

# Retropharyngeal edema secondary to superior vena cava syndrome revealing Behcet's disease

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## Abstract

Retropharyngeal edema, uncommon in superior vena cava syndrome, can exceptionally represent the first manifestation of Behcet's disease, which should be taken into account in the differential diagnosis of this condition.

## KEYWORDS

Behcet's disease, retropharyngeal edema, superior vena cava syndrome

## 1 | INTRODUCTION

Behcet's disease is an inflammatory disease with vascular tropism; its clinical presentation is polymorphic, and its diagnosis is based on a set of criteria.<sup>1</sup> It is characterized by recurrent oral ulcers, genital ulcers, uveitis, suggestive skin lesions (pseudofolliculitis, erythema nodosum, etc), and vascular thrombosis.<sup>2</sup> Its diagnosis is based on minor and major criteria: oral and genital ulcerations, skin lesions, uveitis, arthritis, and neurological and digestive signs according to the International Study Group for Behçet's Disease.<sup>1</sup> Behcet's disease is more common in the Mediterranean region and the Middle East.<sup>2</sup> Superior vena cava syndrome is a group of clinical signs resulting from obstruction of blood flow through the superior vena cava. The most common symptoms are cervico-facial swelling, arm swelling, dyspnea, distended neck veins, and collateral venous circulation. Mediastinal malignancies and iatrogenic causes (central catheter or pace maker) are the most common causes of superior vena cava syndrome.<sup>3</sup> The superior vena cava syndrome is rarely observed in the context of Behcet's disease, and it is exceptionally revealed by retropharyngeal edema. In this context, we report a clinical case of a patient with retropharyngeal edema

revealing Behcet's disease. To the best of our knowledge, this could be the first case reporting a retropharyngeal edema secondary to superior vena cava syndrome revealing Behcet's disease.

## 2 | CASE REPORT

A 31-year-old Arab man, with no past medical history or family history of Behcet's disease, presented to our emergency department with a 4-day history of a diffuse cervico-facial edema associated with odynophagia without fever, dyspnea, or dysphonia.

The clinical examination revealed a diffuse cervical edema extended from the submandibular region to the supraclavicular and suprasternal regions. The edema was bilateral, symmetrical, soft, poorly limited, and noninflammatory. It was a nonpitting edema (Figure 1). There was no cervical lymphadenopathy. The chest examination showed a collateral venous circulation. The cardiopulmonary auscultation was normal.

Oropharyngeal and oral cavity examination showed multiple painful canker sores on the tongue (Figure 2) with a



**FIGURE 1** Cervico-facial edema, collateral venous circulation (✓)

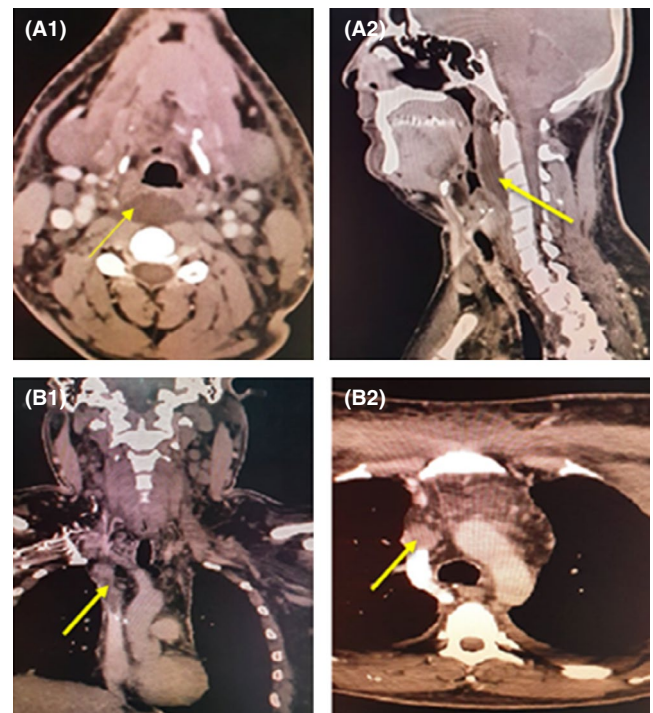


**FIGURE 2** Canker sores on the tongue (✓)

regular, symmetrical, and noncompressive bulging of the posterior wall of the oropharynx. There was no limitation of the mouth opening or tonsil bulging. The rest of the examination showed no signs of local infection.

The laboratory workup revealed a biological inflammatory syndrome (C-reactive protein 70 mg / l and sedimentation rate 40). Complete blood count was normal.

The cervico-thoracic computed tomography (CT) scan with contrast showed a retropharyngeal thickening extended to the parapharyngeal space with thrombophlebitis of the superior vena cava (Figure 3). Cervical magnetic resonance imaging (MRI) showed a retropharyngeal hyperintense lesion on diffusion weighted-sequence: the radiologic findings were consistent with a diagnosis of a retropharyngeal edema (Figure 4). Laboratory thrombophilia testing was normal.

