CASE REPORT

Behçet’s Disease and Popliteal Artery Aneurysm: a Case Report and Review of the Literature

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Key Words: Behçet’s disease; Popliteal aneurysm.

Introduction

Behçet’s disease (BD) is a multisystem disorder characterized by vasculitis. It was first described in 1937 by Hulusi Behçet, a Turkish dermatologist, and consists of a triad of recurrent ulcers of the oral and genital mucosa with relapsed uveitis.¹ Since the first description, additional organ involvements have been reported. Skin, central nervous system, kidney and pulmonary involvement, vasculitis, epididymitis, arthralgia, arthritis, gastrointestinal features, and AA type amyloidosis are other symptoms or signs seen in BD, and also a positive family history is an important criteria of diagnosis.²⁻⁷ Vascular involvement is the leading cause of death in BD with an approximate prevalence of 25%.⁵,⁸ Vascular involvement is more frequent in males than in females (36% versus 14%). Three forms of vascular disease (venous occlusions, arterial aneurysms and/or arterial occlusions) are found in BD. Venous lesions occur more frequently than arterial lesions (88% versus 12%). Symptoms of vascular disease vary depending on the site of involvement.¹,²,⁵ Subcutaneous thrombophlebitis is the most frequent (47%) type of venous involvement.³ The following most common venous lesions are superior and inferior vena cava occlusions.

Budd–Chiari syndrome is a relatively common complication of BD and the presence of hepatic vein thrombosis is one of the major prognostic factors affecting survival.⁹ Aneurysm and/or occlusion of the arteries of the upper or lower extremities are the most frequent type of arterial lesions.¹⁰,¹¹ Pulmonary arterial vasculitis presenting with dyspnea, cough, chest pain, and hemoptysis is not rare.¹² Ruptures of large arterial aneurysms may lead to death.

In this report, we present a patient with BD complicated by popliteal artery aneurysm and discuss the literature on BD in association with popliteal artery aneurysm.

Case Report

A 44-year-old white man was hospitalized because of a tender and pulsatile mass in the left popliteal fossa for a month. His other complaints were recurrent oral and genital ulcerations, erythema nodosum and acneiform lesions (for 8–10 years). The patient also noted to have somewhat left lower leg swelling for the past 3 months.

On admission, physical examination was as follows. The vital signs were essentially within normal limits. The oral mucosa had multiple aphthous lesions. The cardiovascular system was normal. The abdomen was benign. The left lower extremity was found to be

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Our patient had oral and genital aphtae, acneiform lesions and erythema nodosum which fulfilled the International Study Group (ISG) Criteria\textsuperscript{13} for BD. According to the history, physical examination, and laboratory findings; no other causative factors of popliteal artery aneurysm (polyarteritis nodosa, Takayasu’s disease, atherosclerosis, trauma, infection) other than BD were discovered.

There are few controlled studies reported on the management of BD. As BD usually runs an undulating course of exacerbations and remissions, it is generally difficult to evaluate the efficacy of therapy. The treatment of BD is symptomatic and varies according to the clinical manifestations.\textsuperscript{7,14} Although definitive data do not exist, there is a tendency among physicians to prescribe colchicine to all patients.

A controlled, long-term study regarding medical and surgical treatment of vascular BD has not yet been reported. Antiplatelet agents such as low-dose aspirin and diprydamole are recommended for venous involvement, but a controversy exists on this subject.\textsuperscript{5,14} Routine anticoagulation with heparin or oral anti-coagulants is not advised if deep vein thrombosis is not present. Severe vasculitis in BD is treated with systemic corticosteroids and immunosuppressives as in other forms of vasculitis.

It is generally accepted that surgical treatment of arterial aneurysms due to vasculitis is not favorable. Pathergy test is an abnormal tissue reaction to needle pricks and it is one of the ISG diagnosis criteria.\textsuperscript{13} Venous puncture, intravenous infusion, rapid injection of a large bolus of contrast material, and insertion of a venous catheter may initiate venous thrombosis in BD.\textsuperscript{15,16} An increased incidence of aneurysm formation at the puncture site has been reported after venography and arterial puncture.\textsuperscript{5,10,16,17} Besides, aneurysmal rupture results in severe haemorrhage with a death rate of 60\%.\textsuperscript{2} Since popliteal artery aneurysms in BD are at high risk of rupture, certain cases may warrant an emergency intervention. The efficacy of medical therapy to prevent from rupturing is not known.

According to our review, only 37 patients with BD and popliteal artery aneurysm have been described in the literature before (Table 1).\textsuperscript{2,5,15,16,37} The analysis of these cases has shown the following results:

1. The presence of another vascular lesion was a common feature.
Behçets Disease and Popliteal Artery Aneurysm

Table 1. Cumulative analysis of patients with Behçet’s disease and popliteal artery aneurysm.

