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Case Report

“Transient perivascular inflammation of the carotid artery (TIPIC) syndrome” as a rare case of laterocervical pain: Multimodal diagnosis [☆]

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ABSTRACT

“Transient Perivascular Inflammation of the Carotid artery (TIPIC) syndrome” is an unusual cause of unilateral neck pain, due to a nonspecific inflammation of the carotid artery. This entity has been for long known as “carotidynia” and described as a syndrome rather than a distinct pathologic entity. Recently, the presence of structural abnormalities of the carotid artery wall has been demonstrated, leading to the introduction of radiological criteria which, in the appropriate clinical context, allow to diagnose TIPIC syndrome. TIPIC syndrome is a rather rare disease and, since its first description by Fay in 1927, only a small series of patients have been published. The interest of our case lies in the fact that diagnosis and follow-up were assessed on ultrasound and magnetic resonance imaging, demonstrating that a correlation between clinical evolution and radiological findings does exist. In addition, DWI sequence was performed at the time of diagnosis and at resolution. To our knowledge, such an assessment has never been reported in the previous literature.

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Case report

We are reporting the case of a 49-year-old man presented with a 1-week history of pain in the right laterocervi-

cal region, over the carotid bifurcation. The pain was described as a severe dull sense of discomfort, irradiated to the ipsilateral ear, and triggered by head and neck movements (“Fay sign” [1]). This was preceded by mild constitutional symptoms, with fever, fatigue, and myalgia.

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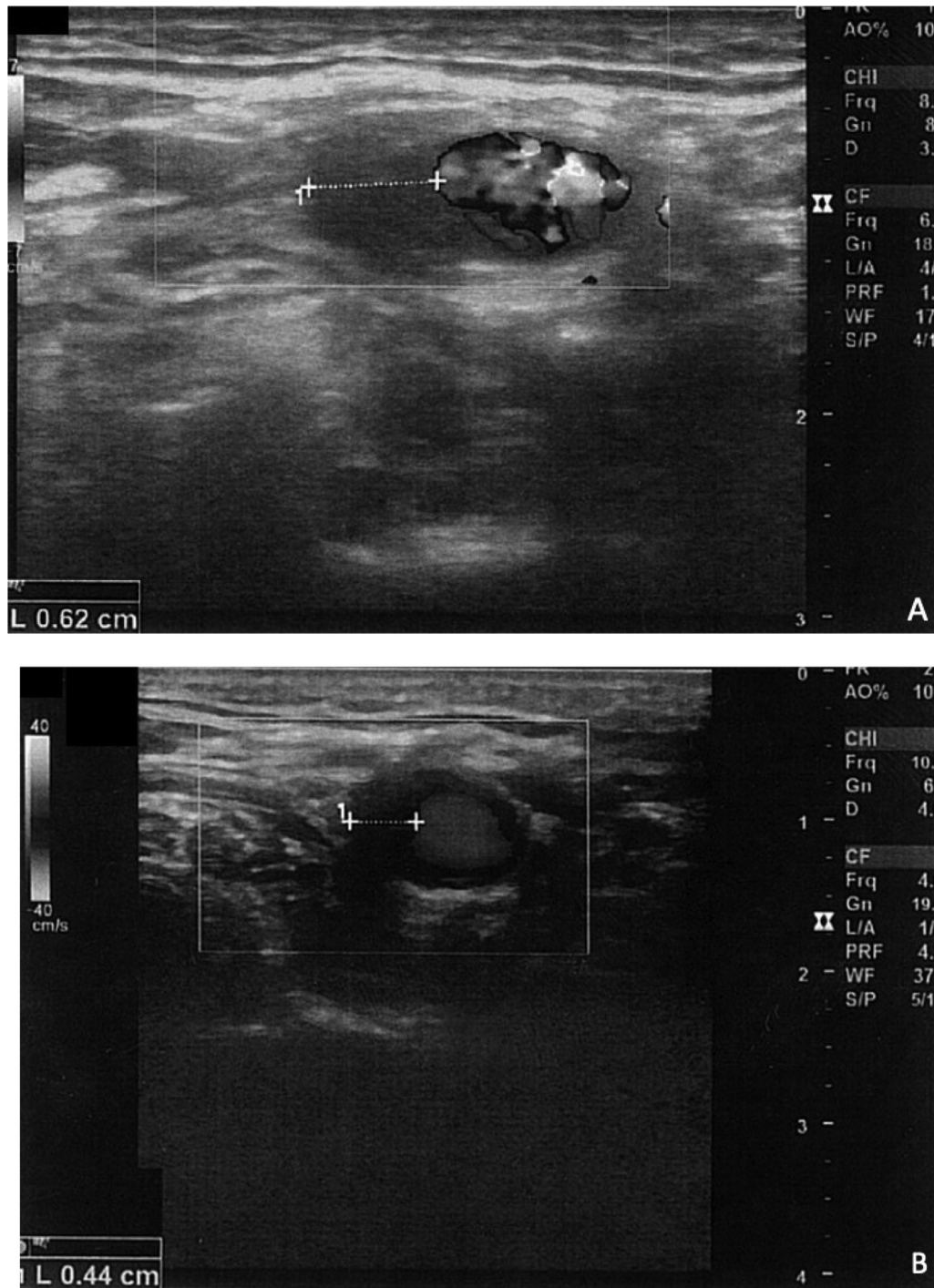


