

Case report

Successful endovascular correction of common carotid pseudoaneurysm secondary to Behçet's disease: case report and review of the literature

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ABSTRACT

Behçet's disease (BD) is a chronic, multisystemic, inflammatory disease characterised by recurrent mucocutaneous, ocular, musculoskeletal, central nervous system, gastrointestinal and vascular manifestations, which may affect blood vessels of any size (1). Venous involvement is more common, but arterial involvement accounts for the major cause of mortality (2, 3). Choosing the adequate technique for correcting aneurysms in BD and the timing to do it is still challenging. The authors report a case of 37-year-old male patient with common carotid pseudoaneurysm at the time of diagnosis, which was successfully treated by an endovascular stent placement after adequate immunosuppression. A review of the literature about this issue was also done.

Case report

A male patient, 37 years old, no previous comorbidities, presented with sudden bulging pulsatile mass in the right cervical region (Fig. 1A). For the past 7 years, the patient had a history of recurrent oral ulcers, folliculitis in the trunk, painful skin lesions in the lower limbs, and past of intense headache, which later was diagnosis of chronic central venous thrombosis. Admitted to the emergency due to the cervical mass. On physical examination, presence of oral ulcers and pulsatile mass in the right cervical region. Laboratory showed elevated C reactive protein (CRP) at 208 mg/L. A CT angiography of the cervical region evidenced a right common carotid pseudoaneurysm with partial thrombus, measuring 4.0 x 4.0 x 3.8 cm, with a pedicle of 0.9 cm (Fig. 2). As the patient presented 5 points according to the International Criteria for Behçet's

Syndrome (IBCD), he was diagnosed with BD and pulse therapy with 500mg of methylprednisolone was performed for 3 days and sustained with oral prednisone 60mg/day and colchicine 1mg/day. Once he quickly normalised the inflammatory tests (CRP: 0.8mg/L, ESR: 3mm/h) and solved the oral ulcers, it was chosen not to administer cyclophosphamide. After two weeks of glucocorticoid immunosuppression, he underwent a minimally invasive procedure with successful endovascular expandable balloon stent placement (Be-Graft 6mm X 38mm). Post-operatively, aspirin and clopidogrel were administered and immunosuppression was intensified with azathioprine 75mg/day, that was increased to 200mg/day (2.8 mg/kg/day) after normalisation of liver enzymes. Control imaging was performed after 5 days, with presence of central laminar flow within the stent. Patient returned after 2 months of the endovascular procedure, asymptomatic and with complete disappearance of the mass in the cervical region (Fig. 1B). The patient repeated carotid ultrasound after 5 months of the procedure, being visualised patent stent, without new local lesions.

Discussion

According to the literature, vascular involvement occurred in 14.3% of 2319 Turks BD patients, including arterial lesions (13%), superficial venous thrombophlebitis (53%), deep vein thrombosis (29%) (2). Another study summarised the main features of arterial lesions in 101 patients, the prominent type of arterial involvement was aneurysms (47%), occlusion (36%) and stenosis (13%), and the most recurrent site was femoral (15%), pulmonary

