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Case Report

Radiographic findings on inflammatory myofibroblastic tumor of the carotid artery: report of two cases

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Abstract: MRI and/or CT scan images were reviewed in two cases of inflammatory myofibroblastic tumor (IMT) of the carotid artery. The MRI study showed that IMT of carotid artery presented itself as an avascular fusiform mass around the carotid bifurcation, with or without focal narrowing of carotid artery. The lesion was isointense or slightly hyperintense on T1 or T2-weighted MRI, strongly enhanced on Gadolinium-enhanced T1-weighted MRI. This was not enhanced by contrast on CT imaging. These results indicate that the radiographic characteristics of IMT of the carotid artery shall be taken into account during diagnosis.

Keywords: Inflammatory myofibroblastic tumor, carotid artery, magnetic resonance imaging, computed tomography, diagnosis

Introduction

Inflammatory myofibroblastic tumor (IMT) of carotid artery is an extremely rare disease which involves the vascular compartment and surrounding soft tissue of the neck [1]. According to the documented papers, the lesion has the propensity to involve the carotid bifurcation though it's not the most common site in the head and neck [2-5]. Here we report some radiographic findings on IMT of carotid artery in two patients, which are based on the magnetic resonance imaging (MRI), computed tomography (CT) and magnetic resonance angiography (MRA).

Case report

Case 1

A 56-year-old male had a 3-year-history of recurrent syncope. The symptom had been becoming worse and worse for one month by the time the patient was admitted to West China Hospital, for he experienced repeated syncope and falls at shortened intervals. A mass was discovered under the sternocleidomastoid muscle in the upper left neck.

Color duplex imaging demonstrated a lesion with a weak echo of uneven thickness which wrapped around the carotid bifurcation. Besides, blood flowed more slowly in the right vertebral artery than in the left one. MRI T1 or T2-weighted imaging indicated a 3×4 cm mass isointense to skeletal muscle in cross section, and these imaging also showed a strongly enhanced pericarotid fusiform mass, with circular hypointense signals around the terminal segment of the common carotid artery and the proximal ends of both the internal and external carotid arteries in coronal section, which was also observed in Gadolinium-enhanced T1-weighted imaging (**Figure 1A-D**). A suspected narrowing of carotid artery was demonstrated on MRA imaging (**Figure 1E**). Preoperative examinations revealed his cardiological and neurological conditions, total serum IgG and IgG4 levels were within normal limits. Surgical exploration was performed. During surgery, the lesion was found to be scar-like with rich blood supply. The vagus nerve and sympathetic chain were also trapped in the lesion (**Figure 1F**). Histologically, the lesion revealed spindle myofibroblast and inflammatory cells in the myxoid or collagen background. Immunohistochemically, the myofibroblastics pindle cells were positive

