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Case Report

Vasculo-Behçet's Disease Mimicking a Metastatic Neck Mass

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—— This article presents a case of Behçet's Disease (BD) with vascular involvement of the neck, which mimicks a metastatic neck mass in the initial presentation. A 58-year-old man presented with dysphagia, weight loss, bulging on the lateral wall of the left pyriform sinus, and a firm and fixated neck mass suggestive of metastasis. Computed tomography of the neck demonstrated a solid mass, around the bifurcation of the carotid artery together with a pseudoaneurysm of the left external carotid artery. The mass was about four centimeters in diameter and extended to hypopharynx medially. Biopsy from neck mass and hypopharynx revealed no specific pathology. During follow-up the firm and fixated mass changed into a completely pulsatile one in the following three weeks. Reassessment of the patient's past history in detail revealed that he had had recurrent oro-genital ulcers, arthralgia and recurrent skin lesions. The pathergy test was positive. The patient was diagnosed to be BD and treatment consisting of colchicine 1 mg/day, peroral was started. He had a favorable outcome after treatment and was asymptomatic at follow-up of 24 months. It is unusual for BD to present as a neck mass but yet it must be considered in the differential diagnosis of neck masses. The present case report demonstrates how such a mass may mimic metastatic tumoral involvement and cause diagnostic dilemma. ———— Behçet's disease; vasculitis; neck mass
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Behçet's disease (BD) is a multisystemic disease, characterized by genital ulcers healed with scarring, recurrent oral aphthous ulcers, arthritis, skin lesions and ocular manifestations (e.g., uveitis). BD frequently occurs in Mediterranean countries and Japan, but it has been described worldwide. It has an undulating course with exacerbation and remission periods. The underlying disease presents as small and large vessel vasculitis in various organs (Tuzun et al. 1997). Vascular complications of BD may include venous thrombosis, arterial occlusion and arterial aneurysm formation (Kuzu et al. 1994).

This article presents a case of BD with vas-

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cular involvement of the neck, which mimicked a metastatic neck mass in the initial presentation.

Case Report

A 58-year-old male was referred for evaluation of a mass in the neck with the suspicion of a metastatic lymphadenopathy from an unknown primary origin. His complaints were sore throat, dysphagia, weight loss (10 kg in two months) and a swelling on the left side of his neck, which he had noticed approximately two months ago. He is an ex-smoker for two years with a history of cigarette smoking, 1 package per day for 33 years. During the physical examination, a quite firm, fixated mass with poorly defined margins was palpated on the left carotid bifurcation region. Examination of the larynx revealed a bulging on the lateral wall of the left pyriform sinus. Appearance of the mucosa was normal. An arteriogram was performed with suspicion of carotid body tumor. The angiogram showed a pseudoaneurysm of the left external carotid artery (Fig. 1) and another of the left femoral artery (Fig. 2).

Computed tomography of the neck demonstrated a contrast enhancing solid mass, around the bifurcation of the carotid artery together with a pseudoaneurysm of the left external carotid artery. The mass was about 4 cm in diameter with poorly defined margins and extended to hypopharynx medially and the skull base superiorly (Fig. 3). We suspected of a metastatic lymphadenopathy leading to carotid aneurysm formation with these clinical and radiological findings. Direct laryngoscopy and biopsy from hypopharynx was performed under general anesthesia. During the operation, an aspiration biopsy from the neck mass was obtained with 25-gauge needle. In order to avoid rupturing, biopsy was taken from the posterior part of the mass, away from the aneurysm. In early postoperative period, the patient woke up with right hemiplegia. Neurological consultation and management were immediately provided and the patient was followed-up for hemiplegia, which gradually improved with almost complete recovery in several days. Cytopathologic examination of the specimens revealed non-specific inflammation. During follow-up the firm and fixated mass changed into a completely pulsatile one with relatively sharp boundaries in the following three weeks. Scrotal



Fig. 1. Digital substraction angiography of the left common carotid artery. Pseudoaneurysm arising from the left external carotid artery (∗) can be seen.

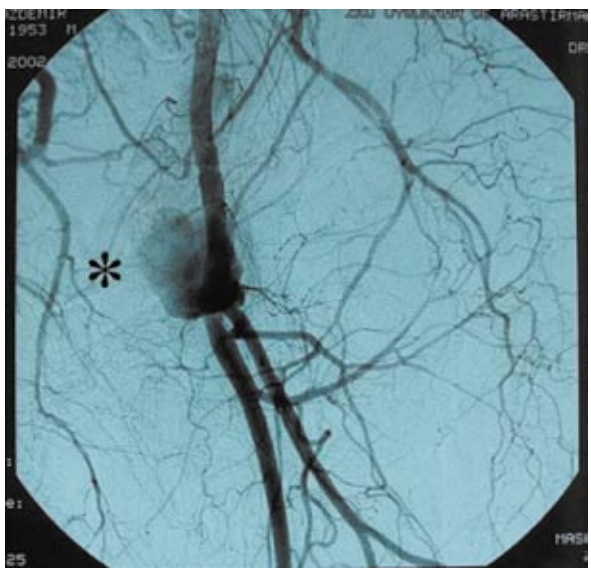


Fig. 2. Digital substraction angiography of the left common femoral artery. Pseudoaneurysm originating from the distal bifurcation of left common femoral artery (∗) can be seen.