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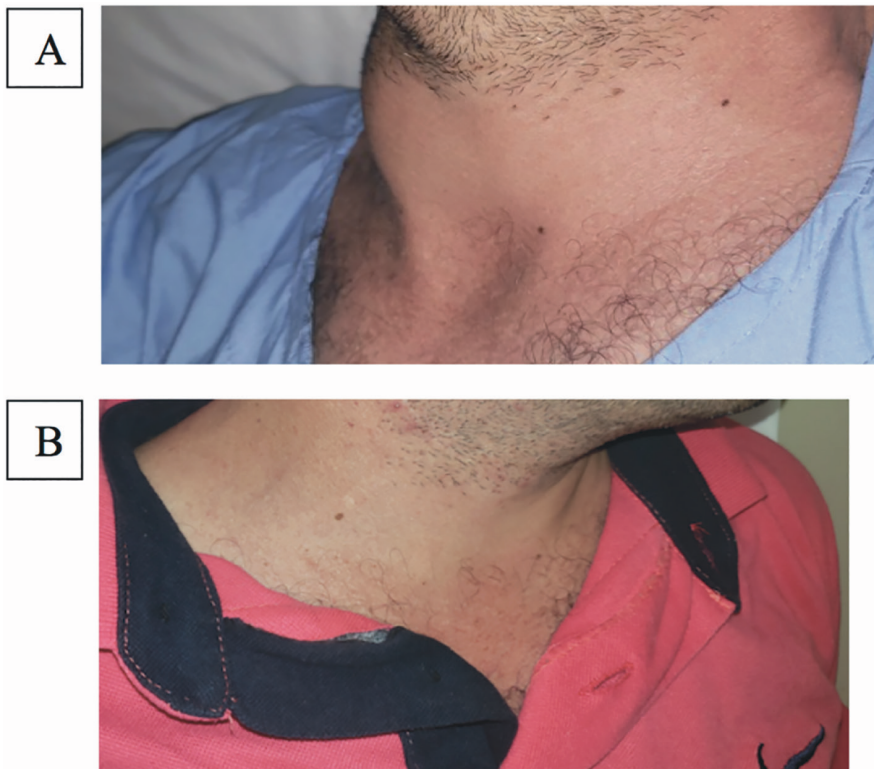


Fig. 1. A palpable pulsatile 4 cm bulging is observed at the right anterior cervical region before endovascular procedure (A), while no more signs are visible after treatment (B).

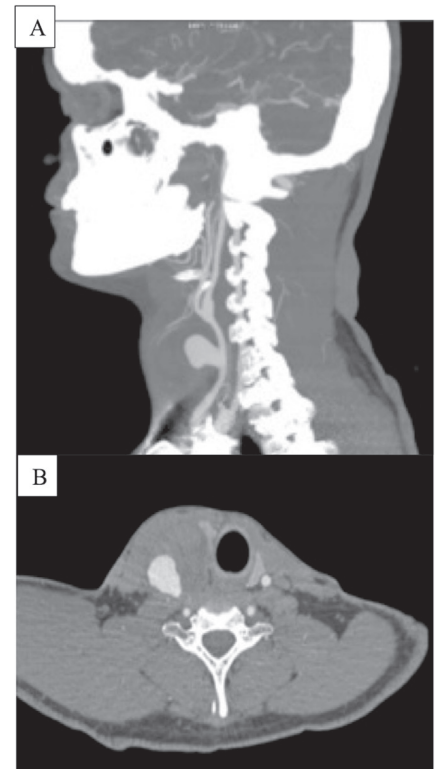


Fig. 2. A 4.0 x 4.0 x 3.8 cm saccular pseudoaneurysm with a 0.9 cm pedicle of the right common carotid is seen on CT-angiography on sagittal (A) and axial (B) cuts.

(14%), iliac (13%) and abdominal aorta (11%) (3). Vascular manifestations are the main predictors of mortality and morbidity in BD, the survival rate was significantly lower in patients with arterial involvement than in those without arterial lesions ($p < 0.0001$) (3, 4). Recently, according to the recommendation of the European League Against Rheumatism (EULAR) 2018 for management of BD, medical treatment with glucocorticoid and cyclophosphamide is necessary for vascular BD involvement (5). The literature has shown that clinical immunosuppressive treatment is necessary before surgery or stenting, and should be continued after the procedure to avoid early and late post-procedure complications (4, 6-8), however invasive treatment should not be delayed if the patients are symptomatic.

Unfortunately, there is no universally accepted method to evaluate the disease activity in these patients, and there is yet no consensus in the literature about the best time for vascular correction. Kalko *et al.* considered to be in active stage of the disease, patients with an

ESR >15 mm/h, CRP >5 mg/L, and/or white blood cell count $>10,000/L$, so this patients received immunosuppressive therapy to induce remission for at least 15 days before the surgery (4). Liu *et al.* also recommend aggressive immunosuppression therapy with the objective of normalising the values of ESR and CRP in the preoperative (9). Although the inflammatory tests are used as activity parameter to choose the best time to perform the aneurysm correction, Kim *et al.* reported two cases of pseudoaneurysms developed at the puncture site in patients with normal ESRs (7) and Saadoun *et al.* described that only 85% of patients had elevated inflammatory markers at diagnosis of arterial involvement (3). Therefore, this may reflect a limitation of using this test to assess disease activity in patients with BD.