Fig. 1 – Doppler ultrasound (A) shows an eccentric and laterally developed hypoechoic adventitial thickening of the right internal carotid. Intimal plaque and stenosis are absent. Regression of the hypoechoic thickening is found after 3 wk of treatment with non-steroidal anti-inflammatory drugs (B).

Patient denied history of migraine or other neurological symptoms.

Clinical examination found an apyretic patient in a good general condition, with localized swelling and pain at the level of the right carotid bifurcation, worsened by palpation. There was no palpable cervical node, no palpable induration along the jugular vein and no carotid bruits were audible.

Basic laboratory hematological examination showed normal white blood cell count ($9910/\text{mm}^3$) and C-reactive protein ($0.10 \text{ mg}\%$). Erythrocyte sedimentation rate (20 mm/h) was slightly increased. The rest of the laboratory investigations was normal.

US and MRI of the neck were performed approximately 1 week after symptoms onset.

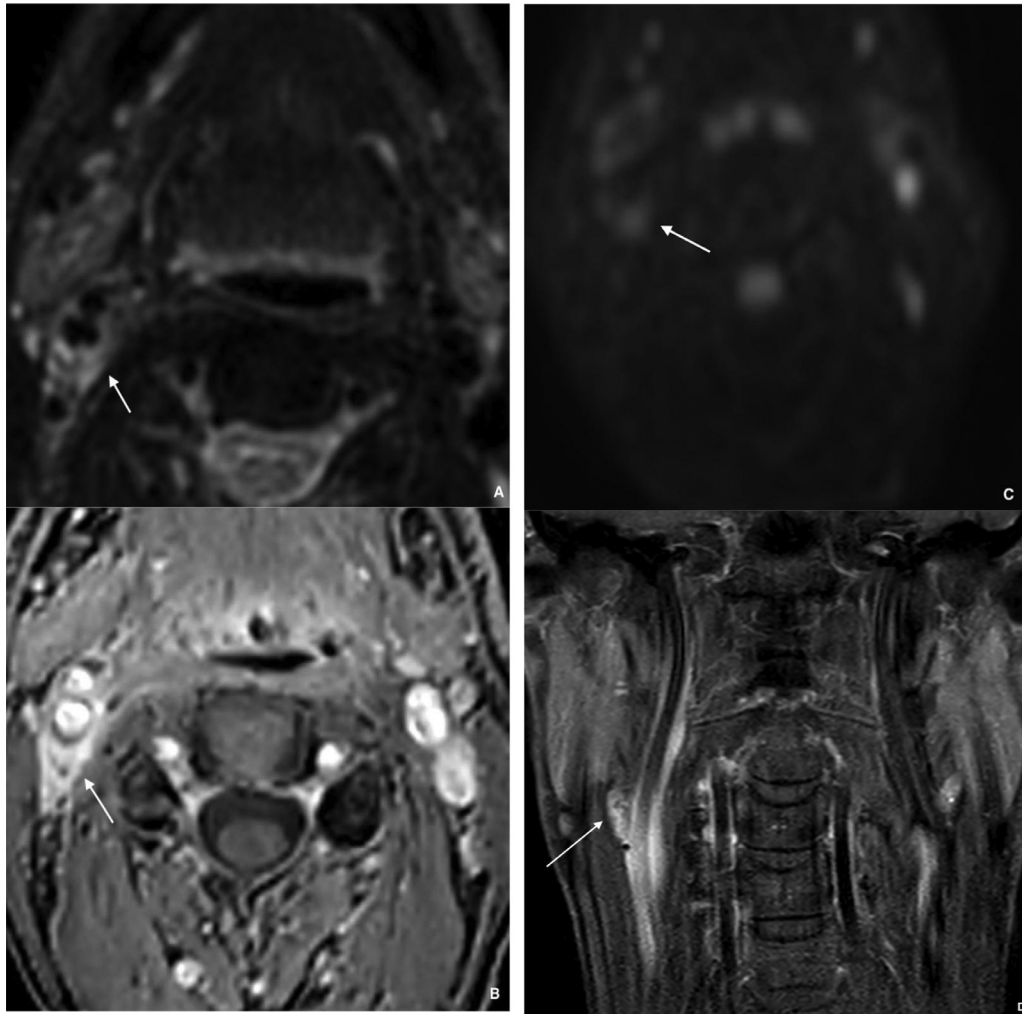


Fig. 2 – (A, B, C and D) axial STIR, axial Ultrafast spoiled GE, DWI and coronal postcontrast images at the level of the right carotid artery show an abnormal soft tissue signal surrounding the carotid artery.

US showed the presence of an asymmetric hypoechoic thickening of the right carotid wall, localized at the level of the distal right common carotid artery, extending into the proximal internal carotid artery (Fig. 1A).

On Doppler studies, the affected carotid artery on the right side demonstrated normal flow parameters (Fig. 1A). No left carotid artery or other vascular abnormalities (stenosis or dissection) were found.