<table>
<thead>
<tr>
<th>Author (Reference no.)</th>
<th>Year</th>
<th>No. of patients</th>
<th>Coexisting vascular lesion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Enoch B. A.⁶¹</td>
<td>1968</td>
<td>1</td>
<td>Subcutaneous thrombophlebitis</td>
</tr>
<tr>
<td>Chavataz D.¹⁹</td>
<td>1974</td>
<td>1</td>
<td>—</td>
</tr>
<tr>
<td>Jenkins A. M.²⁰</td>
<td>1976</td>
<td>1</td>
<td>Superficial femoral artery aneurysm</td>
</tr>
<tr>
<td>Piers A.²¹</td>
<td>1977</td>
<td>1</td>
<td>Subcutaneous thrombophlebitis, Deep vein thrombosis</td>
</tr>
<tr>
<td>Chamberlain M. A.²²</td>
<td>1977</td>
<td>1</td>
<td>—</td>
</tr>
<tr>
<td>Reza M. J.²³</td>
<td>1978</td>
<td>1</td>
<td>Femoral artery occlusion</td>
</tr>
<tr>
<td>Shimizu T.²</td>
<td>1979</td>
<td>1</td>
<td>—</td>
</tr>
<tr>
<td>Wang Z. G.²⁴</td>
<td>1985</td>
<td>1</td>
<td>Axillary and femoral artery aneurysms</td>
</tr>
<tr>
<td>Muftuolu U. A. U.²⁵</td>
<td>1986</td>
<td>3</td>
<td>—</td>
</tr>
<tr>
<td>Dhobb M.²⁶</td>
<td>1986</td>
<td>2</td>
<td>—</td>
</tr>
<tr>
<td>Yamana K.²⁷</td>
<td>1988</td>
<td>2</td>
<td>Left common femoral artery aneurysm (2)</td>
</tr>
<tr>
<td>Bartlett S. T.²⁸</td>
<td>1988</td>
<td>1</td>
<td>Recurrent femoral artery aneurysm</td>
</tr>
<tr>
<td>Koç Y.²⁹</td>
<td>1992</td>
<td>1</td>
<td>Carotid artery aneurysm</td>
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<td>Senzen E.²⁹</td>
<td>1992</td>
<td>2</td>
<td>Superficial femoral artery aneurysm (1)</td>
</tr>
<tr>
<td>Chaillou P.³⁰</td>
<td>1992</td>
<td>1</td>
<td>Posterior tibial artery aneurysm</td>
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<tr>
<td>Sherif A.³¹</td>
<td>1992</td>
<td>1</td>
<td>Femoropopliteal phlebitis</td>
</tr>
<tr>
<td>Kuzu M. A.³²</td>
<td>1994</td>
<td>3</td>
<td>—</td>
</tr>
<tr>
<td>Kaynak K.³³</td>
<td>1996</td>
<td>3</td>
<td>—</td>
</tr>
<tr>
<td>Tüzün H.³⁴</td>
<td>1997</td>
<td>4</td>
<td>Left posterior tibial artery aneurysm (1)</td>
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<tr>
<td>Canova C. R.³⁵</td>
<td>1997</td>
<td>1</td>
<td>Internal carotid artery aneurysm. Radial artery aneurysm</td>
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<tr>
<td>Gürler A.³⁶</td>
<td>1997</td>
<td>3</td>
<td>—</td>
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<tr>
<td>Berkmam T.³⁵</td>
<td>1998</td>
<td>1</td>
<td>Pulmonary artery aneurysm</td>
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<td>Sasaki S.³⁷</td>
<td>1998</td>
<td>1</td>
<td>—</td>
</tr>
<tr>
<td>Our report (Akar H.)</td>
<td>1999</td>
<td>1</td>
<td>Deep vein thrombosis, popliteal vein thrombosis</td>
</tr>
</tbody>
</table>

2. The treatment of choice was surgery with or without immunosuppressive drugs.
3. The surgical procedures were PTFE graft interposition (most common), saphenous vein interposition and ligation.
4. Recurrence of popliteal artery aneurysm and amputation due to necrosis after surgery were rare.

This report shows that popliteal artery aneurysm may be more common than previously suspected in BD. BD should be considered in the differential diagnosis of popliteal artery aneurysms. The treatment of these patients includes surgery and immunosuppressive drugs.

However, controlled, long-term studies evaluating the effects of anticoagulation or immunosuppressive drugs on the treatment of vascular BD are needed. Clarification of the etiopathogenesis of BD can lead to better treatment options.

Acknowledgments

We want to express our appreciation to Ibrahim Seki, MD, Emergency Physician at Natchez Regional Medical Center, Natchez, Mississippi for his commitment to the study.

References

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