

## Why not to look for a vasculitis etiology in a patient presenting with coronary aneurysm, aortic pseudoaneurysm, and deep vein thrombosis such as Behçet's disease?

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We read with great interest the manuscript of Megaly et al.<sup>[1]</sup> in which the authors presented a case with ascending aortic pseudoaneurysm who was treated with percutaneous endovascular techniques. However, we would like to mention certain issues that should be discussed in the paper.

The patient was a 40-year-old male with a history of a 7-cm aneurysm of the left anterior descending artery (LAD) treated with excision of the aneurysm and coronary artery bypass grafting with the use of a saphenous vein graft. He presented to the clinic with deep vein thrombosis and a large pseudoaneurysm of the ascending aorta two years later.<sup>[1]</sup> Considering the patient's young age and atypical presentation, it is interesting not to see any work-up for a possible vasculitis syndrome, such as Behçet's disease in the manuscript.

Behçet's disease is an autoimmune vasculitis, mostly encountered at the third and fourth decades of life affecting both venous and arterial systems.<sup>[2]</sup> Cardiovascular involvement of the disease is the major determinant for the mortality and the morbidity of the patients. Superficial and deep vein thrombosis are the most frequent vascular pathologies and affects approximately 15 to 40% of the patients.<sup>[3]</sup> In addition, arterial true and false aneurysms are frequent in the course of the syndrome. They may occur spontaneously, being more common at

the surgical or percutaneous intervention sites.<sup>[4]</sup> Any emergency intervention should be carefully performed with the administration of high doses of steroids preoperatively which should be converted to long-term immunosuppressive therapy with close follow-up.<sup>[2,3]</sup>

It is not clear in the manuscript that whether the authors suspected any vasculitis syndromes and performed any histopathologic examination from the excised LAD aneurysm material. The left internal thoracic artery to LAD bypass is the gold standard revascularization method for the myocardium, which was not preferred by the authors, and revascularization was performed with the use of a saphenous vein graft. Was the proximal anastomosis of the saphenous vein performed on to the aorta? Did the authors histopathologically investigate the excised presumably healthy aortic tissue?

According to the manuscript, aneurysm resection and coronary revascularization was performed under cardiopulmonary bypass. However, we were not able to see the saphenous vein graft to LAD bypass on the angiography image. It would be more informative for the readers, if the authors presented the selective coronary angiography images to clearly understand the longevity of the saphenous vein graft. It is even more important, if the patient had Behçet's disease or another immune-mediated systemic vasculitis disease.

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What was the exact origin location of pseudoaneurysm at the ascending aorta? Could it be the anastomosis site of the saphenous vein or could it be the ascending aorta cannulation site when it was 4 cm above the sinotubular junction?

The last issue about the paper is the femoral puncture sites and we believe these regions should be carefully monitored for possible arterial pseudoaneurysms or occlusions in the long-term follow-up.

Once again, authors are to be congratulated for the successful management of their challenging case performed by endovascular stent grafting of the ascending aorta.<sup>[1]</sup> However, it would be more informative, if the authors could clarify the issues aforementioned above.

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#### Authors' reply

We read with interest the Letter to the Editor regarding our manuscript titled "Percutaneous thoracic endovascular aortic repair of an ascending aortic pseudoaneurysm".<sup>[1]</sup> First, we thank the authors for their knowledgeable comments on our paper.

In our case, the patient presented with a coronary aneurysm, which is a rare presentation for a middle age male with no diagnosed underlying disease in the past. Vasculitis was considered, but work-up was not mentioned in the original manuscript, as it is out of the scope of the learning objectives. Our patient underwent

extensive work-up for common known causes of vasculitis such as titers for ANA, RPR, MPO Ab, and Protein 3 Ab which were all negative. The C-ANCA and P-ANCA patterns suggestive of small-to-medium vessel vasculitis were not detected. Vasculitis causing infectious work-up such as HIV, Hepatitis C, and B virus were also negative. Behçet's disease may also present as a vascular stenosis, aneurysm or thrombosis similar to our patient for which hypercoagulability work-up was done revealing no significant factors deficiency acquired or inherited placing him at a higher risk of arterial and venous thrombosis.<sup>[2]</sup>

The diagnosis of Behçet's disease is merely based on the presentation of recurrent aphthous and genital ulcers with characteristic systemic manifestations, including vasculitis related end-organ damage.<sup>[2]</sup> The patient had no history of recurrent aphthous or genital ulcerations, eye symptoms suggestive of uveitis or skin lesions. The excised part of the left anterior descending artery (LAD) was sent for histopathological analysis, which showed pericardial tissue with necrosis, acute and chronic inflammation, granulation, fibrin deposits and fat necrosis with organizing blood clot. As per treating clinicians' discretion, though this was attributed to a trauma, it was not possible to rule out non-specific vasculitis manifesting with chronic inflammation such as Behçet's disease based on the pathological findings.

The original LAD aneurysm (Figure 1) was excised, proximal LAD was oversewn at the entry



Figure 1. Coronary angiogram showing left anterior descending aneurysm.

point with distal vein graft bypass (SVG). The surgeon preferred using an SVG over a left internal mammary graft, and the choice was made intraoperatively due to the urgency of the surgery and for anatomical reasons. Later on, the ascending aortic pseudoaneurysm ensued at the site of SVG anastomosis.

After three months, the patient complained of a left groin swelling and pain for which arterial Duplex was done and showed a 2×1-cm pseudoaneurysm of the left femoral artery at the site of vascular access. Under general anesthesia, open exploration, debridement, and repair of the aneurysm were done.

In summary, to diagnose Behçet's disease, the patient has to fulfill two of the five international Behçet's diagnostic criteria including recurrent oral aphthous ulcers, vascular thrombosis or pseudoaneurysm, skin findings, eye symptoms, pathergy test positivity or genital ulceration. Although our patient lacks the other requirements, the patient was recommended for follow-up with rheumatology for further diagnosis and treatment of a possible vasculitis syndrome.

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