Rheumatology International (2019) 39:1061–1067
https://doi.org/10.1007/s00296-019-04283-y

Rheumatology
INTERNATIONAL

OBSERVATIONAL RESEARCH

# Clinical course of abdominal aortic aneurysms in Behçet disease: a retrospective analysis

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Received: 29 January 2019 / Accepted: 13 March 2019 / Published online: 19 March 2019


## Abstract

Arterial aneurysms are rare manifestations of Behçet Disease (BD) with high morbidity and mortality. This study aimed to investigate the clinical course of BD patients with abdominal aortic aneurysms (AAA). We retrospectively searched charts of BD patients, followed up between 1988 and 2011, to identify those with AAA with at least 6-month clinical and radiological follow-up data. Chart review revealed 12 patients (11 males) with AAA amongst 1224 patients; follow-up data from 11 patients were available. The most common symptoms were lower back and abdominal pain. The only pre-treatment complication was a spontaneous rupture. All but one patient received corticosteroid and cyclophosphamide pulses for the induction, and corticosteroid and azathioprine for the maintenance treatment; one patient received only the maintenance treatment. Two patients had surgical graft interposition, without postoperative complications. Seven patients had endovascular stenting; five of them (71.4%) showed radiological regression after 32.5 (13.4–53.8) months, while four (57%) had clinical improvement after 11.8 (0.2–29.4) months. However, one non-responsive patient developed stent infection and exsanguinated during percutaneous drainage, and one patient developed femoral artery pseudo-aneurysm at the catheter insertion site. Another patient developed a new aneurysm under the maintenance treatment. Medical treatment alone yielded radiological regression in one of two patients. Current immunosuppressive, surgical or endovascular approaches can provide clinical and radiological improvements lately in BD patients with AAA. Furthermore, complication rates seem to be high with interventional approaches. These findings suggest an unmet need for safer alternative treatments.

**Keywords** Behçet disease · Vasculitis · Aortitis · Abdominal aortic aneurysm · Endovascular stent · Surgical graft interposition

## Introduction

Behçet disease (BD), a multi-system inflammatory disorder of unknown etiology, is classified as "variable vessel vasculitis" with a particular predilection to both venous and arterial sides of systemic as well as pulmonary circulations, with a lower frequency for arterial side (1–12%) [1–4]. The most common form of arterial disease is characterized by pseudo-aneurysm formation, and abdominal aortic aneurysms are responsible for approximately one-third of all arterial aneurysms [4–7].

Despite the availability of some studies on the characteristics of pulmonary arterial aneurysms, peripheral arterial aneurysms, or overall arterial involvement, detailed information about the course of AAAs in BD is scarce. Also, no randomized controlled trial data are available about the management of arterial aneurysms in BD, and the current

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approaches are largely based on case reports and expert opinions [8, 9]. In the light of emerging data on the treatment and prognosis of AAAs due to atherosclerosis, we herein aimed to identify the distinctive clinical features and course of AAAs in BD patients.

## Methods

### Patients

Charts of BD patients followed up in the outpatient clinic between 1988 and 2011 were retrospectively screened to identify those with well-documented AAA. All screened patients fulfilled the classification criteria for BD according to the International Study Group Diagnostic Criteria [10].

Imaging data obtained by computerized tomography (CT) and/or magnetic resonance imaging (MRI) were used for the diagnosis and radiological follow-up of AAAs, and patients who had at least 6 months of clinical and radiological follow-up were included in the analysis. An expert radiologist reviewed all of the available CT and MR images, and chart notes were also used for the missing data during the course.

A standard form was used to record the clinical findings, treatment and follow-up data. Data were collected from patients' charts, face-to-face interviews, and telephone contacts were used when possible. The last visits were done between February 2011 and April 2012.

## Results

There were 27 patients (25 males) (2.2%) with non-pulmonary arterial aneurysms amongst 1224 BD patients. Twelve (11 males) (44.4%) of them had AAAs. Follow-up data were available in 11 patients for retrospective review.

