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Research Article

TIPIC SYNDROME AND AMAUROSIS FUGAX IN A PATIENT WITH GIANT CELL ARTERITIS AND POLYMYALGIA RHEUMATICA

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Abstract

TIPIC Syndrome (Transient perivascular inflammation of the carotid artery) is a rare idiopathic neck pain syndrome, which characterized by unilateral pain in the region of the carotid bifurcation.We present the case of a 60-year-old patientwith Giant cell arteritis and Polymialgia Rheumatica who had a retinal transient ischemic attack, complaining of a rare simptom such as carotidynia, among others. TIPIC Syndrome has been reported in 7% [1]of Giant cell arteritis cases. TIPIC syndrome is a diagnosis of exclusion, and the following diagnoses should be excluded: aterothrombosis, fibromuscular dysplasia, aneurysm, dissection, lymphadenitis, submandibular gland disease, and neck cancer. Stroke is uncommon in Giant cell arteritis, but transient retinal ischemic attacks occur often in this entity[2]. Neurologists should always consider Giant cell arteritis in the differential diagnosis of a new-onset severe headache in patients ≥ 50 years-old with an elevated erythrocyte sedimentation rate (ERS).

Keywords: TIPIC Syndrome, Carotidynia, Giant cell arteritis, Amaurosis fugax

INTRODUCTION

Case Report

A 68-year-old man with Polymialgia Rheumatica, presented with a 2- week history of left cervical pain radiating from the carotid artery bifurcation to the left ear, exacerbated by head movements and swallowing, fatigue and loss of appetite. He also experienced severe throbbing bilateral temporo-parietal headache, scalp allodynia, which had not disappeared completely despite treatment with NSAIDs. After 2 weeks, symptoms such as distal paresthesia of the extremities on both sides and jaw claudication were added. The patient appeared for neurologic consultion one day after experiencing an episode of left amaurosis fugax lasting approximately 30 minutes. Physical examination revealed a pulseless, thickened left temporal artery and a tender left carotid artery. Laboratory tests revealed leukocytosis (WBC 12.20g/l), elevated platelets count (PLT 570 g/l), mild microcytic anemia (HGB 12.8 g/dl), raised erythrocyte sedimentation rate (ESR 55 mm/h), increased concentration of the systemic inflammatory marker (C-reactive protein 89.5 mg/l), fibrinogen 547 mg/dl. The thyroid hormones were within the normal limits. Blood cultures were sterile. Investigations for connective tissue diseases and autoimmune diseases (ANCA, ANA) were all negative. Cervical ultrasound showed a hypoechoic thickening of the vessel wall of the left carotid artery, in the absence of atheromatous plaques. In the angio-computed tomography of the head, except for bilateral calcification in traps of the internal carotid artery, there were no other lesions. Based into the information obtained from the clinical presentation and the overall diagnostic studies, the diagnosis of Horton's disease with Carotidynia was established.

Treatment began with 60 mg daily of prednisone and aspirin 100 mg daily. The patient was also started on celexocib, with symptoms improvement within approximately 17 days.

Introduction

Giant cell arteritis (GCA) is a systemic inflammatory autoimmune disease of large and medium sized- blood vessels. Horton's disease (temporal arteritis) is a distinct entity, caused by giant cell arteritis of the temporal arteries, causing a wide range of neuro-ophthalmological complications. The intracranial vessels are usually not affected[3]. GCA mostly affects the temporal superficial artery, aorta, the carotid, subclavian and iliac arteries. Women are more affected than males (ratio 2:1) and the mean age of onset is above 55 years old[4]. Neurologists should always consider GCA in the differential diagnosis of a new-onset severe headache in patients ≥ 50 years-old with an elevated ERS.

Systemic symptoms are common in GCA and vascular involvement can be widespread, causing stenosis and aneurysm of affected vessels.

Clinical manifestations of Giant cell arteritis

The onset of symptoms in giant cell arteritis is subacute, acute presentations over a few days have also been raported in literature. Although many of the clinical manifestations of GCA are nonspecific, some characteristic findings strongly suggest the diagnosis.

Some of the clinical feaures of Giant cell arteritis are shown on Table 1[5].