Treatment options in patients with arterial aneurysm include endovascular and surgical intervention, and there are no established safe surgical treatments for vascular BD. Park *et al.* showed that a positive pathergic reaction at the time of surgery and surgeries that

were not followed by treatment with glucocorticoids and immunosuppressive agent was an independent risk factor for the occurrence of postoperative complications (10), including suture pseudoaneurysm formation at anastomotic and graft occlusion (4, 7, 10). Liu *et al.* concluded that surgical treatment of arterial complications in BD was unsatisfactory, suggesting that endovascular treatment could provide a viable and less invasive alternative to surgery and may prove to be the treatment of choice in patients with major risk factors for open surgery (9).

Some case series have shown that endovascular treatment of aneurysms with stent placement is safe and effective with a long durability when disease activity is strictly controlled (6-9). Thus, pseudoaneurysms at margin of stent, such as in angiographic punctures sites, can recur in BD patients even after endovascular grafting (6-9, 11); postoperative follow-up with images method after vascular intervention is thus recommended.

Many investigators have reported some

Table I. Endovascular treatment of carotid artery aneurysms in patients with Behçet's disease.

Author (Year)	Age (yrs) / Sex	Location of the aneurysm	Endovascular technique	Follow-up (months)	Time and Drugs prior to endovascular intervention	Outcome
Present study	37 / M	Right CCA	Stent graft	5	15 days before intervention; Methylprednisolone 500mg /day for 3 days + Prednisone 60mg/day + Colchicine 1mg/day	Good evolution
Takamiya <i>et al.</i> (2016)	40 / M	Right ICA	Stent graft and coil embolisation	Not reported	Not reported	Good evolution
Yamaoka <i>et al.</i> (2015)	44 / M	Right ICA	Coil embolisation	24	Not reported	Good evolution
Ozveren <i>et al.</i> (2009)	36 / M	Left ICA	Coil embolisation	9 days	Not reported	Good evolution
Ohshima <i>et al.</i> (2008)	56 / M	Left CCA	Stent graft	12	Not reported	Good evolution
Agrawal <i>et al.</i> (2007)	32 / F	Right ICA	Coil embolisation	12	Not reported time; Course of high dose of prednisolone	Good evolution
Caballol <i>et al.</i> (2005)	29 / M	Left ICA	Stent graft	27	Not reported time; Corticosteroids + Immunosuppressive	Good evolution
Koo <i>et al.</i> (2003) // Kim <i>et al.</i> (2009)	32 / M	Right CCA	Stent graft	6	4 days before intervention; Prednisone 60mg/day + Azathioprine 100mg/day	Stent graft occlusion and pseudoaneurysm of the femoral artery
Park <i>et al.</i> (2001)	32 / M	Right CCA	Stent graft	6	Not reported	Stent graft occlusion
Bonnotte <i>et al.</i> (1999)	47 / F	Left ICA	Stent graft and Coil embolisation	Not reported	Not reported time; Azathioprine 100mg/day	Good evolution

M: male; F: female; ICA: internal carotid artery; CCA: common carotid artery.

successful endovascular treatment of carotid artery aneurysm (6, 7, 11, 12-17) (Table I). Only 2 reports evolved with unfavourable outcome, one of them related to discontinuation of immunosuppressive medication and the other probably due to the disturbance of the flow caused by double lesions in a small diameter vessel (6, 17).

Conclusion

Surgery intervention should be avoided for arterial lesions in the acute phase of inflammation whenever possible due to frequent postoperative complications. Pre- and post-operative immunosuppressive therapy should be always performed. To prevent complications, endovascular intervention has given promising results. However, there are still many questions that need to be answered in the future to improve the management of patients with BS: the choice of the type of intervention, the necessary time of immunosuppression before the vascular correction, dose

and the optimal duration of immunosuppressants after vascular intervention.

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