

Vocal cord palsy as a clinical manifestation of Behçet's disease[☆]



Parálisis de cuerda vocal como manifestación clínica de enfermedad de Behçet

To the Editor,

Behçet's disease (BD) is a rare autoimmune disease, with distinctive features like recurrent oral ulcers and, in most cases, genital ulcers. The symptoms can also include ophthalmological, neurological, dermatological, vascular, musculoskeletal, intestinal and pulmonary manifestations.¹ Involvement of the larynx is not common. We report the case of an individual with BD that affected a vocal cord.

The patient was a 58-year-old man who had stopped smoking 3 years earlier and had no other medical history of interest. He came to the department of internal medicine with recurrent oral and genital ulcers that had developed 2 months earlier. They were accompanied by fever and he felt generally unwell. Moreover, he also had a 2-week history of dysphonia, with no other associated symptoms.

Physical examination revealed dysphonia and the presence of painful scrotal and oral ulcers. In addition, he had an acneiform eruption on his back and lesions compatible with erythema nodosum on lower limbs. He presented no other problems.

Laboratory analyses disclosed leukocytosis, neutrophilia and an elevation of acute-phase reactants: 14,000 leukocytes/mm³ (78% neutrophils) and a C-reactive protein level of 150 mg/L.

Cranial computed tomography (CT) showed no evidence of ischemic or hemorrhagic cerebral vascular involvement. Cervicothoracic CT (Fig. 1) revealed paralysis of left vocal cord, whereas abdominal CT disclosed infrarenal abdominal aortic aneurysm measuring 47 mm × 48 mm, with no other findings.

The study of ear, nose and throat confirmed paralysis of left vocal cord. There was no ophthalmological involvement. Tests for autoimmunity included antinuclear antibodies, antineutrophil cytoplasmic antibodies, antiphospholipid antibodies, rheumatoid

factor and anti-cyclic citrullinated peptide antibodies, all negative. Serological tests ruled out bacterial infectious diseases like syphilis, Lyme disease and chlamydiosis and polymerase chain reaction for herpesvirus 1 and 2 and cytomegalovirus was negative. Blood cultures were also negative.

The patient underwent a pathergy test in right arm, which was positive, and a punch biopsy of the skin in the region of the acneiform eruption on his back showed evidence only of pseudofolliculitis. Given the clinical suspicion of BD, treatment was begun with oral glucocorticoids at a dose of 20 mg a day. The treatment with corticosteroids improved the ulcers and skin lesions, but there was no change in the dysphonia. The aneurysm observed in CT images subsequently required surgical treatment, which was successful.

Behçet's disease is an immune-mediated systemic vasculitis, characterized by the presence of recurrent orogenital ulcers, ocular inflammation and skin lesions. The etiology and pathogenesis of BD are unknown, although it has been demonstrated that the presence of human leukocyte antigen (HLA) B51 constitutes an important genetic predisposing factor, especially among patients from the Middle and Far East.¹

Being a multisystemic vasculitis, almost any part of the organism can be affected. The oral ulcers, recurrent and painful, are present in 90%–100% of the patients with BD. Other frequent clinical manifestations are genital ulcers (60%–80%), ocular lesions (67%–95%), skin lesions (41%–94%), arthritis (47%–69%) and neurological involvement (8%–31%).² The cutaneous manifestations of the patient we describe was nearly diagnostic of BD.

In contrast to central nervous system involvement, which is common in BD, peripheral nervous system manifestations are rare. There have been isolated cases of Guillain-Barré syndrome, mononeuritis multiplex, sensorimotor neuropathy, autonomic neuropathies and subclinical nerve conduction abnormalities.³ However, vocal cord involvement is unusual and, to date, only 2 cases have been reported in the medical literature.^{4,5}

The most probable explanation for this occurrence could be the presence of vasculitis at the level of the recurrent laryngeal nerve.⁵