The thyroid gland, parathyroid glands and salivary glands were normal, and no neck mass or cervical lymphadenopathy was noted.

Further evaluation with MRI was performed to characterize the lesion found on US. MRI demonstrated the presence of a thin amount of hyperintense tissue on STIR sequence, which restricted diffusion on DWI sequence, and showed contrast enhancement after contrast medium administration (Fig. 2A, B, C, D). Significant luminal narrowing was absent.

The findings described confirmed the hypothesis of inflammatory tissue in the periadventitial area of the right carotid artery and the diagnosis of TIPIC syndrome was formulated.

The patient was treated with anti-inflammatory medications and had a full clinical recovery within 14 days. On follow-up, 3 weeks after presentation, US (Fig. 1B) and MRI (Fig. 3A, B, C, D) showed the regression of the eccentric perivascular tissue.

Discussion

Carotidynia is an idiopathic unilateral neck pain syndrome, caused by a nonspecific inflammation of the carotid artery. It usually lasts less than 2 weeks, being self-limited or resolving with nonsteroidal anti-inflammatory drugs or steroids.

It was described for the first time by Fay in 1927 as a clinical entity characterized by tenderness and pain at the level of the carotid bifurcation [1]. In 1988, it was included in the first International Classification of Headache Disorders [2] and, in 2004, the International Headache Society published modified criteria for carotidynia, classifying it as a syndrome rather than a distinct pathologic entity: the criteria specified that pa-

