CC107. Painful unilateral facial swelling due to superficial temporal artery thrombosis: a rare presentation of Antiphospholipid Syndrome

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Introduction

Persistent painful unilateral temporal swelling is rarely seen in clinical practice. Antiphospholipid syndrome (APS) is an autoimmune and systemic disorder that causes changes in blood clotting homeostasis, marked by arterial or venous thrombosis, gestational morbidity, and high and persistently positive serum levels of antiphospholipid antibodies (aPL). APS is more common in young women and middle-aged adults, with no preference for race. Among its clinical features, temporal artery thrombosis, associated with headache and temporal and hemifacial edema is extremely rare, with few publications worldwide on this topic.

Objectives

This report aims to present the case of a unilateral painful facial edema due to thrombosis of the superficial temporal artery as an unprecedented manifestation of Antiphospholipid Syndrome. Thus, the only record in the English-language literature surveyed in the Pubmed database in July 2022 highlights the rarity of this APS presentation and the consequent challenge in suspecting the correct diagnosis for adequate treatment.

Case description

A 32-year-old woman was presented with pain in the right temporal region of her face. The pain was intense, daily and continuous, pulsating, without irradiation, which worsened with physical activity and presented partial relief with common analgesics. After 20 days, she developed a right pale temporal edema (Figure 1) associated with a significant worsening of pain and intense right unilateral headache attacks triggered by chewing and speaking. She has a history of deep vein thrombosis in the left lower limb. Physical examination and imaging tests showed significant cold edema of the right temporal region with asymmetry of the temporal muscles, which was extremely painful on palpation and made it difficult to open the mouth. There were no other changes in the general physical or neurological examination. During evolution, hypertrophy of the masseter muscles on the right was also noted. She presented erythrocyte sedimentation velocity (ESR) tests with high values, subcutaneous edema in the right temporal region on magnetic resonance imaging (MRI) of the brain, and a biopsy of the right temporal artery revealed a residual histological picture of the previous thrombosis. In the case of

suspected hematological disease, serial investigations were performed for aPL markers, which were positive, and for systemic lupus erythematosus, which was negative. Evolved with improvement of edema and pain with the use of indomethacin and low molecular weight heparin. Data disclosure was authorized by the patient through an informed consent form.

Conclusions

This report demonstrates a persistent painful unilateral temporal swelling due to a temporal artery thrombosis resulting from APS. This is a very rare presentation of this condition, highlighting the importance of a high level of clinical suspicion for directing laboratory investigation and appropriate treatment.

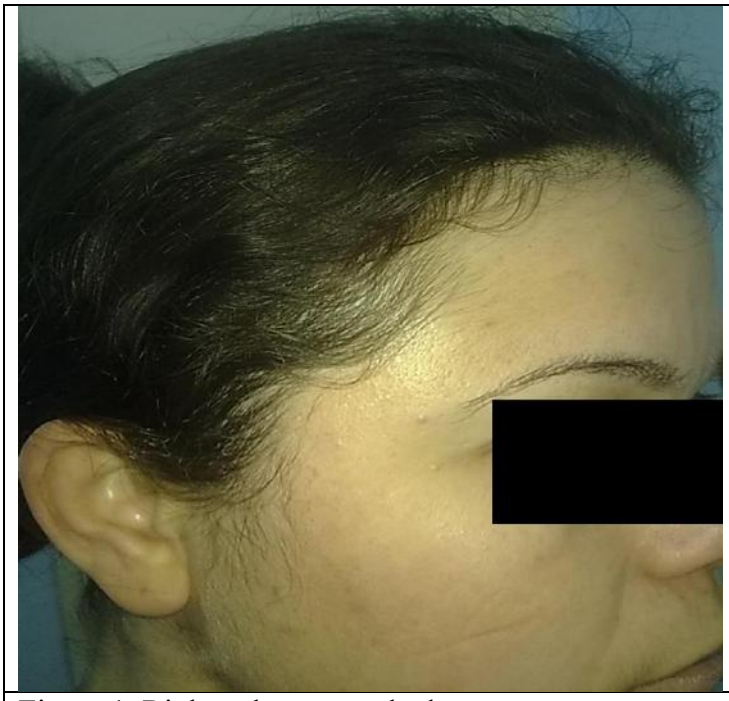

Figure 1. Right pale temporal edema.