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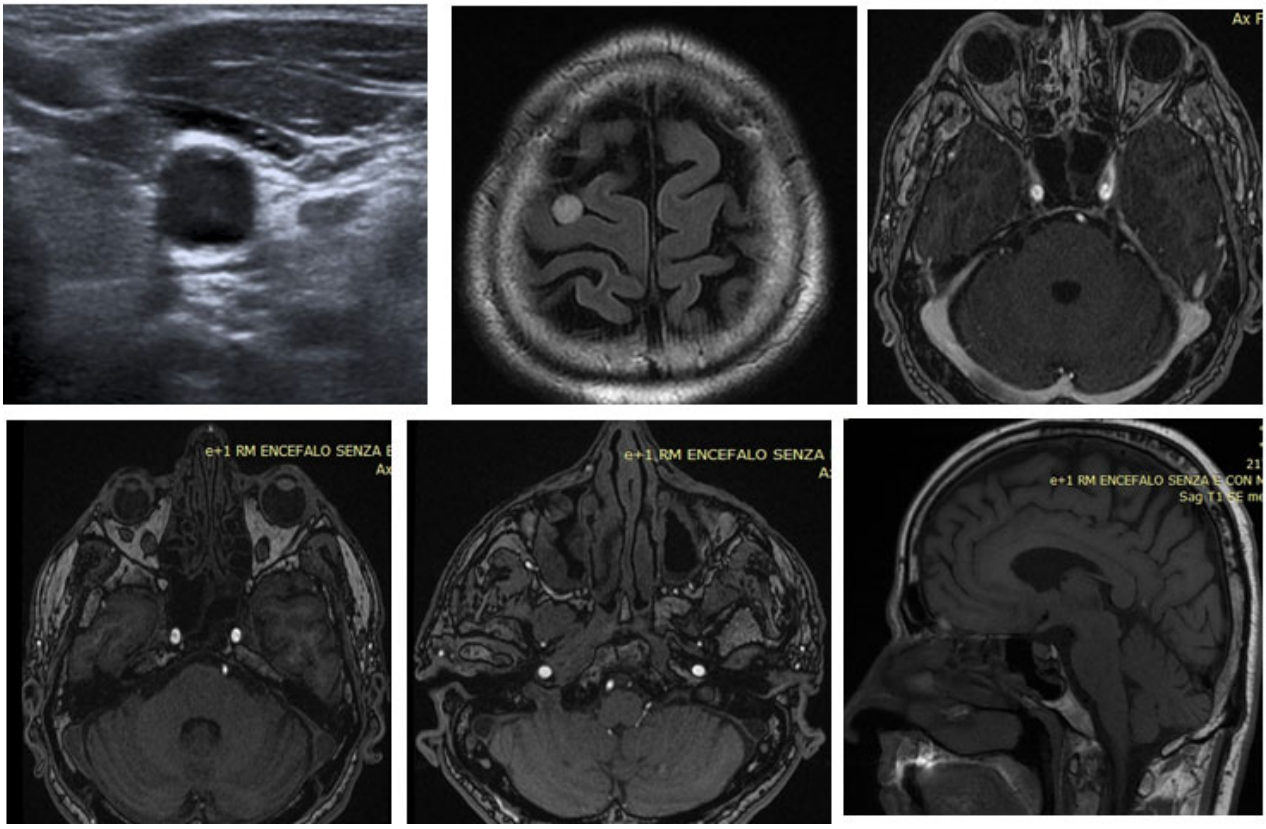

Figure 1. Imaging of the patient. Cervical ultrasound showed a hypoechoic thickening of the vessel wall of the left carotid artery, in the absence of atheromatous plaques . Magnetic Resonance revealed right frontal meningioma, right Schwannoma n.VII and right intraorbital cavernous angioma. Intracranial vessels appear normal

