Vascular Behcet's disease with multiple arterial aneurysms

A 30-year-old man presented with history of swelling in the left infraclavicular region and upper abdominal discomfort after food intake. He had a history of Behcet’s disease (BD) diagnosed 4 years ago. His complaints related to BD were limited to recurrent aphthous ulcerations in the genital region and oral mucosa. Examination revealed aneurysms of the left subclavian artery (figure 1A) and abdominal aorta. Selective subclavian artery angiogram revealed 15×40 mm fusiform aneurysm (figure 1B, see online supplementary movie 1). A 64-slice CT angiogram showed fusiform aneurysm of the upper abdominal aorta (73×40×42 mm) and a partially thrombosed saccular aneurysm of the superior mesenteric artery (67×78×75 mm). The right common femoral artery and both superficial femoral arteries were totally occluded (figure 2A, B). An aortic angiogram was done, which showed fusiform aneurysm of the abdominal aorta and partially thrombosed aneurysm of the superior mesenteric artery (figure 2C, D, see online supplementary movie 2). While planning for endovascular repair he developed sudden rupture of aortic aneurysm and expired.

BD is a rare form of vasculitis. The frequency of vascular involvement is estimated to range from 2–46%. Its vascular manifestations are thrombophlebitis, and less frequently arterial lesions such as occlusions or stenoses and pseudoaneurysms.1

The abdominal aorta is the most frequent site for pseudoaneurysm. Aneurysm of the superior mesenteric artery is very rare. The vascular involvement significantly affects the patient’s morbidity and mortality. The major cause of death in BD is rupture of pseudoaneurysm.2

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Figure 1 (A) Swelling seen in the infraclavicular region (arrow). (B) Selective subclavian artery angiogram showing fusiform aneurysm (15×50 mm).

Figure 2 (A) 64-slice CT angiogram anteroposterior view showing fusiform aneurysm of abdominal aorta (73×40×42 mm) and saccular aneurysm of superior mesenteric artery (67×78×75 mm). Right common femoral and both superficial femoral arteries occluded with distal reformation. (B) 64-slice CT angiogram—lateral view: saccular aneurysm of superior mesenteric artery is clearly seen. (C) Aortic angiogram—lateral view: showing abdominal aortic aneurysm (aneurysm wall marked). (D) Aortic angiogram—anteroposterior view: showing abdominal aortic aneurysm superiorly and superior mesenteric artery aneurysm inferiorly (aneurysm wall marked). AAA, abdominal aortic aneurysm; SMA, superior mesenteric artery aneurysm.