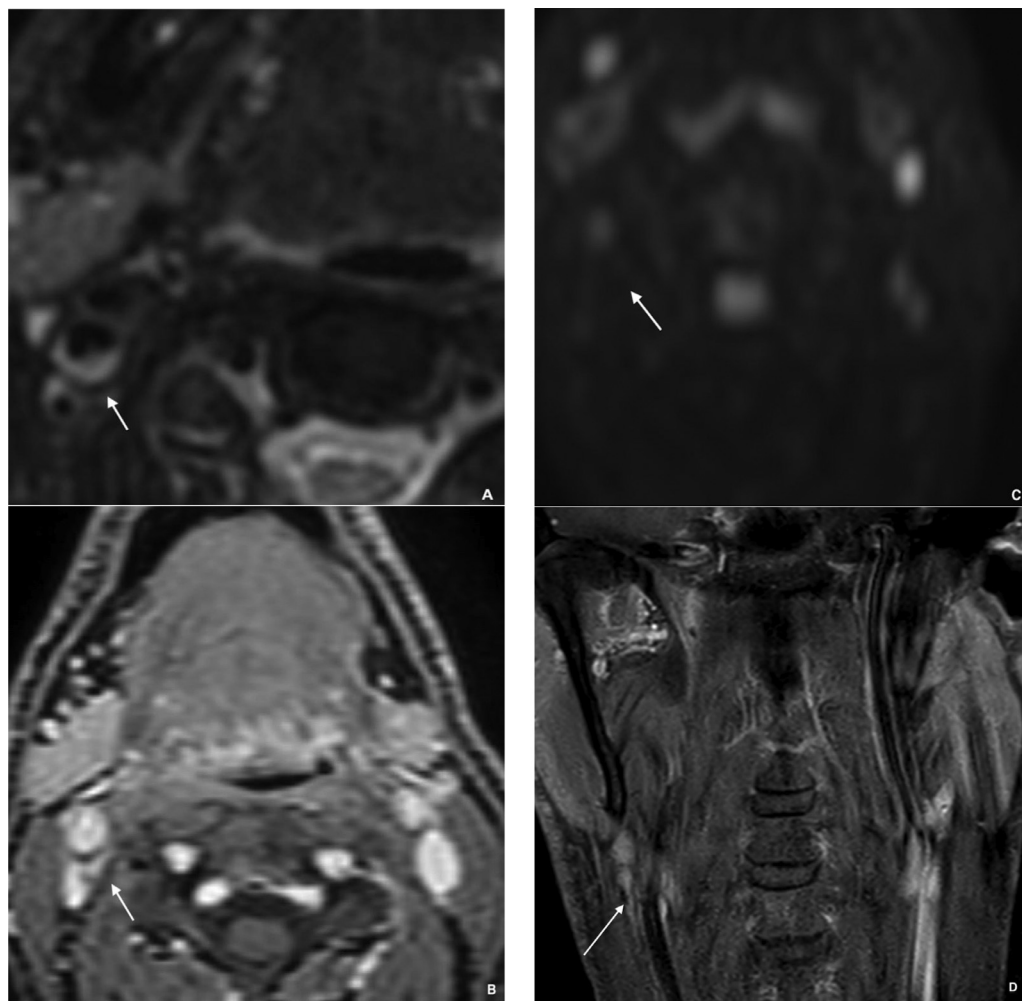


Fig. 3 – (A, B, C and D) axial STIR, axial Ultrafast spoiled GE, DWI and coronal postcontrast images at the same level of Fig. 2, obtained 3 wk later, when the patient had become asymptomatic, show resolution of the abnormal soft tissue signal.

tients with carotidynia should not have had structural abnormalities of the carotid artery [3].

Recently, consistent imaging findings were reported, particularly on ultrasound (US) and magnetic resonance imaging (MRI), demonstrating that radiological abnormalities of the carotid bifurcation zone, evidencing an inflammatory process, are present [4]. As a result, the condition of carotidynia is currently defined as the combination of specific clinical and imaging findings and the acronym “Transient Perivascular Inflammation of the Carotid artery (TIPIC)” syndrome has been introduced to describe the entity as thoroughly as possible [5,6].

TIPIC syndrome is a rather rare disease. Precise epidemiological data is not available. A large study of 47 patients with acute neck pain, published by Lecler et al., reported a prevalence of 2.8% [5].

The etiology and the pathogenesis of the inflammatory process is not clear and only 1 study reported histologically proved findings of non-specific vascular inflammation of the carotid adventitia [7].

Clinical presentation includes unilateral cervical pain of acute onset, occasionally with temporal irradiation, triggered

by palpation or head and neck movements. Transient neurological symptoms or constitutional symptoms are rarely reported.