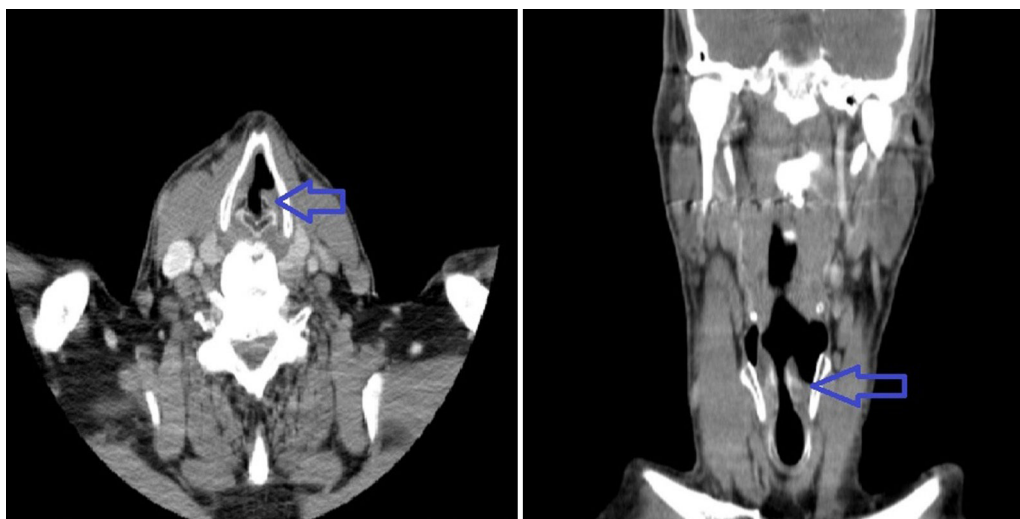


Fig. 1. On the left side, a cross-sectional slice of a cervical CT showing the paramedial position of the left vocal cord (arrow). On the right side, an axial slice showing the same position of the vocal cord (arrow), findings compatible with paralysis.

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In the case of this patient we found no other condition to explain the dysphonia.

In conclusion, BD should be taken into account in the differential diagnosis of sudden dysphonia in the absence of other more common findings, especially in regions in which the disease is frequent.

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Osteosarcoma in Tophaceous Gout: A Case Report and Literature Review[☆]



Osteosarcoma en gota tofácea: descripción de un caso clínico y revisión de la literatura

To the Editor,

Gout is a disease that involves the deposit of monosodium urate (MSU) crystals, which mostly affects men, and constitutes the most common cause of arthritis in that gender. On occasion, it can lead to accumulations of crystals of variable size and location referred to as tophi. Considered to be a “mimicking” disease, gout can simulate, among others, infectious or neoplastic processes. We report the case of a patient with gout who developed an osteosarcoma arising at the site of a preexisting tophus.

The patient was a 65-year-old man with tophaceous gout who had been followed at another center and was referred to us by way of the medical oncology department for the management an acute attack of polyarthralgia and olecranon bursitis after a transfusion while undergoing chemotherapy. He had been diagnosed a year earlier with osteosarcoma on the dorsum of his foot arising at the location of a tophus that had been removed 7 months before, after pathological confirmation (Fig. 1) and negative findings in the study of the extension. It required infracondylar amputation and preoperative and postoperative chemotherapy with cisplatin and adriamycin. He had been taking allopurinol for 3 years (up to 300 mg/day), and employed colchicine and/or nonsteroidal anti-inflammatory drugs during attacks. He had levels of uric acid in blood of up to 8 mg/dL, and began to have symptoms at the age of 40 years with podagra, and later developed tophi in the olecranon bursa and the dorsum of his foot. The inflammatory attack was resolved with low-dose prednisone and aspiration of the olecranon bursa. Gram staining and culture of the fluid obtained were negative, and a study with polarized light microscopy revealed the presence of MSU crystals. He was discharged and began colchicine at 1 mg/day after gradual interruption of prednisone therapy. During follow-up, he changed from allopurinol to febuxostat (at a dose

of 80 mg/day), which achieved optimization of his uric acid level in blood and progressive dissolution of the olecranon tophi. Several months later, colchicine was discontinued and he continued to take febuxostat at the same dose. He has not had any further attacks and maintains his level of uric acid at 5 mg/dL. In May of 2016, he developed an “in-transit” recurrence of the neoplastic disease, with a superficial mass in ipsilateral thigh that required local resection and chemotherapy (ifosfamide).

Gout is the most common cause of arthritis in men, and is originated by deposits of MSU crystals in tissues. Some patients develop tophi (subcutaneous, periarticular or intraarticular crystal accumulations) which can have a marked inflammatory activity and present calcifications or ossifications as they increase in size.^{1–3} Moreover, tophi can mimic infectious or neoplastic processes and become superinfected or undergo malignant transformation, having an adverse effect on the prognosis of the patient.

A number of publications have reported tumors that arise at the sites of tophi.^{4–9} All occurred in extremities of middle-aged or elderly men and involved recurrences or metastasis. There are descriptions of benign neoplasms such as giant cell tumors or malignant diseases such as different sarcomas (undifferentiated pleomorphic sarcoma, cutaneous angiosarcoma or fibrosarcoma).

The authors of these articles provide interesting hypotheses such as the role of prolonged hyperuricemia and the severity of the MSU deposit as factors leading to secondary cell transformation toward inflammation,⁸ or the theory of sarcoma associated with a foreign body, similar to those originated by other materials.^{7,10}

Sarcomas are mesenchymal tumors originating from connective tissue (fibrosarcoma), cartilage (chondrosarcoma), bone (osteosarcoma), endothelium (angiosarcoma), etc. Although there may be a certain underestimation of the incidence of these neoplasms in gout, sarcomas seem to be uncommon, but occasionally are aggressive. Thus, they should be included in the differential diagnosis in the case of a subcutaneous or periarticular mass in a patient with gout or with the atypical complication of a preexisting tophus.

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