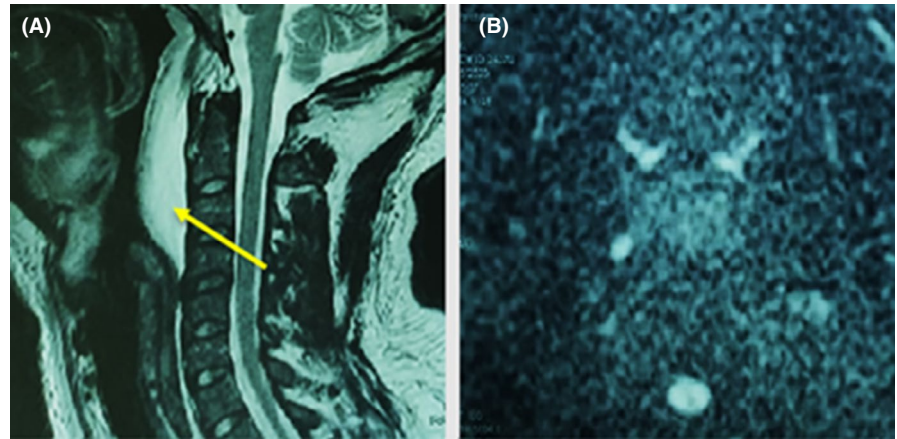


**FIGURE 3** CT scan with injection of contrast: A1, A2: retropharyngeal edema, B1, B2: thrombophlebitis of the superior vena cava and left internal jugular vein

Upon further questioning, we found that the patient had recurrent oral and genital aphthosis lasting for 6 months. Ophthalmologic examination was normal (didn't reveal uveitis). Skin examination was unremarkable. The diagnosis of Behçet's disease was evoked, then confirmed after grouping a set of clinical and biological criteria (oral and genital ulcers, thrombophlebitis, and positive pathergy test).

The patient was treated with prednisolone and colchicine, as well as anticoagulation (vitamin K antagonist). Clinical evolution was favorable after 3 months of follow-up.

**FIGURE 4** Cervical MRI. A: T2 weighted-sequence, retropharyngeal hypersignal, B: Diffusion weighted-sequence, retropharyngeal hypersignal



### 3 | DISCUSSION

The most observed pathologies in the retropharyngeal space are infectious diseases like abscesses and phlegmons. Sometimes, retropharyngeal edema can simulate a collection; it can be caused by radiation therapy, or retropharyngeal calcium tendonitis.<sup>4</sup> The differential diagnosis of retropharyngeal edema is mainly the infectious diseases (abscesses and phlegmons); retropharyngeal tumors are very rare. Abscess has a peripherally enhancing rim with a centrally low attenuation. Phlegmons have the same radiologic features as edema, but caused by infection in spaces surrounding the retropharyngeal space. MRI has superior contrast resolution than CT scan in studying the retropharyngeal space.

Although an obstructed superior vena cava causes increased venous pressure in the upper body, the collection of fluid in the retropharyngeal space is an unusual finding in superior vena cava syndrome. Fluid accumulation is thought to be due to impaired lymphatic drainage or excessive production of lymph.<sup>5</sup> To the best of our knowledge, this could be the first case in English and French literature reporting a retropharyngeal edema secondary to superior vena cava syndrome revealing Behcet's disease.

The superior vena cava syndrome is caused by the obstruction of the superior vena cava or its afferents by an intraluminal occlusion, a compression, or its invasion by a benign or malignant neof ormation, which results in a craniocervical venous return. The most frequent causes of superior vena cava syndrome are intra thoracic malignancies, iatrogenic causes (central catheter or pace maker),<sup>3</sup> and vasculitis as Behcet's disease. The differential diagnosis of superior vena cava syndrome includes right ventricular dysfunction, cardiac tamponade, and mediastinitis.

Behcet's disease is a vasculitis of unknown etiology; it is common in the Middle East and in young patients. Its diagnosis is based on minor and major criteria: oral and genital ulcerations, skin lesions, uveitis, arthritis, and neurological and digestive signs according to the International Study Group for Behçet's Disease.<sup>1</sup> Dr Hulusi Behçet in 1937 described

a diagnostic triad: recurrent oral and genital ulcerations and uveitis.

The underlying mechanism of thrombosis in patients with Behcet's disease remains unknown. The main mechanism responsible for thrombosis is thought to be endothelial damage induced by inflammation. Prothrombotic factors, abnormal fibrinolysis, altered platelet function, and genetic risk factors have also been suggested.<sup>6</sup> The venous system is more affected in Behcet's disease than the arterial system. Large venous trunks are rarely affected. The most affected veins are those of the lower extremities. Upper vena cava syndrome is rare; it is due to vascular disease; the absence of thrombosis is exceptional. Behcet's disease manifests as superior vena cava syndrome in only 2% of cases. The time taken to diagnose venous thrombosis in the case of Behcet's disease is 3.1 years.<sup>7</sup> The frequency of vascular lesions decreases after 5 years of progression of the disease.

### 4 | CONCLUSION

Superior vena cava obstruction is rarely the inaugural manifestation of Behcet's disease. The collection of fluid in the retropharyngeal space is an unusual finding in superior vena cava syndrome. Retropharyngeal edema in the context of Behcet's disease is an exceptional possibility which must be considered in the event of a superior vena cava syndrome in a young patient outside of a context of thrombophilia.

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### CONFLICT OF INTEREST

None declared.

## AUTHOR CONTRIBUTIONS

FM and IA: wrote the manuscript with support of AZ. WT, MAC, and BH. KBM: analyzed imagery. MM and IC: approved the final version of the manuscript.

## ETHICS APPROVAL AND CONSENT

Obtained from the patient in written.

## DATA AVAILABILITY STATEMENT

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

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