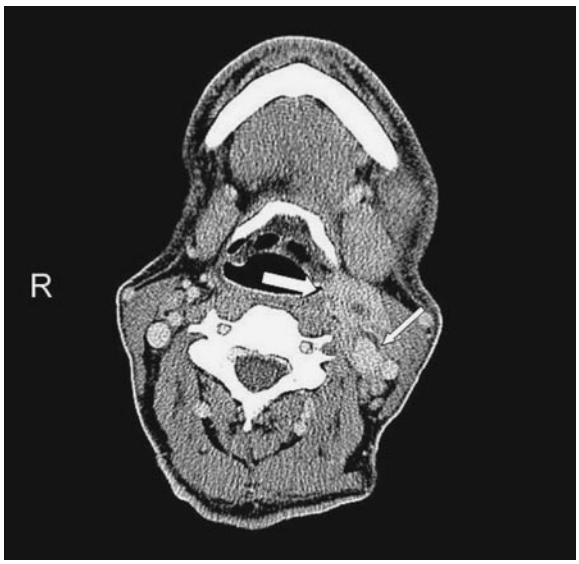


Fig. 3. Contrast enhanced computed tomography imaging of the neck. Pseudoaneurysm of the left external carotid artery (*thin arrow*) can be visualized in this axial slice computed tomography image. Contrast enhanced solid mass around the left carotid bifurcation extending to hypopharynx medially (*thick arrow*) can be seen.

scarred ulcers, pustular skin lesions and a left femoral pulsatile mass were observed in physical examination. Thereupon, the past history was explored in detailed fashion with suspicion of a systemic disease. The patient noted that, he had recurrent oro-genital ulcers, arthralgia, and frequently reappearing skin lesions. The pathergy test was positive. Examination of the eye did not show any pathologic findings. The patient was diagnosed to be BD and treatment consisting of colchicine 1 mg/day, peroral was started. We observed improvement of the pustular lesions of the skin and weight gain in the next two months period. He was asymptomatic at follow-up of 24 months. The patient refused any further surgical therapeutic modality involving vascular reconstruction.

Discussion

BD, in general, occurs more commonly in men and predominantly affects people during their second or third decades (Kuzu et al. 1994). The common histopathological features of BD present in the base of nonspecific vasculitis of small and large vessels. Cardiovascular involvement appears in only 7-29% of patients. Venous thrombosis, aneurysm formation, and arterial occlusion are well-documented features of BD. Among these vascular lesions, thrombophlebitis is the most common one. Aneurysm is more frequent than occlusion with arterial involvement in BD. The most common localization site of aneurysm is the abdominal aorta, followed by the pulmonary and femoral arteries (Tuzun et al. 1997).

So far, 16 cases with involvement of carotid arteries due to BD have been reported in the world literature. Among these patients, aneurysms of carotid artery are associated with other large artery aneurysms such as, abdominal aorta, right cerebral artery, left cerebral artery, middle cerebral artery, anterior communicating artery, popliteal artery, and radial artery. Our case is the second report in which carotid artery aneurysm is associated with femoral artery aneurysm. Previously, Canova et al. (1997) reported internal carotid artery aneurysm associated with femoral artery and radial artery aneurysms.

In the affected artery active arteritis initially occurs, inflammatory cells composed of neutrophils, lymphocytes and plasma cells infiltrate the media and adventitia, mainly around the proliferated vasa vasorum. With the destruction of elastic and muscle cells in the media the vessel wall weakens and a process of aneurysmal dilatation starts. Occlusion of the vasa vasorum speeds this process by transmural necrosis. Ultimately, perforation of the vessel wall and pseudoaneurysm formation ensues (Tuzun et al. 1997).

Aneurysm formation may also be associated with tumoral involvement of the carotid artery. There have been several reports of aneurysm formation due to metastasis of organ cancer. Two examples are, carotid artery aneurysm due to metastatic choriocarcinoma, and tonsillary and epipharyngeal carcinoma with metastasis which have been reported by Toyama et al. (1986) and Karov (1996), respectively.

Our patient presented with a non-pulsatile, quite firm neck mass, fixated to the surrounding tissues with poorly defined margins. These find-

ings were associated with obliteration of the pyriform sinus, weight loss and dysphagia. Computed tomography of the neck showed a solid mass around the carotid bifurcation region. In such circumstances, it is mandatory to rule out malignancy. For that reason, fine-needle aspiration biopsy and hypopharyngeal punch biopsy were performed to confirm the possible diagnosis. To the best of our knowledge, this is the first case report in the literature in which BD mimics hypopharyngeal carcinoma with typical features like weight loss, dysphagia and obliteration of the pyriform sinus together with an apparently metastatic neck mass.

In BD with vascular presentation, the clinical presentation of the aneurysm depends on the stage of the disease. Tuzun et al. (1997) reported iliac and extremity aneurysms diagnosed in the active stage of the disease, which were characterized by pulsatile and tender mass. In our case, a non-pulsatile, quite firm mass, fixated to the surrounding tissues with poorly defined margins has been observed in the initial period of the disease. In this stage, it reminded of a metastatic neck mass. However, during follow-up, the lesion changed to a pulsatile mass with relatively sharp boundaries, in contrast with the pattern described in previous reports. We claim that the firm nature of the mass in the initial period was due to severe active inflammation related to BD. As the active phase of the inflammation has subsided, the enduration encircling the mass has ceased and the pulsatile mass due to the aneurysm has become evident.

The success rate of surgery is rather poor, since aneurysm formation at the site of anastomosis is likely as a result of vasculitis in BD (Canova et al. 1997; Tuzun et al. 1997). We did not consider surgical repair due to the low success and high complication rates and the recent history of cerebrovascular accident in our patient. Endovascular insertion of a stent graft may be a reasonable alternative to avoid complications of surgical repair. We planned to perform endovascular intervention following the improvement of the general status of our patient but he refused to undergo further treatment for vascular reconstruction.

Conclusion

It is unusual for BD to present as a neck mass but yet it must be considered in the differential diagnosis of neck masses. Present case report demonstrates how such a mass may mimic metastatic tumoral involvement and cause diagnostic dilemma.

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