Median age of patients was 43 (21.4–47.6) years when AAA was diagnosed. The median time elapsed between onset of BD symptoms and the diagnosis of AAAs was 7.3 (2.5–20.6) years, and all patients developed other manifestations of BD before the appearance of AAA. Median follow-up time from the onset of AAA was 4.3 (0.7–9.8) years. Pulmonary and venous involvements were present in 9% and 72% of the patients, respectively.

Lower back pain was the most frequent symptom of AAA (72%), followed by abdominal pain (54%), groin pain (18%), and weight loss (9%). One patient was asymptomatic at the time of diagnosis, who was being treated for pulmonary artery aneurysms, and AAA was incidentally found on a follow-up imaging.

All aneurysms were in the infra-renal segment of the abdominal aorta, except for one patient who had a second aneurysm located superior to the celiac trunk (Fig. 1A). The median transverse diameter of aneurysms was 4.3 (1.3–8.2) cm. All aneurysms were saccular and irregular in shape, and contained mural thrombus. Erosive changes were seen in the lumbar vertebral body adjacent to AAA in one patient (Fig. 1b).

The median erythrocyte sedimentation rate was 35 (6–108) mm per hour in seven patients who had this measurement prior to any treatment.

All patients, except for three, received immunosuppressive agents for the induction (IV methylprednisolone and cyclophosphamide pulses) and/or maintenance (oral methylprednisolone plus azathioprine or cyclophosphamide) treatments (Table 1). One patient (patient 10) developed nystagmus and nausea/vomiting after the first dose of 1000 mg of intravenous cyclophosphamide, and these adverse effects were eliminated by reducing the dose to 750 mg/month. Oral methylprednisolone could be tapered to 4–6 mg/day in all but one patient (patient 9). Of seven patients who had endovascular stenting (EVS), six received EVS plus induction and maintenance and one had EVS plus maintenance regimen.

Two patients underwent surgical resection and polytetrafluoroethylene (PTFE) graft interposition due to spontaneous rupture of their aneurysms (one was severe, and the other was contained rupture). Induction followed by maintenance immunosuppressive treatments was given immediately after the surgical interventions. One of the two surgical interventions was done in another center. Their postoperative follow-ups were eventless and symptom free.

All EVSs, except for one, were inserted by the same radiology–surgery team in our center. Five of seven patients (patients 3, 4, 5, 6, and 7), who had EVS showed complete radiological resolution of pseudo-aneurysm findings after a median of 32.5 (13.4–53.8) months following the intervention, while four patients (patients 3, 4, 5, and 7) experienced symptomatic improvement after a mean of 11.8 (0.2–29.4) months following the intervention. One patient (patient 6), who remained symptomatic despite radiological response, developed femoral artery pseudo-aneurysm at the catheter insertion site (Fig. 1C), and died of cervix cancer after 2.7 years of follow-up. Two patients (patient 5 and 7), who had complete radiological and symptomatic response, started to complain from lower back pain when bending forward after EVS.

Patient 8 had also familial Mediterranean fever (FMF) with compound heterozygous p.Met680Ile/p.Val726Ala mutations in the MEFV gene. His initial induction immunosuppression was started in our clinic, but he continued to his treatment at another center and had EVS intervention. According to the data retrieved from that center, he developed stent infection and abscess around the stent following EVS (Fig. 1d), and fatal intra-abdominal

