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(CASE REPORT)

Ortner's Syndrome Revealing an Aortic Arch Aneurysm in Behçet's Disease: A Case Report

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Abstract

Background: Behçet's disease is a systemic vasculitis that can affect vessels of any size. Arterial involvement is uncommon but may lead to severe complications. Ortner's syndrome, defined as hoarseness caused by recurrent laryngeal nerve compression from cardiovascular structures, is an exceptional mode of presentation.

Case Presentation: We report the case of a 34-year-old man with progressive hoarseness, fever, weight loss, arthralgias, and scrotal ulcers. Laboratory findings showed elevated inflammatory markers. CT angiography revealed a large saccular aneurysm of the aortic arch with mural thrombus compressing the left pulmonary artery. The diagnosis of Behçet's disease was retained. Because of active vascular inflammation, surgical repair was postponed. The patient was treated with high-dose corticosteroids followed by cyclophosphamide, leading to clinical and biological improvement.

Conclusion: This case illustrates the rare occurrence of Ortner's syndrome as the first manifestation of Behçet's disease. It underlines the importance of considering large-vessel vasculitis in young patients with unexplained hoarseness.

Keywords: Behçet's Disease; Ortner's Syndrome; Aortic Arch Aneurysm; Vasculitis; Case Report

1. Introduction

Behçet's disease (BD) is a chronic, relapsing vasculitis with a broad clinical spectrum. It is typically characterized by recurrent oral and genital ulcers, skin lesions and ocular inflammation, but can also involve joints, the gastrointestinal tract, the central nervous system and the vascular system. Venous thrombosis is the most frequent vascular manifestation. Arterial lesions are far less common, but when present they carry a high risk of rupture and mortality. The abdominal aorta is the most frequent site of aneurysm formation, while involvement of the thoracic aorta and its branches is exceptional.

Ortner's syndrome, or cardiovocal syndrome, corresponds to hoarseness caused by compression of the recurrent laryngeal nerve by cardiovascular structures. Initially described in patients with left atrial enlargement, it may also occur in association with aneurysms of the thoracic aorta. Its occurrence in the context of Behçet's disease is particularly rare.

We report here the case of a 34-year-old man in whom Ortner's syndrome was the revealing sign of a giant saccular aneurysm of the aortic arch. This unusual presentation illustrates the diagnostic challenge of large-vessel involvement

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in BD and emphasizes the need for early recognition and appropriate immunosuppressive therapy before surgical intervention.

2. Case Presentation

A 34-year-old man with no particular past medical history was referred for progressive hoarseness lasting four months. The symptom was associated with intermittent fever, weight loss, anorexia and diffuse arthralgias. On clinical examination, he had multiple scrotal aphthous ulcers; no ocular involvement was found. Cardiopulmonary auscultation was normal.

Laboratory investigations showed an inflammatory syndrome with elevated C-reactive protein. Thoracic CT angiography demonstrated a large saccular aneurysm of the aortic arch measuring 70 × 65 × 61 mm, with a mural thrombus compressing the left pulmonary artery (Figure 1).