Clinical manifestations of Giant cell arteritis

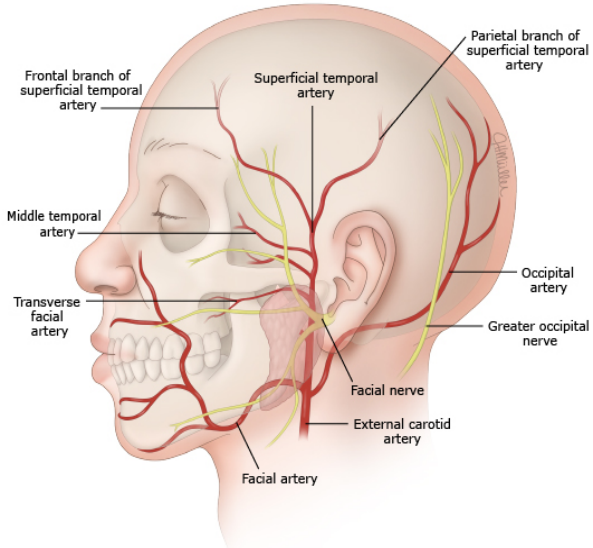

Figure 2. Superficial temporal artery

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Superficial temporal artery is a terminal branch of the external carotid artery. The superficial temporal artery originates in the parotid gland. It divides into two branches: a. Frontal (anterior temporal) and b. Parietal (posterior temporal)

Superficial temporal artery gives off several branches that include: - Parotid branch - Transverse facial artery - Anterior auricular branch - Zygomatico-orbital artery - Middle temporal artery - Frontal branch - Parietal branch

Polymialgia Rheumatica and Giant cell arteritis

Polymialgia Rheumatica (PMR) is a form of inflammatory arthritis that mainly affects the shoulder girdle that causes pain and stiffness, which are worse in the morning. Approximately 5-15% of the patients with Polymialgia Rheumatica have Giant cell arteritis[6]. The precise nature of the relationship between GCA and PMR is not completely understood. In some patients, symptoms and signs of the two conditions occur simultaneously, while in others they appear separately over time.

The American College of Rheumatology (ACR) has established criteria for the diagnosis of GCA[7] (Table 2)

TIPIC syndrome (Transient Perivascular Inflammation of the Carotid Artery)

Carotidynia, also known as TIPIC syndrome (Transient Perivascular Inflammation of the Carotid Artery) is characterized by unilateral neck pain in the region of the carotid bifurcation. It is a rare syndrome, first described in 1927[8] by Temple Fay, an American neurologist and neurosurgeon. Carotidynia worsens with palpation in the area of the carotid bifurcation, with cervical movements, coughing or swallowing. International Headache Society (IHS) classified TIPIC Syndrome as an idiopathic neck pain syndrome, with tenderness over the carotid bifurcation but without structural abnormality. TIPIC is an idiopathic and self-limited clinical entity with a prevalence of 2.8% [9]in patients presenting with acute neck pain. Symptoms resolve in about 2 weeks and in 10% of the cases they appear on both sides.

Table 1. Clinical manifestations of Giant cell arteritis

Clinical manifestations of Giant cell arteritis	
Constitutional symptoms	Fever, fatigue, weight loss
Headache	2/3 of the cases, mostly temporal
Jaw claudication	½ of the cases, mandibular pain or fatigue brought on by mastication, tongue claudication
Ocular involvement	
• Transient visual loss (amaurosis fugax)	An early manifestation of GCA
• Permanent vision loss	15 to 20 percent of patients
• Anterior ischemic optic neuropathy (AION)	At least 85 percent of cases of vision loss in patients with GCA are caused by AION.
• Central retinal artery occlusion (CRAO)	CRAO is a less common cause of visual loss in GCA.
• Posterior ischemic optic neuropathy (PION)	is an unusual occurrence in GCA,
Cerebral ischemia	occipital lobe infarction resulting from a lesion in the vertebrobasilar circulation.
Ophthalmic syndromes	
• Diplopia	in approximately 5 percent of patients with GCA
• The Charles Bonnet syndrome	rare, visual hallucinations
Musculoskeletal involvement	proximal polyarthralgias and myalgias
Large vessel involvement	Aorta and its major proximal branches, especially in the upper extremities. Aortic aneurysm or dissection
Central nervous system involvement	Stroke is uncommon in GCA.
Upper respiratory tract symptoms	Nonproductive cough, Vasculitis of the bronchial arteries
Atypical features	Dysarthria, Sensorineural hearing loss, Pericarditis

Table 2. Diagnostic criteria of Giant cell arteritis

The presence of three or more out of five criteria
age ≥ 50 years at disease onset
temporal artery tenderness or decreased temporal artery pulse
new onset of localized headache
ESR ≥ 50 mm/h
artery biopsy

Many inflammatory markers are elevated in GCA, including :

- C-reactive protein (CRP)
- Erythrocyte sedimentation rate (ESR)
- Serum amyloid A protein (SAP)
- Fibrin degradation products and D-dimer
- Eccentric thickening of the carotid wall/perivascular tissues, without any hemodynamic alterations
- Enhancement (CT, and more so MRI)- An increased uptake of gadolinium in the carotid wall and the absence of atheromatous plaques can be seen in the MRI.