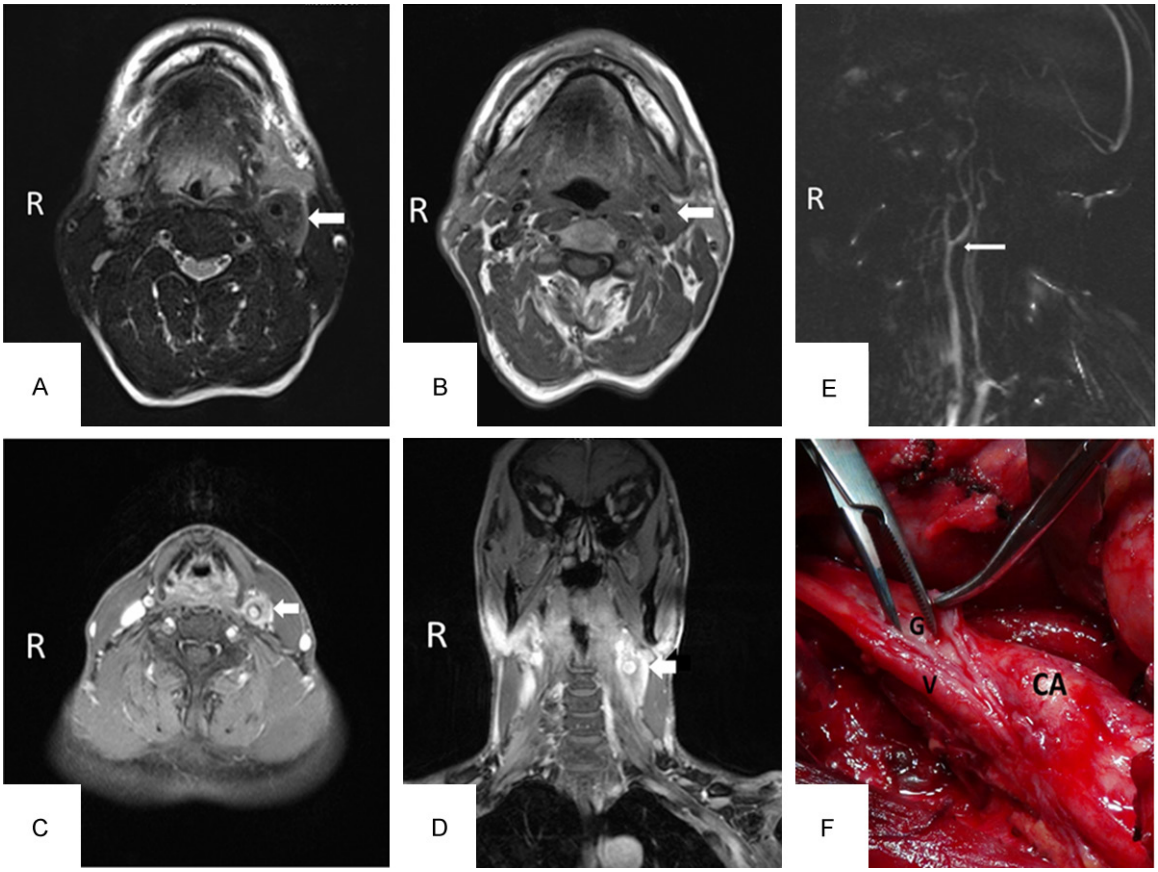

Figure 1. MRI and operative field of case 1 MRI demonstrated a 3×4 cm mass with isointense signals to skeletal muscle in T1 (arrow) (A) and T2-weighted imaging (arrow) (B) at the left side; A strongly enhanced pericarotid round mass with circular hypointense signals around the left carotid artery in Gadolinium-enhanced T1-weighted imaging (arrow) (C: cross section, and D: coronal section); Obscure narrowing of the proximal end of internal carotid artery (arrow) (E); Vagus nerve (V) and sympathetic chain were trapped in the scar-like lesion (F) (CA: carotid artery, G: granulomatous tissue).

for smooth muscle actin (SMA), partially positive for human mitotic spindle apparatus (MSA), and weakly positive for anaplastic lymphoma kinase (ALK). An IMT diagnosis was thus confirmed. The patient was treated with short-term corticosteroids and traditional Chinese herbs. Then during a 2-year follow-up, neither the symptoms nor obvious imaging recurrence was observed.

Case 2

A 69 years old male was admitted to West China Hospital due to a 3-month history of pharyngalgia and dizziness. The latest MRI revealed that lunar isointense signals to the skeletal muscleon T1-weighted imaging and slight hyperintense signals on T2-weighted imaging located posteriorly to the left carotid artery. Meanwhile CT of the neck demonstrated some lymph nodes without significant enlargement (**Figure 2A**, **2B**). Total serum IgG and IgG4 levels were also within normal limits. Apart from aforementioned symptoms, the patient also suffered from severe hypertension, bilateral nasal sinusitis, bilateral mastoiditis, anemia, and colitis gravis. General systemic support, corticosteroids, and antibiotics were intermittently given for 3 years and provided some symptom relief.

One month later after his admission, follow-up CT revealed oval bulging around the left carotid artery, which was not enhanced by contrast (**Figure 2C**, **2D**). Furthermore, MRI demonstrateda lesion with isointense signals on T1 or T2-weighted imaging, and strongly-enhanced circular hypointense signals around the carotid artery on Gadolinium-enhanced T1-weighted imaging. The adventitia of the carotid artery