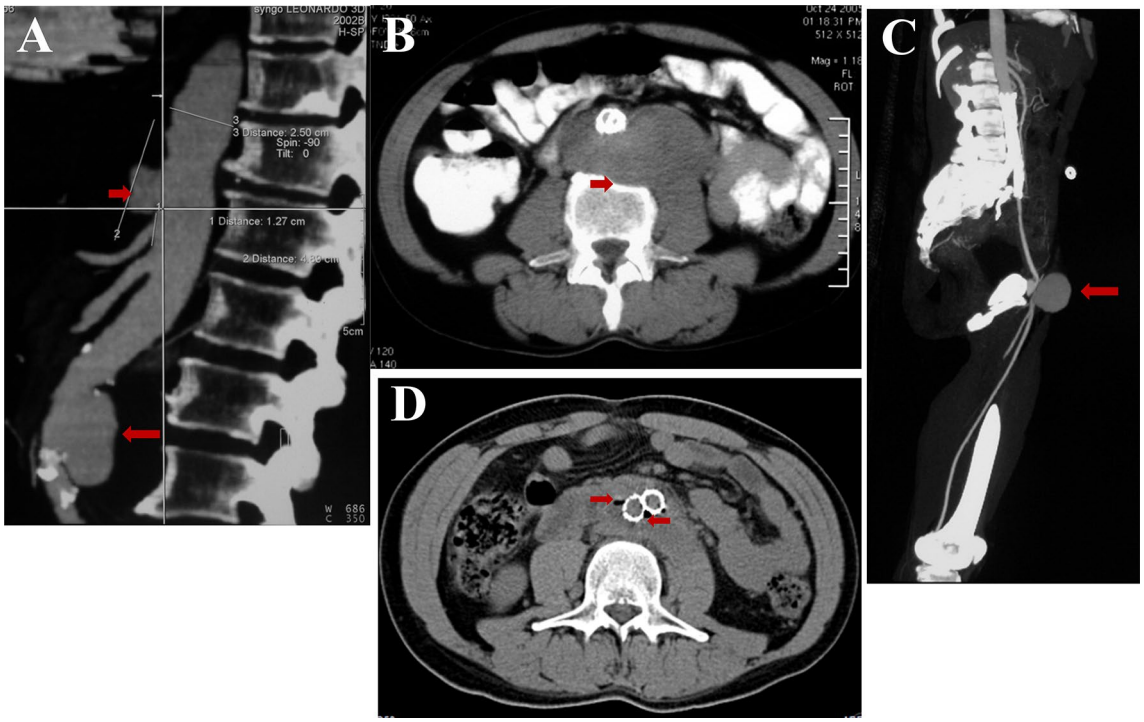


**Fig. 1** **a** Sagittal CT image of a patient with two aortic aneurysms at the celiac trunk (short arrow) and the infarenal segments (long arrow). **b** Axial CT image of an erosion of a lumbar vertebral body (short arrow) in a patient with BD. **c** Three-dimensional reconstruction CT angiography of a femoral artery pseudo-aneurysm that developed at the catheter insertion site (long arrow). **d** Axial CT image showing air bubbles (arrows) within the aneurysmal pouch due to stent infection, which was developed following endovascular stent insertion at another center

hemorrhage occurred as a result of percutaneous abscess drainage attempt.

Patient 9 had a history of surgical resection and graft interposition for his right common iliac artery aneurysm 4.7 years before the development of AAA, and remained without follow-up after a short course of corticosteroids and colchicine postoperatively. Induction treatment was followed by EVS and cross-femoral bypass operation because of continuing symptoms under immunosuppressive treatment. One month later the stent was found to be thrombosed, and the patency was reinstituted via transcatheter alteplase infusion. However, the patient's symptoms, elevated acute phase response and the size of the aneurysm did not improve despite 17.4 months of follow-up under standard immunosuppressive treatment. Furthermore, a new aneurysm (3 cm by 4 cm) developed at the proximal site of the endovascular stent. FMF was suspected as a possible contributor to his higher inflammatory response and refractory disease course, but only a functional polymorphism of p.Glu148Gln in heterozygous state was detected in the MEFV gene analysis. Azathioprine was stopped, and daily injections of anakinra (a recombinant IL-1 receptor antagonist) was started. His symptoms were dramatically improved within 10 days along with later normalization of acute phase response, and the dose of corticosteroid could be tapered to 4–6 mg/day. The original aneurysm disappeared at the 35th month of follow-up, whereas the aneurysm that developed at the proximal site of the EVS expanded to 3.5 cm by 4.4 cm during the follow-up.

