CASE REPORT

Internal jugular vein thrombosis in Behcet's disease: a rare complication

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SUMMARY

Behcet's disease (BD) is a systemic inflammatory disorder characterised by oral/genital ulcers, ocular lesions, and gastrointestinal, musculoskeletal, neurological and major vessel involvements. Venous manifestations are more common than arterial involvements. In this case report, we present a patient with internal jugular vein thrombosis, which is a very rare complication of BD.

BACKGROUND

Although internal jugular vein thrombosis is a very rare complication in patients with Behcet's disease (BD), its diagnosis should be established, and physicians should be aware of its possibility and significance.

CASE PRESENTATION

A 32-year-old man presented to our outpatient rheumatology clinic with a 2-week history of progressive dyspnoea, back pain and oedema on his face, neck and bilateral limbs. He was diagnosed with BD 5 years ago, and had a 1-year history of oral and genital ulcers and uveitis. He was treated with azathioprine (150 mg/day) and prednisolone (15 mg/day); the steroid dose was decreased gradually. On physical examination, he had a blood pressure of 100/70 mm/Hg, a temperature of 36.4°C, a heart rate of 84 bpm, bilateral oedema and tenderness on the limbs, neck and face.

INVESTIGATIONS

Laboratory data revealed a haemoglobin of 14.4 g/dL, a leucocyte count of 12 800/mm³, a thrombocyte count of 286 000/mm³, a blood urea nitrogen of 32 mg/dL, a serum creatinine of 0.8 mg/dL, an aspartate aminotransferase of 34 U/L, a C reactive protein (CRP) of 152 mg/L, an erythrocyte sedimentation rate (ESR) of 84 mm/h, an international normalisation ratio of 0.99, and an activated partial thromboplastin time of 28.5 s.

Some tests were ruled out for thrombosis. Lupus anticoagulans, antiphospholipid-anticardiolipin antibodies, antithrombin III, protein C and protein S levels were normal. Antinuclear antibody, immunofluorescent antibody and anti-dsDNA were negative. However, the patient had elevated levels of CRP and ESR. There was no history of surgical or medical interventions to his neck or limbs.

Left internal jugular vein thrombosis together with aneurysm and right internal jugular vein thrombosis extending down to the brachiocephalic vein have been revealed by Doppler ultrasonography. A Doppler study of the lower extremities

Figure 1 The thrombosis in the right internal jugular vein extending down to the brachiocephalic vein.

was normal. Computed tomography angiography (CTA) was performed for neck, chest and abdomen. The thrombosis in the right internal jugular vein extending down to the brachiocephalic vein (figures 1) and left internal jugular vein thrombosis together with aneurysm (figures 2, 3) were confirmed by CTA.

TREATMENT

We considered vascular inflammation associated with BD as an underlying cause for the thrombosis, started a 3-day pulse methylprednisolone (1000 mg/day) treatment and continued with prednisolone 60 mg/day. The patient was also administered cyclophosphamide (1000 mg/monthly). After recurrence of thrombosis, we started interferon therapy.

OUTCOME AND FOLLOW-UP

After a week, the patient's symptoms were alleviated. The oedema on his neck, face and limbs resolved. Laboratory data revealed an ESR of 10 mm/h and a CRP of 4.3 mg/dL. Two months later, his symptoms recurred and Doppler ultrasonography was performed again. The right internal

Figure 2 Left internal jugular vein thrombosis together with aneurysm.

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Figure 3 Left internal jugular vein thrombosis together with aneurysm.

jugular vein thrombosis and the left jugular vein thrombosis were still present. We started interferon treatment. His oedema and pain decreased again.