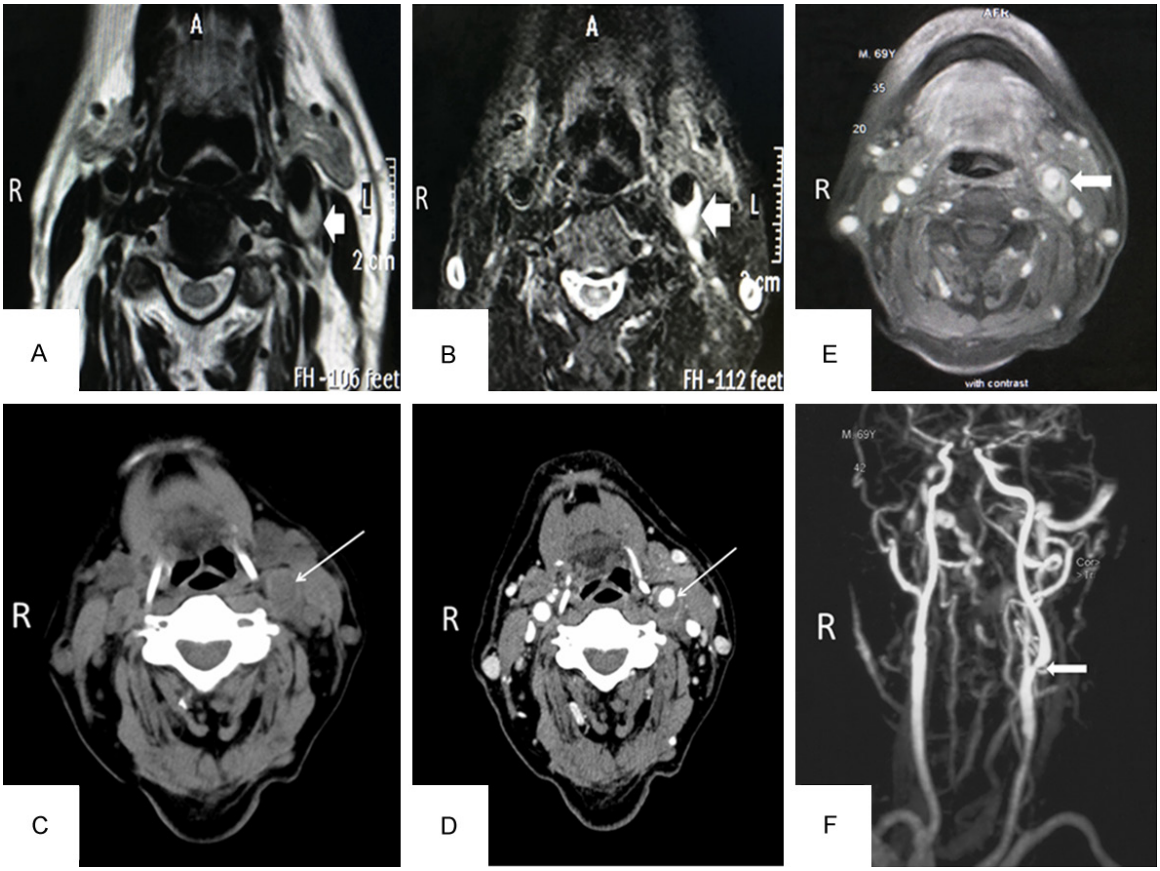

Figure 2. MRI and CT imaging of case 2 MRI revealed a lunar isointense signals posterior to the carotid artery in T1-weighted imaging (arrow) (A); Hyperintense signals in T2-weighted imaging (arrow) (B); Oval bulging around the left carotid artery is not enhanced in contrast-enhanced CT (arrow) (D) in comparison with that in plain CT (C); A strongly enhanced pericarotid lesion with circular hypointense signals around the left carotid artery in Gadolinium-enhanced T1-weighted imaging (arrow) (E); Focal carotid narrowing at the proximal end of both internal carotid arteries in MRA (arrow) (F).

was interrupted (**Figure 2E**). The MRA imaging demonstrated focal carotid narrowing at the proximal end of internal carotid arteries of both sides and showed the slightly rough lumen of the external carotid arteries (**Figure 2F**). An incisional biopsy was performed and the diagnosis of IMT was finally established. Here corticosteroid was again administered for this patient, after which pharyngalgia recurred sometimes but the dizziness gradually disappeared in the subsequent 2 months. The lesion also markedly decreased in size after six months.

Discussion

IMT is also called inflammatory pseudotumor, a round or spindle cell proliferative lesion, which occurs mostly in the lung. Besides, it also occurs in different extra-pulmonary locations such as the abdomen, retroperitoneum, pelvis, heart, head and neck, upper respiratory tract, trunk, bladder and extremities [6, 7]. After the first report by Rice in which this disease was described as sclerosing inflammatory pseudotumor arising from the carotid artery, several papers reported that it was prone to occurring at the carotid bifurcation [1, 3-5, 8]. Only two cases have been reported in the Chinese population up to now [9, 10].

