Left Subclavian Artery Aneurysm in Behçet's Disease Presenting as Thromboemboli and Brachial Plexopathy

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Abstract

Behçet's disease is an uncommon multisystemic disorder that appears most often in the third or fourth decade of life. It is characterized by recurrent orogenital ulcers and ocular and cutaneous inflammatory lesions. Cardiovascular involvement, which may be arterial or venous, is rare but carries a particularly poor prognosis. Arterial involvement is less frequent, constituting 12% of cardiovascular complications. The arterial findings in this disease may be occlusions and aneurysms or pseudoaneurysms of the aorta and pulmonary, brachial, carotid, subclavian and visceral arteries. In this report, we present a 41-year-old man having suffered from Behçet's disease for more than 12 years, complicated by a big bi-saccular aneurysm of the 1st and 2nd portions of the left subclavian artery. The presenting symptoms and signs were thromboembolic arterial occlusion and brachial plexus compression ***(Iranian Heart Journal 2005; 7 (3): 70-75).***

Key words: Behçet's disease Ë subclavian arterial aneurysm Ë thromboemboli Ë brachial plexopathy

Behcet's disease is a multisystemic disorder characterized by recurrent oral and genital aphthous ulcers, ocular manifestations and skin lesions. Cardiovascular involvement occurs in only 7-29% of the patients.[1] However, it is the most common cause of death in these patients.[2,3] Among the vascular lesions, venous involvement is more common than the arterial, which accounts for only 12% of cardiovascular complications in Behçet's disease. Arterial lesions mostly develop in the aorta, pulmonary artery and their major branches. Saccular aneurysms and occlusion of multiple large vessels in young adults are the most common types of arterial involvement. Subclavian artery aneurysm is not frequent among the peripheral artery aneurysms, accounting for 1% of them.[8-10] There are some reported cases of subclavian artery aneurysm in the available literature.[8, 11] The most common etiologies are thoracic outlet syndrome (TOS) and atherosclerosis.[12,13] Other uncommon causes include trauma, injections and arthritis.

To our knowledge, Behçet's disease complicated by subclavian artery aneurysm has not been reported in the literature thus far.

In this report, we present an adult man suffering from Behçet's disease presenting with acute ischemia of the left upper extremity and signs of brachial plexus compression.

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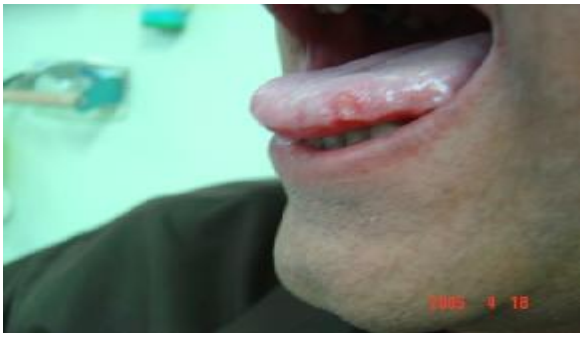
Case report

We present a 41–year–old man with a 12-year history of Behçet's disease diagnosed based on a positive pathergy test, recurrent ulcers in the mouth (resembling canker sores) and uveitis (inflammation of the uvea of the eye), which caused blurred vision (Fig. 1).



A

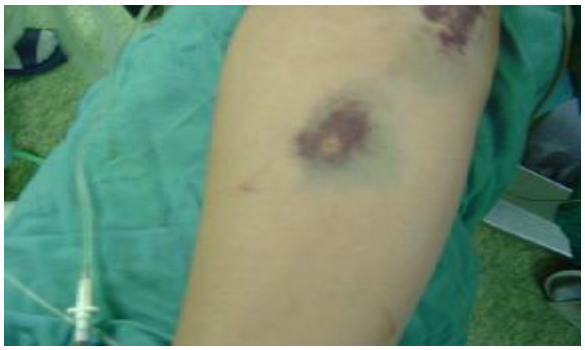


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Fig.1. A, Ulcer of tongue (two months after surgery) and (B) pathergy test in Behçet's disease presenting with subclavian artery aneurysm.

The patient referred to our center because of acute ischemia of the left upper extremity, severe pain in the left shoulder and arm combined with paresthesia of the ulnar nerve dermatome. The patient was successfully treated with acetylsalicylic acid (100mg/day) and colchicine (1mg/day).