Radiological follow-up data permitted to analyze the response of six aneurysms in five patients to immunosuppressive agents prior to EVS; the initial median transverse diameter was 4.5 (2.0–7.0) cm and it expanded to 7.15 (4.2–11.5) cm after a median of 7.3 (4–57.1) months under immunosuppression.

Two patients did not receive any surgical intervention. Patient 10 had an aneurysm of 1 cm by 1 cm, which showed complete resolution after 47.2 months under immunosuppression. Patient 11 had an aneurysm of 3.5 cm by 4.7 cm, and his abdominal and lower back pains persisted, along with expansion of the aneurysm to 4 cm by 4.8 cm despite 56.8 months of immunosuppressive treatment.

## Discussion

In this study, AAA was observed at frequencies similar to the previous reports as a late manifestation of BD mainly observed in males [1–3, 6, 7]. Male predominance has also

**Table 1** Summary of the clinical, radiological, and treatment features of the patients

| Patient's number (age, sex) | Diameter (cm) | Induction prior to intervention | Intervention | Induction after intervention | Maintenance treatment | Radiological response | Symptomatic response | Complication | Total follow-up time (year) |
|---|---|---|---|---|---|---|---|---|---|
| 1 (38.3 M) | 4.2 | No | Surgery | Steroid (3 g in 3 days MP)/cyclophosphamide (1 gr/month for 12 months) | Steroid (60 → 4 mg/day MP) + cyclophosphamide (100 mg/day) (oral) | N/A | Yes | No | 7.2 |
| 2 (43 M) | 4.5 | No | Surgery | Steroid (3 g in 3 days MP)/cyclophosphamide (1 gr/month for 7 months) | Steroid (60 → 4 mg/day MP) + AZA (150 mg/day) | N/A | Yes | No | 4.7 |
| 3 (47 M) | 4.0 | No | Stent | Steroid (3 gr in 3 days MP)/cyclophosphamide (1 g/month for 10 months) | Steroid (40 → 6 mg/day MP) + AZA (100 mg/day) | Yes | Yes | No | 4.1 |
| 4 (44.8 M) | 7.0 | Steroid (40 → 4 mg/day MP) + AZA (150 mg/day) (maintenance) | Stent | No | Steroid (40 → 4 mg/day MP) + AZA (150 mg/day) (maintenance) | Yes | Yes | No | 4.3 |
| 5 (47.5 M)<br>Two aneurysms | 2.0<br>4.0 | Steroid (3 gr in 3 days MP)/cyclophosphamide (500 mg/month for 2 months) | Stent | Steroid (32 → 4 mg/day MP) + cyclophosphamide (75 mg/day for 23 months) then AZA (125 mg/day for 32 months) (treatment before stenting) | Steroid (4 mg/day MP) + AZA (125 mg/day) | Yes | Yes | No | 9.7 |
| 6 (33.6 F) | 4.2 | Steroid (1500 mg in 3 days)/cyclophosphamide (1 gr/month for 7 months) | Stent | No | Steroid (32 → 4 mg/day MP) + AZA (100 mg/day) | Yes | Yes | Femoral artery aneurysm at the catheter insertion site | 2.7 |
| 7 (45 M) | 3.3 | Steroid (3 gr in 3 days MP)/cyclophosphamide (1 gr/month for 7 months) | Stent | Cyclophosphamide (1 g/month for 5 months) | Steroid (32 → 4 mg/day MP) + AZA (150 mg/day) | Yes | Yes | No | 3.5 |
| 8 (29 M) | 5.0 | Steroid (3 gr in 3 days MP)/cyclophosphamide (1 gr/month for 8 months) | Stent | Steroid (60 → 16 mg/day/cyclophosphamide (500 mg/month total dose?) | Died | No | No | Stent infection | 0.7 |
| 9 (29.8 M) | 2.5 | Steroid (3 gr in 3 days MP)/cyclophosphamide (1 gr/month for 5 months) | Stent | Cyclophosphamide (1 g/month for 7 months) | Steroid (60 → 32 mg/day) + AZA (150 mg/day) after 9.5 months Steroid (8 mg/day MP) + Anakinra | No | No | New aneurysm formation at the proximal site of the stent | 4.7 |