DISCUSSION

We aim to present a case with a rare complication of BD, internal jugular vein thrombosis. Vascular involvement is observed in approximately 40% of BD cases. Venous manifestations are more common than arterial involvements. Venous thromboses affect the lower extremities more commonly than the upper extremities and internal jugular vein thrombosis is a very rare manifestation of BD.^{1 2} In a study of 1200 patients with BD, 173 (14.4%) had venous manifestations and 19 (1.6%) had arterial involvement. Among the patients with venous manifestations, there were 154 (12.8%) with venous thrombosis, 17 (1.4%) with superior vena cava syndrome, 5 (0.4%) with inferior vena cava syndrome, 5 (0.4%) with varices, two with upper extremity venous thrombosis, and one with internal jugular vein thrombosis.³ However, our patient had no history of deep-vein thrombosis and the Doppler study of his lower extremities was normal. The clinical presentation of internal jugular vein thrombosis consists of leucocytosis, cervical pain, mass or neck swelling, superior vena cava syndrome, headache and back pain.^{4 5} Patients with BD may present solely with oedema on the face, neck and bilateral limbs without deep vein thrombosis at lower extremities. In our case, there was no catheterisation history that could have precipitated thrombosis. Venous thrombosis generally occurs approximately 2 or 3 years after the beginning of BD.⁶ Our patient presented with thrombosis 5 years after the diagnosis of BD. Therefore, we should be alert to the possibility of jugular vein thrombosis while following up a patient with BD even 5 years after the diagnosis. No evidence of any thrombophilic factors predisposing to thrombosis in BD has been established to date.^{7–9} The coagulation profile of our patient was also completely normal. Treatment of thrombosis in BD is not well established. Currently, treatment includes glucocorticoids, cyclophosphamide, cyclosporine, azathioprine and anticoagulants.¹⁰ Immunosuppressive agents have significantly reduced the incidence of venous thrombosis relapse in patients with BD. The safety profile of anticoagulation therapy was satisfactory, with haemorrhagic complications in 2% of these patients.¹¹ In a study from Korea, it was suggested

that immunosuppressive therapy is essential and anticoagulation therapy might not be required for the treatment of deep venous thrombosis associated with BD.¹² We conclude that patients should first be started on immunosuppressive therapy when thrombosis is detected. Lastly, we emphasise that internal jugular vein thrombosis may be the only vascular symptom on admission.

Learning points

- ▶ Behcet's disease is a significant disorder with possibly fatal complications such as internal jugular vein thrombosis.
- ▶ If patients with a concurrent diagnosis of Behcet's disease present with progressive dyspnoea, back pain and oedema on the face, neck and bilateral upper limbs, internal jugular vein thrombosis must be included in our differential diagnosis.
- ▶ We emphasise that internal jugular vein thrombosis may be the only symptom on admission.
- ▶ As anticoagulant treatments are neither sufficient nor effective in this case, immunosuppressive drugs should be prescribed first as treatment.

Competing interests None.

Patient consent Obtained.

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REFERENCES

- 1 Kural-Seyahi E, Fresko I, Seyahi N, *et al.* The long-term mortality and morbidity of Behcet syndrome: a 2-decade outcome survey of 387 patients followed at a dedicated center. *Medicine* 2003;82:60–76.
- 2 Melikoglu M, Ugurlu S, Tascilar K, *et al.* Large vessel involvement in Behcet's syndrome: a retrospective survey. *Ann Rheum Dis* 2008;67(Suppl 2):P67.
- 3 Kuzu MA, Ozaslan C, Koksoy C, *et al.* Vascular involvement in Behcet's disease: 8-year audit. *World J Surg* 1994;18:948–54.
- 4 Ascher E, Salles-Cunha S, Hingorani A. Morbidity and mortality associated with internal jugular vein thromboses. *Vasc Endovascular Surg* 2005;39:335–9.
- 5 Fuhrman T, Balatbat J, Frakes J. Internal jugular thrombosis causing increased intracranial pressure and upper airway edema. *Internet J Anesthesiol* 2000;4:3.
- 6 Bayraktar Y, Balkanci F, Bayraktar M, *et al.* Budd-Chiari syndrome: a common complication of Behcet's disease. *Am J Gastroenterol* 1997;92:858–62.
- 7 Fresko I, Melikoglu M, Tunc R, *et al.* Behcet's syndrome: pathogenesis, clinical manifestations and treatment. In: Ball GV, Louis Bridges S, eds. *Vasculitis*. Oxford; New York: Oxford University Press, 2002:406–32.
- 8 Sengul N, Demirel S, Yerdal MA, *et al.* Comparison of coagulation parameters for healthy subjects and Behcet disease patients with and without vascular involvement. *World J Surg* 2000;24:1584–8.
- 9 Mader R, Ziv M, Adawi M, *et al.* Thrombophilic factors and their relation to thromboembolic and other clinical manifestations in Behcet's disease. *J Rheumatol* 1999;26:2404–8.
- 10 Radke PW, Schwarz ER, Groesdonk H, *et al.* Thrombosis in Behcet's disease: report of a case followed by a systematic review using the methodology of evidence-based medicine. *J Thromb Thrombolysis* 2001;11:137–41.
- 11 Desbois AC, Wechsler B, Resche-Rigon M, *et al.* Immunosuppressants reduce venous thrombosis relapse in Behcet's disease. *Arthritis Rheum* 2012;64:2753–60.
- 12 Ahn JK, Lee YS, Jeon CH, *et al.* Treatment of venous thrombosis associated with Behcet's disease: immunosuppressive therapy alone versus immunosuppressive therapy plus anticoagulation. *Clin Rheumatol* 2008;27:201–5.

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