He discontinued his medication without consultation with his doctor. At the time of admission, there was no clinical sign of the evolution of Behçet's disease. On physical examination, a pulsatile fullness was palpable in the left supraclavicular fossa (Fig. 2).

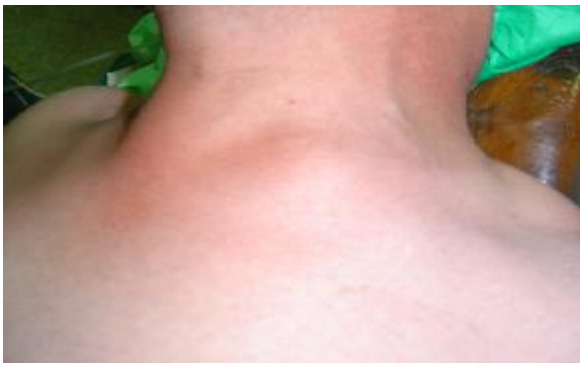

Fig. 2. Fullness of supraclavicular fossa in Behçet's disease presenting with subclavian artery aneurysm.

Distal pulses of the affected limb were absent. Pulses of the other extremities were normal. Chest x-ray revealed a widened upper mediastinum (Fig. 3).

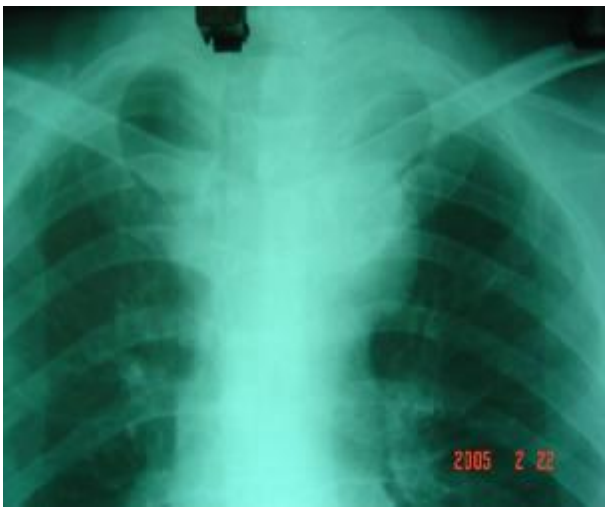

Fig.3. Chest X-ray in Behçet's disease presenting with subclavian artery aneurysm.

Computed tomographic angiography showed the aneurysm to be 5 cm in size in the first portion of the left subclavian artery with a

significant rim of clots (Fig. 4).

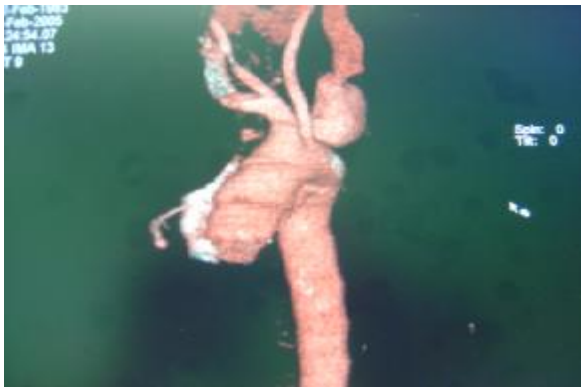

Fig. 4, A. Three-dimensional CT angiography in Behçet's disease presenting with subclavian artery aneurysm.

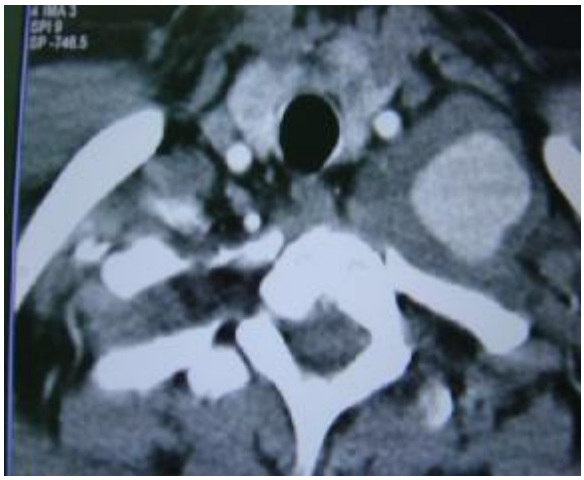

Fig. 4, B. Two-dimensional CT angiography in Behçet's disease presenting with subclavian artery aneurysm.

