50. VASCULAR BEHÇET’S DISEASE IN A 20-YEAR-OLD MAN PRESENTING WITH AN ABDOMINAL AORTIC ANEURYSM

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Introduction: Behçet’s Disease (BD) is a complex, multisystem autoimmune disorder. The most frequent manifestations are oral aphthous ulcers, genital ulcers and uveitis. Vascular involvement is less common but one of the major causes of mortality and morbidity. Unlike the international study group (ISG) criteria for Behçet’s disease the international criteria for Behçet’s disease (ICBD) do include vascular lesions. The possibility of BD needs to be considered, especially in patients presenting with potential vascular involvement. For both aortic and peripheral artery aneurysms the 2018 update of the EULAR guidelines recommend medical treatment with cyclophosphamide and corticosteroids before surgical intervention. If there is ongoing arterial disease activity despite cyclophosphamide anti-TNF and IL6 inhibition can be considered and have shown some success. Endovascular surgery is increasingly being preferred to open surgery, although there is a risk of further aneurysm formation, particularly at graft peripheries.

Case description: A 20-year-old man with a Moroccan father, presented to A&E with abdominal pain, constipation and night sweats, following recurrent admissions with lower abdominal and back pain. On this occasion he was noted to have a CRP of 160 mg/L and a subsequent CT scan of the abdomen showed a 4 cm pseudo-aneurysm arising from the right lateral infrarenal abdominal aorta which was initially repaired with open surgery and a vein graft. This subsequently leaked and so he had further surgery and an EVAR procedure.

On further questioning it was noted that he had had frequent mouth ulcers for seven years and also episodes of epididymo-orchitis and folliculitis and the possibility of BD was considered. He was seen in the Birmingham Behçet’s Centre of Excellence and started azathioprine together with oral prednisolone. Six months later he required embolization of a left renal artery pseudoaneurysm and stent-grafting of the same artery. Given these new changes he was commenced on IV cyclophosphamide.

Despite six pulses of cyclophosphamide and high doses of prednisolone he continued to have a raised CRP and so was switched to tocilizumab infusions. His inflammatory markers initially improved, but after about two years he attended for an infusion and complained of worsening abdominal pain. At that point it was noted that his inflammatory markers had been intermittently raised over the preceding few months. There had been a few breaks in treatment due to possible infections and non-attendance. A further CT scan was performed that showed disease progression at the distal site of the graft with small new aneurysms. After an MDT discussion it was decided to switch him to infliximab. He has now been on infliximab for over six months and his inflammatory markers have, reassuringly, remained low.

Discussion: Arterial involvement in BD is relatively uncommon, presenting variably with stenosis, thrombotic occlusions and aneurysms, as the vasculitic process can affect perivascular and endovascular tissues. Aneurysms are typically seen a number of years after the initial diagnosis and are believed to be due to an obliterator endarteritis within inflammation causing destruction of elastic and muscle cells in the media. The aneurysms tend to be more saccular, and the abdominal aorta is more frequently involved than the thoracic aorta or pulmonary artery.

In this case the initial surgical procedure was performed prior to the diagnosis of BD, so it was not possible to give something such as cyclophosphamide prior to surgical intervention which would be advised. The EULAR guidelines recommend cyclophosphamide and corticosteroids as first line in treatment in arterial involvement. However, no further guidance is given regarding subsequent immunosuppression if needed. In this case the decision to start tocilizumab was made based on successful use in other patients with vascular Behçet’s disease.

Subsequently infliximab was chosen as it has been used more widely in BD including in patients with vascular BD. Biochemically and symptomatically the patient has been well since infliximab was started, but we are currently awaiting a repeat CT scan. This case has also proved challenging as the patient has struggled with the diagnosis and need for frequent hospital appointments. He has had some help from the psychologist available through the Behçet’s service but has continued to DNA appointments and turn up for infusions at times that suit him. We are hopeful that a longer period of stability will also be beneficial from that point of view.

Key learning points: Vascular is less common than other features of BD but can sometimes be a presenting feature and is a major cause of mortality and morbidity. Unlike the international study group (ISG) criteria for Behçet’s disease the international criteria for Behçet’s disease (ICBD) do include vascular lesions. The possibility of BD needs to be considered, especially in patients presenting with potential vascular involvement. For both aortic and peripheral artery aneurysms the 2018 update of the EULAR guidelines recommend medical treatment with cyclophosphamide and corticosteroids before surgical intervention. If there is ongoing arterial disease activity despite cyclophosphamide anti-TNF and IL6 inhibition can be considered and have shown some success. Endovascular surgery is increasingly being preferred to open surgery, although there is a risk of further aneurysm formation, particularly at graft peripheries.

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