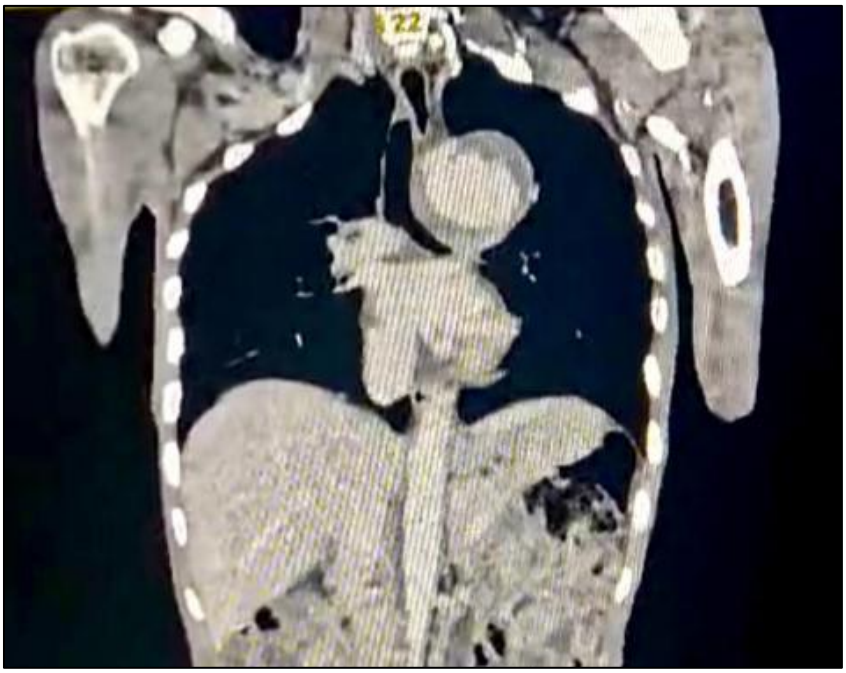

Figure 1 Contrast-enhanced CT angiography demonstrating a saccular aneurysm of the aortic arch with mural thrombus, compressing the left pulmonary artery and responsible for recurrent laryngeal nerve involvement leading to Ortner's syndrome

Transthoracic echocardiography was unremarkable.

Considering the association of recurrent oral and genital ulcers, systemic features and vascular involvement, the diagnosis of Behçet's disease was retained. Because of active vascular inflammation, surgical intervention was postponed. The patient was treated with high-dose intravenous corticosteroids followed by monthly cyclophosphamide pulses, which led to clinical and biological improvement.

3. Discussion

Arterial involvement in Behçet's disease is much less common than venous thrombosis, but when present it is associated with considerable morbidity and mortality [1]. Aneurysms are the most feared arterial lesions because of their risk of rupture. They usually involve the abdominal aorta, whereas thoracic and aortic arch aneurysms are distinctly uncommon [2,3]. Their clinical presentation can be misleading, often with non-specific or indirect symptoms, which may delay diagnosis.

Ortner's syndrome, or cardio vocal syndrome, was first described in relation to left atrial enlargement. It results from compression of the recurrent laryngeal nerve by cardiovascular structures. Its association with large-vessel vasculitis,

and especially with Behçet's disease, is exceptional [4]. In our case, progressive hoarseness was the only initial clue to a giant aortic arch aneurysm, underlining how easily the diagnosis can be missed.

Management of vascular Behçet's requires control of inflammation before considering surgery. Operative repair in the acute inflammatory phase carries a high risk of complications, including pseudoaneurysm formation. High-dose corticosteroids combined with cyclophosphamide remain the standard first-line induction treatment [1]. For refractory or severe cases, biologic agents, particularly anti-TNF therapies, have shown increasing efficacy [5–7]. Early immunosuppressive therapy is crucial, as it can stabilize the aneurysm, reduce the risk of rupture, and improve surgical outcomes [8,9].

This observation highlights an unusual but important message for internists: unexplained hoarseness in a young patient should not be underestimated. When associated with systemic features, it should prompt consideration of large-vessel involvement in Behçet's disease, where timely immunosuppressive therapy may be life-saving.

4. Conclusion

Ortner's syndrome is an exceptional mode of presentation of arterial involvement in Behçet's disease. This case illustrates how a seemingly isolated symptom such as persistent hoarseness can reveal a life-threatening vascular complication. Recognizing this unusual manifestation is essential, as prompt initiation of immunosuppressive therapy may stabilize the disease and allow safer surgical management.

Compliance with ethical standards

Acknowledgments

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Disclosure of conflict of interest

The authors declare no conflict of interest.

Statement of informed consent

Written informed consent for publication was obtained from the patient.

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