

Takayasu's Arteritis as a Cause of Carotidynia: Clinical and Imaging Features

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Carotidynia is a neck pain syndrome associated with tenderness on palpation over the carotid bifurcation. The differential diagnosis for carotidynia includes large-vessel vasculitides such as Takayasu's and temporal arteritis, arteriosclerosis, thrombosis, dissection and aneurysm. Carotidynia, particularly in a young patient or a patient without risk factors for atherosclerosis, should alert the clinician to the possibility of arterial pain and subsequent large-vessel vasculitides. We report the clinical and imaging findings in a young patient with Takayasu's arteritis as a cause of carotidynia.

Patient Description

A 41 year old woman with no significant past medical history presented with recurrent anterior neck pain that had troubled her for a year. She complained of almost daily bouts of vague, dull, continuous ache, while denying fatigue, malaise, dizziness, weight loss and fever. Her pain was not related to swallowing, coughing or head movement. Treatment with antibiotics and non-steroidal anti-inflammatory drugs was not helpful.

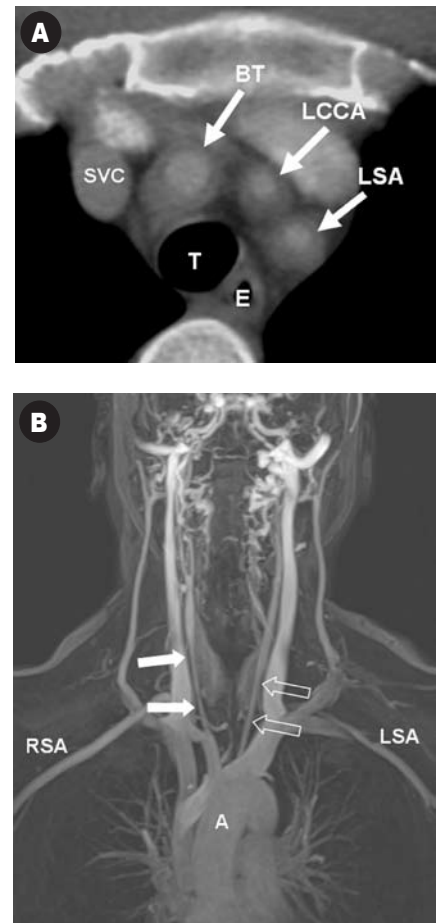
Physical examinations revealed mild tenderness over the carotid bifurcations, normal peripheral pulses with no bruits above the large neck vessels, limbs or aorta. Her examination by the otorhinolaryngologist was unremarkable. Blood pressure was 124/76 mmHg in the right and 136/80 mmHg in the left arm. Laboratory testing revealed normocytic anemia (hemoglobin 9.8 g/dl), thrombocytosis (platelet count 560,000/ μ l), elevated erythrocyte sedimentation rate (85 mm/hour) and C-reactive protein serum level of 60 mg/dl (normally < 10 mg/dl).

Soft tissue ultrasonography of the neck was reported as normal. Contrast-enhanced computed tomography scan showed concentric arterial wall thickening involving the thoracic and abdominal aorta, brachiocephalic trunk, common carotid arteries, and left subclavian artery [Figure A]. Both common carotid arteries and the left subclavian arteries showed various degree of vessel narrowing without arterial wall calcification. Magnetic resonance angiography revealed significant tubular stenosis of both common carotid arteries and of the proximal segment of the left subclavian artery [Figure B]. This presentation was consistent with the diagnosis of Takayasu's arteritis. Treatment with glucocorticosteroids was initiated and led to rapid and complete relief of the patient's neck pain and normalization of her laboratory inflammation parameters.

Comment

Takayasu's arteritis is a large-vessel vasculitis in young women that affects the aorta, its main branch vessels and the pulmonary arteries [1]. Although Takayasu's arteritis occurs mainly in Asia, the distribution of the disease is worldwide. The etiology is unknown but it is believed to be autoimmune. Histologically, it is characterized by a granulomatous infiltrative process of the arterial wall (acute stage) with marked intimal proliferation and fibrosis of the media and adventitia (fibrotic stage), resulting in severe stenosis, occlusion or aneurysmal dilatation (from extensive destruction of elastic fibers in the media).