**Table 1** (continued)

| Patient's number (age, sex) | Diameter (cm) | Induction prior to intervention | Intervention | Induction after intervention | Maintenance treatment | Radiological response | Symptomatic response | Complication | Total follow-up time (year) |
|---|---|---|---|---|---|---|---|---|---|
| 10 (21.4 M) | 1.0 | Steroid (3 gr in 3 days)/cyclophosphamide (first dose 1 gr, then 750 mg/month for 11 months) | No | No | Steroid (40 → 6 mg/day MP) + AZA (100/150 mg/day) | Yes | Yes | No | 6 |
| 11 (47.6 M) | 4.7 | Steroid (3 gr in 3 days)/cyclophosphamide (1 gr/month for 4 months) | No | No | Steroid (60 → 4 mg/day MP) + AZA (150 mg/day) | No | No | No | 4.9 |

*MP* methylprednisolone, *AZA* azathioprine, *M* male, *F* female

been reported in atherosclerotic AAAs, which are usually asymptomatic [11]. However, all BD patients were under 50 with no obvious risk for atherosclerosis; and their aneurysms were saccular and/or irregularly shaped and symptomatic. Pain was the most frequent symptom, and it was possibly due to vascular and perivascular inflammation, and it may persist after controlling the intravascular pressure and the risk of rupture by EVS. Symptoms and signs related to affection of neighboring organs due to perivascular inflammation could also occur, such as aorto-enteric fistula or erosion of vertebral body, as was seen in one of our patients [12–14].

A higher frequency of venous thrombosis in BD patients with arterial involvement was reported previously [2–4]. Similarly, 72% of patients in this study had signs of venous thrombosis. This observation could reflect common pathogenic mechanisms for arterial and venous involvements.

Pulmonary artery aneurysms in BD can rupture into bronchial space and cause massive hemoptysis resulting in 25–50% mortality [15, 16]. But no data have been published yet about the prognosis of AAAs in BD. The risks of rupture and, therefore, mortality of atherosclerotic AAAs are closely related to the transverse diameter of the aneurysm [17]. However, severe spontaneous rupture of AAAs in BD is rare, and it is probably be due to inflammation of the peri-aneurysmal tissues and fibrotic reactions [6, 18]. Two patients in this series had spontaneous ruptures prior to any treatment and one of them was considered severe enough to necessitate an emergency surgery. But both patients had enough time to undergo successful surgical intervention. There were two deaths in this study; one of them was due to cervix cancer developed later during the course of the disease, and the other was due to exsanguination during percutaneous drainage of an infected aneurysm, while no death due to spontaneous rupture of an AAA was seen. Saadoun et al. reported one death among 17 patients with AAA compared to three deaths out of eight patients with thoracic aortic aneurysms and three deaths out of 21 patients with pulmonary aortic aneurysms (4). Although these findings may suggest a better prognosis for AAAs compared to thoracic aortic and pulmonary arterial aneurysms in BD, the total numbers are relatively small for a conclusion and long-term follow-up data from larger series of patients are expected to clarify site-specific differences in the prognosis of BD patients with aortic aneurysms.

There has not been any randomized controlled trial so far to guide the treatment of AAAs in BD. Immunosuppressive agents and surgery are recommended for the treatment of non-pulmonary arterial aneurysms in BD by EULAR in 2008 and 2018 based on expert opinions [8, 9]. The updated 2018 recommendations suggested the medical treatment with cyclophosphamide and corticosteroids as necessary before intervention to repair, but if the patient symptomatic, surgery or stenting is advised without a delay [9].