In the vast majority of patients, biologic examinations show a mild increase of the inflammatory markers.

The diagnosis is, therefore, mainly based on clinical and imaging findings.

Four diagnostic criteria have been proposed:

- (1) Presence of acute pain overlying the carotid artery, which may or may not radiate to the head;
- (2) Eccentric PeriVascular Infiltration (PVI) on imaging;
- (3) Exclusion of another vascular or nonvascular diagnosis with imaging;
- (4) Improvement within 14 days either spontaneously or with anti-inflammatory treatment.

Additionally, a minor criterion could be the presence of a self-limited intimal soft plaque [5].

Our case respected all the criteria.

Imaging findings of TIPIC syndrome include perivascular findings described by the general term “PeriVascular Infiltration (PVI)”, referring to the presence of soft amorphous tissue

Table 1 – Differential diagnosis of unilateral neck pain.

Vascular	Non vascular
Dissection	Neck abscess
Giant cell arteritis	Lymphadenitis
Thrombosis	Thyroiditis
Fibromuscular dysplasia	Migraine
Aneurysm	Neuralgia of trigeminal nerve
Vessel wall or intraplaque hematoma	Eagle syndrome

replacing the fat surrounding the carotid artery, with a hazy aspect of the fat. PVI is primarily located at the level of the carotid bifurcation, most often in a posterior and lateral location, and may extend towards the proximal internal or external carotid artery [2,5]. This lesion does not affect the entire circumference of the carotid system but is usually limited to less than half of the perimeter, thus being characterized as eccentric.

On US, PVI appears as a hypoechoic lesion situated in the medial-adventitial layer of the carotid artery, without hemodynamic changes on Color Doppler technique. The lack of hemodynamic disturbance justifies the absence of audible bruit during auscultation.

MRI evidences a thickened wall of the affected carotid artery, due to the presence of periadventitial soft tissue, which shows enhancement after administration of contrast medium [8] (Fig. 2B and 3B). Additional T2 spectral presaturation with inversion recovery sequences, when performed, reveal a narrow, perivascularly raised, signal corresponding to an inflammatory concomitant oedema, as we observed in our patient (Fig. 2A and 3A). The MR angiograms of the neck vessels do not show any sign of significant lumen constriction [9]. In our case, we performed a previously unreported sequence, DWI, which demonstrated that the perivascular tissue restricted diffusion, strengthening our diagnostic hypothesis. We also used DWI sequence to prove the resolution of pathological findings (Fig. 2C and 3C).

The diagnosis of TIPIIC syndrome requires imaging evidence of the exclusion of other vascular and non-vascular causes of neck pain (Table 1) [10,11]. As compared with US, MRI is particularly useful in differentiating TIPIIC syndrome from intramural hematoma or carotid dissection, that are the principal vascular differential diagnoses. In contrast to these two latter causes of acute neck pain, TIPIIC syndrome has a benign clinical course, as it may be self-limiting or be treated with anti-inflammatory drugs or steroids, showing full clinical recovery within a mean period of 2 weeks. A relapse rate of about 20% was reported [5].

The case we reported represents a characteristic case of TIPIIC syndrome, in terms of clinical-radiological findings and benign course. Its main interest lies in the fact that diagnosis and follow-up were assessed on US and MRI, demonstrating that a correlation between clinical evolution and radiological

findings does exist. In addition, DWI sequence was performed at the time of diagnosis and at resolution (Fig. 2C and 3C). To our knowledge, such an assessment has never been reported in the previous literature.

In conclusion, in the appropriate clinical context, a diagnosis of TIPIIC syndrome as a cause for vascular neck pain may be supported by characteristic radiologic findings. This case report supports the imaging features of TIPIIC syndrome previously described in the literature and underscore the role of Doppler sonography and neck MRI, including DWI sequence, as the modalities of choice to confirm the presumed TIPIIC syndrome diagnosis and to evaluate the natural history of the pathology.

Patient consent

Informed written consent was obtained from the patient for publication of the Case Report and all imaging studies. Consent form on record.

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