In addition to routine preoperative work-up, appropriate consultations including rheumatology and ocular services were provided. Pulse methylprednisolone and azathioprine were prescribed, followed by 25mg prednisolone BID and 50 mg azathioprine BID.

Because of the acute ischemia of the left hand, the patient underwent emergency surgical intervention. The left subclavian artery was explored by means of standard supraclavicular approach. The anterior scalene muscle and the phrenic nerve over it were deviated laterally due to the compression effect of the aneurysmal sac originating from the intrathoracic portion. Major surgery and excision of the aneurysmal sac was impossible at this stage because of acute limb ischemia and surgical limitations. Therefore, the source of embolization was obstructed by ligating the outflow of the aneurysmal sac. Blood supply to the limb was provided by axillo-axillary bypass grafting, using a ringed stretched PTFE tube graft with a 6-mm inner diameter (Fig. 5).

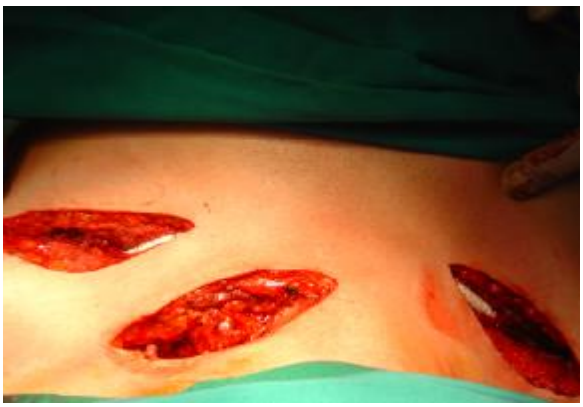

Fig. 5. Operation view of axillo-axillary bypass in Behçet's disease presenting with subclacian artery aneurysm.

Fig. 6. Postoperative digital subtraction angiography in Behçet's disease presenting with subclavian artery aneurysm.

Normal distal pulse returned following bypass grafting. Arch and four-vessel digital subtraction angiography 2 weeks after surgery revealed no outflow of the aneurysmal sac and disappearance of branches of the 1st and 2nd portions of the subclavian artery (Fig. 6). CT angiography one month later demonstrated near total clotting of the aneurysmal sac (Fig. 7).

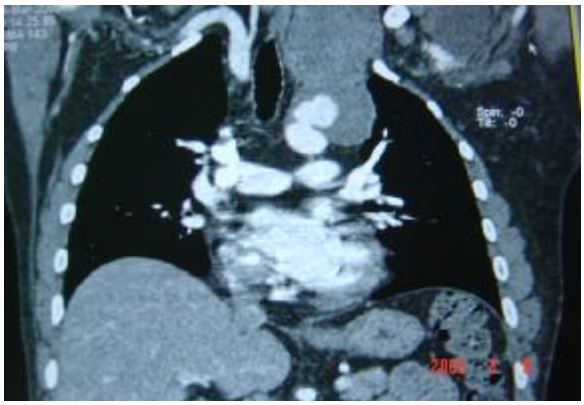

Fig. 7. One month CT angiography in Behçet's disease presenting with subclavian artery aneurysm.

Discussion

In our review of reported cases of subclavian artery aneurysm in the literature, we did not find subclavian artery aneurysm in conjunction with Behçet's disease, nor did we note the association of Behçet's disease with subclavian artery aneurysm. These data include aneurysms of the subclavian artery with their etiology (Table I).