Recurrence of aneurysms or hemorrhagic complications were not seen during early and late postoperative periods in two patients, who underwent surgery in this study. EVS plus immunosuppressive treatment provided late and limited rates of radiological and symptomatic success, at the expense of three major complications (Table 1). Furthermore, two patients had ongoing lower back pain, possibly due to the mechanical impact of EVS on the aortic wall with high pressure. When the literature was reviewed for surgical and endovascular approaches, it appeared that both of them were associated with high-risk complications [4–6, 19, 20]. In a retrospective analysis of 12 patients with AAAs due to BD reported by Kwon et al., the overall recurrence rate after surgical intervention was found 50%, with 14.3% after graft interposition, 62.5% after patch closure and 40% after EVS [21]. Although there may be variations in the efficacy of immunosuppressive regimens applied in different centers, these findings suggest that the current regimens may not be adequate and perhaps alternative treatment strategies are necessary.

One patient in our study and another patient from a recent report [5] had small AAAs that showed complete resolution with immunosuppressive treatment alone, whereas no regression in any of the seven aneurysms with a mean transverse diameter of 4 cm with immunosuppressive treatments was observed prior to EVS. Interestingly, only small pulmonary artery aneurysms in BD showed regression under immunosuppression [22]. These observations suggest that hemodynamic factors may also be involved in the process of healing, as the size of the aneurysm is directly related to wall stress according to Laplace's law [23]. Therefore, protection of large aneurysms from hemodynamic pressures seems to be necessary for the process of resolution. On the other hand, considering the continuation of symptoms even after intravascular hemodynamic control possibly due to perivascular inflammatory changes suggest the potential of additional benefits of more targeted treatments aiming proinflammatory cytokines such as TNF and IL-1 for better management of the patients with AAA [9, 24, 25].

## Conclusions

In conclusion, AAAs in BD patients are relatively late manifestations with high morbidity and mortality, but available information may suggest a better prognosis compared to throracic and pulmonary arterial aneurysms. Endovascular interventions are associated with relatively high complication rates despite effective immunosuppressive treatments, and symptomatic relief could be achieved lately with current treatment options due to ongoing perivascular inflammatory response. Alternative approaches targeting other critical inflammatory molecules are needed for achieving faster and more efficient symptomatic and radiological response in AAAs in BD.

**Author contributions** All of the authors participated sufficiently in the work to take public responsibility for appropriate portions of the content; and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved, according to the updated ICMJE authorship criteria in December 2018. Specifically, TS participated in the conceptual design of the study, acquisition and analysis of data and analyzed the data, and involved in the drafting of the manuscript; BAE involved in the acquisition and analysis of data, critically revised the manuscript; MA and MK involved in the endovascular treatments, analysis and interpretation of data, and critically revised the manuscript; AP participated in the endovascular treatments, analyzed and critically interpreted all radiological data, and critically revised the manuscript; AG conceptually designed and coordinated the study, critically analyzed and interpreted the data, edited and revised the manuscript. All authors read and approved the final version of the manuscript.

**Funding** No funding was received for this study.

**Data availability** The data supporting the conclusions of this article are included within the article and they are stored in the repository of the Medical Faculty patient records.

## Compliance with ethical standards

**Conflict of interest** AG received honoraria from Servier, AbbVie, MSD and Novartis and research grants from Xoma and Novartis. The author TS declares that he has no conflict of interest. The author BAE declares that she has no conflict of interest. The author MA declares that he has no conflict of interest. The author MK declares that he has no conflict of interest. The author AP declares that she has no conflict of interest.

**Ethical approval** The Ethics Committee of Istanbul Faculty of Medicine approved the study protocol (2012/764–1063 on 27 April 2012) and allowed the retrospective analysis of available records of the patients followed up in the outpatient clinic between 1988 and 2011.

**Ethical standards** All procedures performed in this study in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. The Ethics Committee of XYZ approved the study protocol (2012/764–1063 on 27 April 2012) and allowed the retrospective analysis of available records of the patients followed up in the outpatient clinic between 1988 and 2011. The Ethics Committee also consented to use the radiographic images without any identification if it is not possible to get permission from died patients or their next of kin. Patients gave written consent for being participated in the study and using their data and radiographic images. Next of kin of two died patients could not be traced, and their images were used without any identification with the approval of local ethics committee.

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