The clinical manifestations of the disease are divided into early and late phases. The early or pre-pulseless phase



Takayasu's arteritis in a 41 year old woman. [A] Axial contrast-enhanced CT image (obtained at the level of the aortic arch great vessels origin) shows concentric arterial wall thickening involving the brachiocephalic trunk (BT), left common carotid artery (LCCA) and left subclavian artery (LSA). E = esophagus, SVC = superior vena cava, T = trachea. [B] Coronal maximum intensity projection (MIP) 3D magnetic resonance angiogram shows significant tubular stenosis of the right common carotid artery (arrows), the left common carotid artery (open arrows) and the proximal segment of the left subclavian artery (LSA). A = ascending aorta, RSA = right subclavian artery.

is often characterized by non-specific systemic signs and general symptoms of systemic inflammation (such as fever, night sweats, weight loss, weakness, myalgia and arthralgia), whereas the late or pulseless phase (consequences of vascular inflammation or fibrosis) may be associated with symptoms of ischemia of limb (claudication, pulse deficit, bruits), renovascular hypertension, mesenteric angina, retinopathy, aortic regurgitation (when the ascending aorta is involved), myocardial ischemia and neurological symptoms.

Carotidynia is a neck pain syndrome associated with tenderness on palpation over the carotid bifurcation. The pain is of mild to moderate intensity, typically dull, throbbing and continuous. The pain may be aggravated by swallowing, coughing, sneezing, or elevating the head while moving it toward the contralateral side [2]. The differential diagnosis for carotidynia includes large-vessel vasculitides such as Takayasu's and temporal arteritis, arteriosclerosis, thrombosis, fibromuscular dysplasia, dissection, aneurysm, pharyngitis, lymphadenitis, submandibular gland disease, thyroiditis, and neck neoplasm. Carotidynia has been reported in 32% of patients with Takayasu's arteritis [3]. The proposed mechanism of carotidynia in large-vessel vasculitides includes the involvement of the adventitia with the inflammatory process through the vasa vasorum, followed by the media. Of relevance, nerve distribution in most blood vessels is confined to the adventitial-medial border, potentially making the process involving the adventitia and media painful, as opposed to intimal injury which is relatively painless.

The delay in diagnosis of Takayasu's

arteritis due to lack of specificity of early symptoms is not rare and may have serious consequences for the patient in the late fibrotic stage. Therefore, carotidynia alone or with raised inflammatory markers (erythrocyte sedimentation rate and C-reactive protein), claudication of extremities, hypertension, arterial bruit, particularly in a young patient or a patient without risk factors for arteriosclerosis, should alert the clinician to the possibility of arterial pain and subsequent large-vessel vasculitides such as Takayasu's arteritis.

Conventional angiography has been the gold standard imaging tool for the diagnosis and evaluation of Takayasu's arteritis, but it is clearly invasive with associated procedural morbidity and mortality risks. CT angiography, MRI and magnetic resonance angiography are non-invasive good alternative tools for the diagnosis of Takayasu's arteritis with superb resolution of changes in the vessel walls: thickening, luminal narrowing and dilatation [1,4]. MRI may also show enhancement of the thickened walls with gadolinium contrast material, suggestive of the active phase. Positron emission tomography scan with 18F-fluorodeoxyglucose was recently shown to be effective in diagnosing Takayasu's arteritis, differentiating active and late/fibrotic stages of the disease, as well as in dynamic and serial evaluation of patients with Takayasu's arteritis [1].

Corticosteroids remain the mainstay of medical treatment; however, Takayasu's arteritis is often resistant to treatment, so pulse therapy with methylprednisolone should be considered. Several immunosuppressive agents – such as methotrexate, cyclophosphamide, azathioprine, cyclosporine, and mycophenolate mofetil

– have been used as steroid-sparing agents in Takayasu's arteritis patients with varying efficacy. The safety and efficacy of anti-tumor necrosis factor therapy for Takayasu's arteritis were recently reported in an open-label study [1,3]. Obstructive lesions need to be managed by revascularization techniques such as angioplasty and surgery.

In conclusion, carotidynia is a painful symptom that may be an underrated clinical sign of Takayasu arteritis and other large-vessel vasculitides. Non-invasive imaging modalities such as CT and MRI allow earlier diagnosis than conventional imaging. MRI enhanced with gadolinium and PET scan offer additional information regarding the staging, dynamic evaluation and management of Takayasu's arteritis.

References

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PET = positron emission tomography