"King Kong carotid" has been suggested by some neuroradiologists as an imaging sign of TIPIC syndrome, with eccentric thickening of the tissues around carotid artery with narrowing of the lumen.

Carotidynia is associated with multiple diseases, as shown on Table 3.

Table 3. Diseases that are associated with carotidynia

Migraine (most common)
Diseases associated with carotidynia
Thrombosis or atheromatosis
Lymphadenitis
Peritonsillar abscess
Sialadenitis
Aortic dissection
Large vessel vasculitis (giant cell arteritis, Takayasu arteritis)
Cervical neoplasia
Jugular vein thrombosis
Head tumors
Submandibular gland diseases
Thyroiditis

Diagnostic criteria for Carotidynia/ TIPIC Syndrome

- Unilateral neck pain with irradiation to the neck
- Presence of focal tenderness or increased pulsation over the carotid artery
- Absence of a structural lesion
- Recovery within 14 days of the onset of symptoms

DISCUSSION

Transient Perivascular Inflammation of the Carotid Artery (TIPIC) syndrome or Carotidynia or is a rare cause of atypical unusual neck pain. The aetiopathogenesis of this syndrome is not clear. TIPIC syndrome is characterized by a transient perivascular inflammation of the carotid artery, that responds well to non- steroidal anti inflammatory drugs. Imaging is the gold standart for diagnosis of Carotidynia.

Our case is in agreement with the four major criteria of TIPIC syndrome suggested by Lecler et al:

1. The presence of acute pain overlying the carotid artery, with irradiation to the neck
2. Eccentric perivascular inflammation on imaging
3. Exclusion of another vascular disorders
4. Improvement within two weeks

Retinal transient is chemic attack is an early manifestation of Horton syndrome. With transient monocular visual loss (TMVL), affected patients typically note an abrupt partial field defect or temporary curtain effect in the field of vision of one eye. Patients with both polymyalgia rheumatica and GCA are often predisposed to the potential for vision loss.

Transient visual loss can be a harbinger of permanent visual loss, and thus mandates urgent attention in a patient. Giant cell arteritis can result in granulomatous inflammation within the central retinal artery resulting in partial or complete occlusion, and leading to decreased blood flow manifesting as amaurosis fugax.

Stroke is uncommon in Giant cell arteritis. In descriptive cohorts, the frequency of stroke within the ranges from 1.5 to 7.5%. Stoke due to GCA can occur in the distribution of both the internal carotid and vertebrobasilar arteries, but they are more common in the latter location. More than one-half of strokes attributable to GCA occur in the vertebrobasilar system. Intracranial involvement is rare.

Conclusion

In summary, we present an ischemic transitory attack in a patient with Giant cell arteritis and Polymialgia Rheumatica, presenting with a rare simptom such as carotidynia, among others. Carotidynia or TIPIC syndrome is a rare syndrome caused by a transient perivascular inflammation of the carotid artery. International Headache Society (IHS) classified TIPIC Syndrome as an idiopathic neck pain syndrome, with tenderness over the carotid bifurcation but without structural abnormality. TIPIC syndrome is a diagnosis of exclusion, and the following diagnoses should be excluded before the diagnosis is made: giant cell arteritis, thrombosis, arteriosclerosis, fibromuscular dysplasia, dissection, aneurysm, lymphadenitis, submandibular gland disease, and neck cancer. TIPIC Syndrome has been reported in 7% of GCA cases. Stroke is uncommon in Giant cell arteritis, but transient retinal ischemic attacks occur often in this entity. Imaging is the gold-standard investigation for the diagnosis of TIPIC syndrome. Neurologists should always consider GCA in the differential diagnosis of a new-onset severe headache in patients ≥ 50 years-old with an elevated ERS.

Conflicts of interest: None

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