortic pressure was above 50 mmHg so no shunt was used to support the distal circulation while the aorta was clamped. The aortic cross clamp time was 21 min.

Histologic examinations revealed a thinned arterial wall which showed typical medial basophilic degeneration and fragmentation of the smooth muscle.

The post-operative course was uncomplicated and she was discharged home on the eleventh post-operative day. Now 6 months post-operation, the patient has no complaints and her control angiography revealed normal flow both in the aorta and the left subclavian artery (Fig. 2).

3. Discussion

Cervical aortic arch is a very rare anomaly, even rarer is the aneurysm formation. To our knowledge, the case described, cervical aortic arch with aneurysm formation, is the only reported case due to basophilic degeneration, with similar findings to the previously reported cases of aortic aneurysm formation [2–8].

When we looked through the reported cases, all patients were young females. Mostly on referral, the major finding was a palpable mass on the neck or supraclavicular region. In the other reported cases symptoms were either absent [2,3] or included dysphagia [4,5], cough or hoarseness [6], dizziness [7], mild stroke [8] or one patient presented with traumatic aortic rupture [8].

In our case a 24-year-old female presented with a palpable mass on her neck and, when questioned, she had been suffering from dysphagia and weight loss for the previous 6 months.

Table 1 shows the previously reported cases.

There are many questions, still waiting to be answered.

Despite the approximately equal sex distribution of cervical aortic arch [10], only female patients with cervical aortic arch have developed aneurysms, a factor that may influence risk counselling of female patients with cervical aortic arch. Most aneurysms are in the transverse aortic arch, which is the location least affected by other types of aortic aneurysm. Only the case reported by Cooley et al. [2] had an associated cardiac lesion (ventricular septal defect).

Including our case, there were three cases reported with a pathologic diagnosis, [2] was cystic medial necrosis [3,7] and ours was basophilic degeneration.

In general, the pathophysiology of aneurysmal aortic dilatation in cervical aortic arch is not clear. Aortic aneurysms in adults usually result from atherosclerotic changes.

In children, they most often arise from loss of structural integrity due to abnormal collagen, such as in genetic syndromes.

Possible explanations for aortic aneurysms developing in cervical aortic arch include abnormal embryologic development, abnormal connective tissue, altered hemodynamics and aortic wall stress and trauma.

The normal left-sided aortic arch forms from persistence of the fourth left branchial arch, with regression of more cephalad arches. In contrast, cervical aortic arch is most likely due to persistence of the third branchial arch. An aortic arch formed from an embryologic remnant designed to regress may not have normal structural integrity. This, coupled with the hemodynamic alterations in the tortuous cervical aortic arch, may provide a substrate for aneurysm formation. In three patients with left cervical aortic arch, the aneurysms were between the left carotid and subclavian arteries, an area likely to be composed of tissue from the third branchial arch. Another factor supporting this hypothesis is that, although cervical aortic arch is rare, aneurysms complicate about 20% of cases. This relatively frequent



Fig. 2. Post-operative angiogram showing normal flow both in the aorta and the left subclavian artery.

Table 1

Summary of cases of cervical aortic arch with aneurysm formation

Investigators	Age (years)	Sex	Side of arch	Branching	Location of aneurysm	Associated lesions	Pathology
Pitzus and Camoglio [9]	39	Female	Left	BCA, LCC, LSCA	Aortic arch	–	–
Cooley et al. [2]	39	Female	Right	BCA, RCC, RSCA	Aortic arch	VSD	–
Morris and Rutley [4]	36	Female	Left	BCA, LCC, LSCA	Descending aorta	–	–
Cao et al. [7]	21	Female	Left	BCA, LCC, LSCA	Between LCC and LSCA	–	CMN
Van Nooten et al. [3]	17	Female	Left	BCA, LCC, LSCA	Between LCC and LSCA	–	CMN
Kumar et al. [6]	37	Female	Left	BCA, LCC, LSCA	Between LCC and LSCA	–	–
Holland and Fitzpatrick [5]	42	Female	Right	LCC, RCC, RSCA LSCA	At apex of arch	–	–
Pearson et al. [8]	11	Female	Left	BCA, LCC, LSCA	Between LCC and LSCA	–	–
	6	Female	Left	BCA, LCC, LSCA	Between LCC and LSCA	–	–

BCA, brachiocephalic artery; LCC, left common carotid artery; LSCA, left subclavian artery; RSCA, right subclavian artery; VSD, ventricular septal defect; CMN, cystic medial necrosis.

occurrence suggests that a factor, or factors, inherent in cervical aortic arch also may be involved in the pathogenesis of aneurysms, although it still does not explain the appearance of aneurysms only in female patients.

References

- [1] Reid DG. Three examples of a right aortic arch. *J Anat Physiol* 1914;48:174–181.
- [2] Cooley DA, Mullins CE, Gooch JB. Aneurysm of right-sided cervical arch; surgical removal and graft replacement. *J Thorac Cardiovasc Surg* 1976;72:106–108.
- [3] Van Nooten G, Deuvaert F, De Paepe J, Primo G. Left-sided cervical aortic arch. *Acta Chir Belg* 1986;86:248–250.
- [4] Morris R, Rutley M. Left cervical aortic arch associated with aortic aneurysm. *Br Heart J* 1978;40:87–90.
- [5] Holland P, Fitzpatrick JD. Case report: magnetic resonance imaging of a right-sided cervical aortic arch with a congenital aneurysm. *Clin Radiol* 1991;43:352–355.
- [6] Kumar S, Mandalam KR, Unni M, Rof S, Gupta AK, Rao VR. Left cervical arch and associated abnormalities. *Cardiovasc Intervent Radiol* 1989;12:88–91.
- [7] Cao P, Angelini P, Colonna L, Cristallo E, Cooley DA. Cervical aortic arch with mediocystic necrosis. *Bull Texas Heart Inst* 1980;7:188–193.
- [8] Pearson GD, Kan JS, Neill CA, Misgley FM, Gardner TJ, Hougen TJ. Cervical aortic arch with aneurysm formation. *Am J Cardiol* 1997;79:112–114.
- [9] Pitzus E, Camoglio E. La sindrome della orta cervicale. A proposito di un caso clinico. *Giornale Italiano di Cardiologia* 1974;4:217–218.
- [10] Felson B, Strife JL. Cervical aortic arch: a commentary. *Semin Roentgenol* 1989;2:114–120.