aneurysm wall of BD patients are adventitial thickening and fibrosis, perivascular lymphocytic infiltration, elastin and muscle fibre decrement in media, smooth muscle fibre and fibroblast increment in intima. These vessel wall changes lead to formation of true aneurysms by wall distension or pseudoaneurysm by wall perforation [2]. The aneurysms in BD frequently involve medium- and large-sized vessels such as aorta, femoral, pulmonary, iliac and popliteal arteries. Rupture is the most common presenting symptom besides being the most common cause of vascular-related deaths [3].

Pseudoaneurysm formation at the site of arterial puncture for angiography or endovascular treatment has been reported in BD patients [4]. Consequently, non-invasive interventions such as USG, CT and magnetic resonance angiography imaging are recommended rather than conventional angiography for patients to whom endovascular treatment is not being considered [4].

On physical examination, pulsatile mass on the trace of peripheral artery or an intraabdominal mass can be palpated. A systolic murmur on the mass can also be heard. The clinical findings in aneurysms are related to localization, size and growth rate. In general, patients refer after a vascular surgical intervention or with a pulsatile mass, which makes it easy to diagnose. Delayed cases can have local erythema and tenderness as well as nerve and vein compression symptoms due to growth [5].

Previously, open surgical repair was the definitive treatment for vascular lesions, such as aneurysms, in patients with BD. However, the success rate of surgical management has not been high because a false aneurysm often occurs after surgical repair. Recently, endovascular stent graft seems to be more successful than surgical intervention because of lesser complication risk and is suggested in treatment of peripheral aneurysms in BD [6]. As it is less invasive, the morbidity is also lower than the open surgical technique [5]. These are particularly important advantages in surgical candidates with high risk.

Vascular aneurysms should be kept in mind in patients with BD admitted to emergency services. Leg pain, which is frequently seen and diagnosed as myalgia, can be due to a giant aneurysm in these patients. Checking for peripheral pulses should be an important part of physical examination as it can be weaker in affected side forming an important clue for diagnosis.

CONCLUSIONS

Vascular problems in BD, like aneurysm formation, may occur in various states including leg pain. The findings in our case suggest that endovascular therapy combined with immunosuppressive treatment for aortic pseudoaneurysms in patients with BD appears to be a promising alternative and effective management option. A longer follow-up is required to allow confidence of lasting success.

Conflict of interest: none declared.

REFERENCES


eComment. Behçet’s disease or Adamantiades-Behçet’s disease?

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doi:10.1093/icvts/ivr155
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Memetoglu and Kalkan, in their well-written manuscript have reported on the successful percutaneous treatment of an aneurysm in a patient with Behcet's disease [1]. We agree with the consideration of a percutaneous therapeutic modality instead of conventional surgical treatment for the management of aneurysms in Behcet's disease and would like to add a brief comment from a historical perspective.

In 1956, the ophthalmologist Feigenbaum highlighted the first description of Behcet's syndrome by Hippocrates of Kos [2,3]. According to Feigenbaum, Hippocrates wrote in his third “Epidemion” book, case 7, “But there were also other fevers, as it will be described. Many had their mouths affected with aphthous ulcerations. There were also many defluxions about the genital parts, ulcerations, boils (phymata), externally and internally about the groins. Watery opthalmies of a chronic character, with pains, fungous excrescences of the eyelids, externally and internally, called fici, which destroyed the sight of many persons…” [2,3].

Zouboulis and Keitel, in their well-documented historical review, showed that the Greek ophthalmologist Benediktos Adamantiades (1875-1962) brought the ocular signs, genital ulcers and the arthritis in connection with a single disease [3]. On 15th of November 1930, Adamantiades presented a case report entitled “A case of relapsing iritis with hypopyon” at the annual meeting of the Medical Association of Athens and this presentation was published in the Proceedings of the Medical Society of Athens [3,4]. The case report was related to a 20 year old male, whose first clinical symptoms were edema and ulcerations of his left leg with a primary diagnosis of thrombophlebitis. In a two years follow-up, the patient developed recurrent iritis with hypopyon in both eyes, resulting in blindness and atrophy of the optic nerve, scrotal ulcers, oral aphthous ulcers, and sterile arthritis of both knees. The last three signs were recurrent and the bacterial cultures of the knees and eyes were negative [3,4].

Adamantiades went on to publish this case report in the French journal Annales d’Oculistique in 1931 [5]. In 1946, Adamantiades reported on two further patients and defined thrombophlebitis as a fourth sign of the disease [3]. In 1953, he presented the first classification of the disease and proposed the first diagnostic criteria and in 1958, his last work regarding its neurologic complications [3] was published.

On 11th of May 1937, Hulsi (Hulusi) Behcet (1889-1948) presented a case report of a 34 year old female with a seven-year history of recurrent oral aphthous ulcers, genital ulcers, and ocular lesions at a meeting of the Dermatological Association of Istanbul [3]. This case report with the addition of a similar one was published in 1937 [3]. Behcet published three more manuscripts from 1938 to 1940, drawing medical attention [3]. He was aware of the publications by Adamantiades and included these in his references.

Even though the current World Health Organization/ICD-10 standard is Behcet’s disease, we believe that the term Adamantiades-Behcet’s disease honors both first describers (in the modern era) for their contributions [3].

Conflict of interest: none declared.

References