The etiology of IMT in carotid artery is still uncertain. It is known that the ingestion of methysergide or ergot derivatives is linked to cervical sclerosis but it is not traced through careful review of medical history in these two patients [11]. Clinical symptoms are generally caused by the entrapment of cranial nerves such as hypopharygeal and vagus nerves, or related to the hypersensitive carotid sinus [3, 10]. A carcinoma or metastasis at the carotid

bifurcation can involve the carotid sinus and cause neurally-mediated syncope whose clinical presentation is similar to carotid sinus syndrome. A carotid body tumor with such consequences is extremely rare in literature [11-14]. The symptoms in these two patients are different but still can be easily explained by vascular and neural reasons.

IMT of the carotid artery has also been called sclerosing cervicitis [15]. When the term, cervical fibrosis or sclerosing cervicitis, is used, it refers to one form of tumefactive fibroinflammatory lesions of the head and neck, namely an entity that frequently affects the sinonasal tract. Sometimes it also involves other sites such as the mandible, tongue, face, cheek, and infratemporal fossa. It is argued that the disease can represent a localized manifestation of a systemic condition because about 20% of the patients also have other inflammatory fibrosclerosing lesions, such as sclerosing cholangitis, mediastinal fibrosis, retroperitoneal fibrosis, and orbital pseudotumor. Recently, most cases of the inflammatory fibrosclerosing lesions have been found to belong to the family of IgG4-related sclerosing diseases. However, IgG and IgG4 serum levels of these two patients could not confirm this relationship. It is not supported by our previous report about sclerosing sialadenitis of the submandibulargland either [16]. This implies that IgG and IgG4 serum levels don't serve as good indicators for IMT of the carotid artery.

Narrowing of the carotid artery has been reported in some cases due to involvements of the carotid artery wall. Theoretically, when intraluminal growth happens, all layers of the carotid artery are involved. Practically, the IMT does involve the carotid bifurcation and makes the dissection difficult in the first case and narrows the carotid lumen in the second case [3, 10, 11]. It is reported that corticosteroids successfully reduce the IMT lesion and almost make a perfect solution to the arterial stricture [17, 18]. Because we were not able to retrieve any significant signs and symptoms relevant to the suspected carotid artery narrowing on the right side in the second case, and corticosteroids had already been given to the patient for 3 years, it's still questionable whether corticosteroids could bring about a long-lasting solution to arterial stricture.

As to treatment, steroid therapy has been reported to be effective in most cases. However, recurrence after steroid therapy was not uncommon. Surgery is recommended as the major treatment approach for IMT in the head and neck. Besides, radiotherapy can also play a significant role in the treatment modalities. When IMT takes place at the carotid bifurcation, total surgical removal can be tough because the scar-like lesion may make the dissection plane more difficult to confirm than in a carotid body tumor surgery [10]. For those patients with IgG4-related IMT, corticosteroids may be a good choice, but the dosage and administration plan have not been optimized because of the paucity of knowledge on this disease. In case 2, the lesion in the patient combined with other systemic diseases, so surgical removal of the tumor was not accomplished.

Compared with CT, MRI of IMT may provide potential advantages via its multiplanar capabilities and differentiation of three layers of the carotid wall. It has been proposed that the mature fibrotic plaque tends to be hypointense on both the T1- and T2-weighted imaging, while the high signal abnormality is deemed to represent edema or cellular infiltrate [3]. But this can't explain why a mature tissue such as a scar or collage bundles are still more capable of infiltrating the carotid artery wall than tissues with an abundance of inflammatory cells. We could hypothesize that the fusiform lesion resulted from inflammtory cell infiltration, while myofibroblast and fibrous tissue accumulation progressed from both circular and longitudinal direction along the carotid artery. As a matter of fact, we have pathologically studied the entire paraffin-embedded specimen stripped from the carotid artery. In the slices, we found a compound of cell rich and cell sparse regions. This made us question whether or not the lesion of the low signals zone in MR or hypointense zone in CT had been completely resected due to the difficulty of dissection. Therefore, we propose that even if IMT may exhibit a variable appearance in signal intensity according to its staging, the exact cause of the prominent contrast enhancement of the mass on MR is still uncertain. It's interesting that, among all papers that declare radiological findings to be nonspecific, none has contained IMT of the carotid artery [1, 19, 20]. This two rare cases

implies that the CT or MR imaging characteristics of IMT of the carotid artery are more unique than those in other sites of head and neck [2, 20-26]. IMT shall be taken into account when a solitary fusiform mass is found around the carotid bifurcation.

Disclosure of conflict of interest

None.

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