Table 1: Reported cases of subclavian artery aneurysm and its cause

Author	Year	Number	Age	Cause	Presentation	Treatment
Thomas	1972	1	61	ASO	Painless neck mass	Excision and graft
McCollum	1979	2		ASO Also CMN	Thrombosis/ischemia In 1 patient: neck pain, Mass, or no Sx in 2	Mainly excision and graft
Elefteriades	1983	1	50	ASO	Chest pain/rupture	Ligation on L (ruptured)
Coseli	1987	2	42 (mean)	Marfan's syndrome	Chest pain, Horner's Syndrome, tracheal Compression	Ligation, Carotid-SC bypass
Akasaka	1989	1	38	Turner's Syndrome	Chest pain	Excision and graft
Kulpati	1990	1	60	ASO	Hemoptysis	Excision
Bower	1991	4		ASO	Pain in 2, mass in 2, neurological Sx in 2, 1 rupture, 3 thrombosis, 2 emboli/gangrene	Excision and graft (5)
Salo	1990	6	60 (mean)	ASO mycotic	3 symptomatic, 2 brachial embolism, 1 dysphagia, 1 neck mass, 1 chest pain	Excision and graft, ligation and carotid-SC bypass
Lakhar	1992	1	11	Congenital	Cough, asymptomatic thrombosis	Excision
Dougerty	1995	1	62	ASO	Asymptomatic	Excision, SC-carotid implant
Stahl	1999	2	22 (mean)	Congenital	Asymptomatic, chest wall tenderness	Excision and graft

ASO = arteriosclerosis obliterans; CMV = cystic medial necrosis; SC = subclavian; Sx = signs and symptoms

Aneurysms of the subclavian artery are relatively rare – 1% of all peripheral aneurysms. It has been noted that atherosclerosis is the most common cause of true aneurysms (60%), followed by infectious aneurysms (syphilis: 15%; tuberculosis: 10%; bacterial: few). Marfan's syndrome and cystic medial necrosis represent 10% of the cases. The male-to-female ratio is 2:1. The mean age for atherosclerotic aneurysm is 61 years.

Clinical presentation of subclavian artery aneurysm can be asymptomatic. When symptoms occur, chest or shoulder pain is the most common. Brachial plexus neuropathy, like the case we present, can occur. A pulsatile mass or fullness in the supraclavicular fossa with tenderness can be noted on physical examination, and sometimes, the aneurysms present with signs and symptoms of embolization and thrombosis as they did in our case. Other compression effects of subclavian artery aneurysm can lead to dysphagia, dyspnea, dysphonia and Horner's syndrome.

Intrathoracic subclavian artery aneurysm may be evident on chest radiograph as a widened mediastinum, as it did in our case (Fig. 3), and sometimes on computed tomography in routine examination.

Arterial aneurysms are a classic and severe complication of Behçet's disease. These aneurysms, whose incidence ranges from 2% - 6%,[14] are commonly located in the abdominal aorta as well as in the femoral or pulmonary arteries. Aneurysms of the internal carotid arteries are seldom reported.[15]

Operative therapy is usually recommended for the management of arterial aneurysms since rupture is the primary cause of death in patients with Behçet's disease. Before surgery, angiography and computed tomography are required to evaluate the anatomic details. Most of the approaches to the subclavian artery aneurysms are based on the assumption that the subclavian artery arises from the distal arch of the aorta and lays more posterior in the thoracic cavity and that it would be impossible to obtain proximal control from the supraclavicular approach.

In this case, we were reluctant to perform a major surgical operation through sternotomy and/or thoracotomy. Therefore, we opted for lighter surgery in the hope that the patient's acute ischemia would be cured and that the aneurysmal cavity would clot, the clotted sac would organize and shrink and finally, the compression plexopathy would be treated. Adjuvant immunotherapy with azathioprine, with or without high doses of corticosteroids, is usually required to control the formation of new aneurysms.[16,17]

Conclusion

Aneurysms of the subclavian artery are relatively rare – 1% of all peripheral aneurysms – and they may be either intrathoracic or extrathoracic or both.

The location of the aneurysm is usually related to its etiology: those located in the first part of the artery are usually atherosclerotic, and those located in the second and third parts are usually traumatic or due to thoracic outlet syndrome.

They may be asymptomatic or may cause ischemia to the hand by causing thrombosis or by giving rise to embolism and brachial plexopathy.

Duplex scanning should be the first investigation, and CT angiography is a definitive investigation and is very useful in planning the operation.

Resection with graft interposition (vein, PTFE) is the most appropriate procedure, and in many selected cases, resection with primary anastomosis is possible. In special cases, like the case we present, more conservative surgery, such as outflow ligation, arrests the source of distal embolization. Extra-anatomic bypass restores limb perfusion.

Surgical results are generally good; operative mortality is less than 5% for

uncomplicated cases. Nonetheless, in rare cases, such as Behçet's disease, prognosis is not good.

Acknowledgement

The authors would like to thank Mr. Amouzadeh for his review of the manuscript.

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