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NEURO-IMAGES

Giant cell arteritis: the skin as the key to the diagnosis

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Case description

An 82-year-old male patient with HIV-1 infection with undetectable viral load was seen at the Neurology Outpatient clinic due to fluctuating binocular diplopia and difficulty in feeding interpreted as dysphagia. An initial diagnosis of ocular and bulbar myasthenia gravis was assumed, and the patient was started on pyridostigmine and prednisolone 5 mg daily, with temporary resolution of symptoms.

Two months after the first visit, the patient started feeling pain over his right frontoparietal region, where he later developed an extensive necrotic ulceration (Fig. 1) in less than a week. On physical examination, no superficial temporal pulses were felt, suggesting an alternative diagnosis of Giant Cell Arteritis (GCA). By that time, the previously assumed diagnosis of Myasthenia Gravis had already been ruled out as a single fiber electromyography did not show signs of neuromuscular plaque dysfunction, and serum anti-AChR and anti-Musk antibodies were undetectable. An erythrocyte sedimentation rate of 78 mg/l, the presence of the halo sign on temporal artery duplex ultrasonography, and an FDG-PET (Fig. 2) displaying extensive vascular inflammation confirmed the alternative diagnosis. Prednisolone dosage was increased to 1 mg/kg/day, and subcutaneous tocilizumab 162 mg every other week was started.

Comment

Giant cell arteritis is the most common primary systemic vasculitis in elderly people: it is rarely diagnosed in patients younger than 50 years old and peaks in the eighth decade [1]. GCA is characterized by granulomatous inflammation of medium and large vessels with tropism for external carotid arteries branches [2]. Twenty percent of patients may display abrupt onset of symptoms with initial disease manifestations attributable to localized effects of vascular and systemic inflammation, mainly presenting with ischemic cranial manifestations such as headache, scalp tenderness, vision loss, diplopia, and jaw claudication (conditioning feeding difficulty) [1].

Reports on dermatological manifestations of GCA are rare and include the following: ischemic mucocutaneous

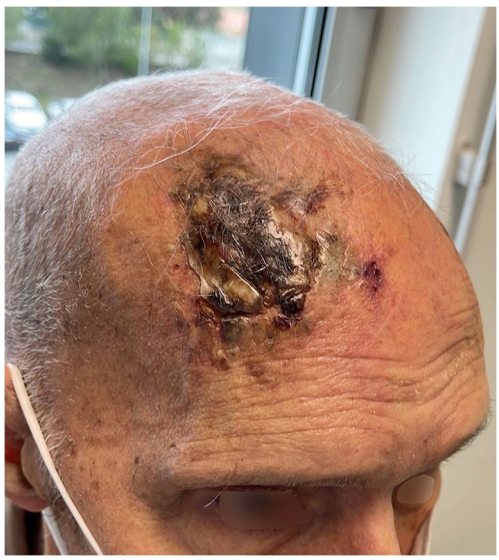

Fig. 1 Scalp necrotic ulceration

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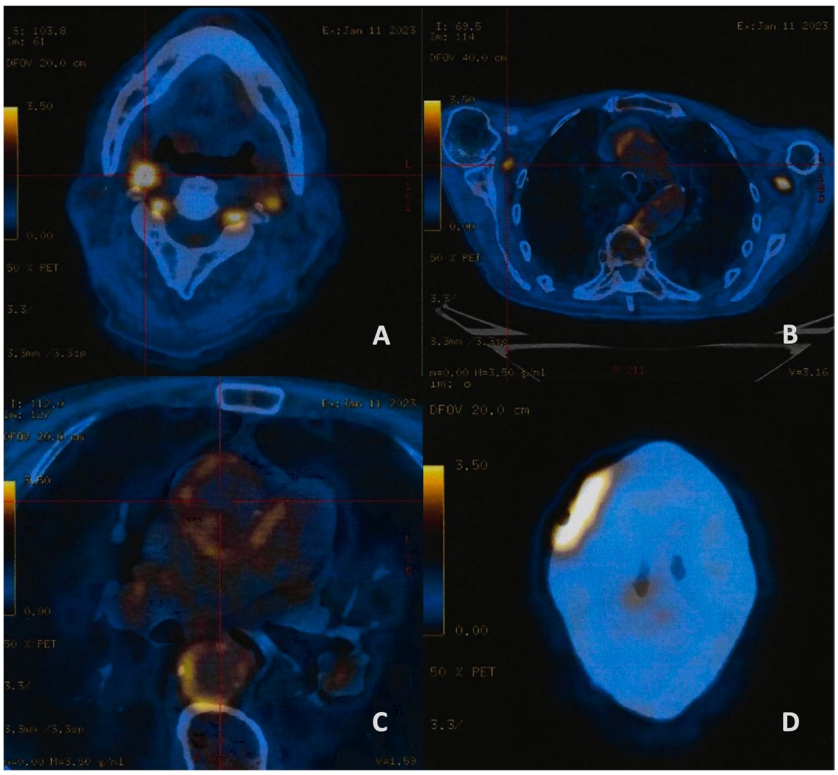

Fig. 2 FDG-PET image shows extensive vascular inflammation with carotid, vertebral (**A**), and aortic (**B**, **C**) involvement. Superficial temporal arteries were also reported to display hypermetabolism. The right anterior aspect of the scalp (**D**) displayed intense uptake (standardized uptake value of 4.7) suggestive of an inflammatory etiology

findings (scalp, tongue and lip necrosis), nodules and panniculitis-like lesions (cutaneous nodules and lesions resembling erythema nodosum), and cutaneous diseases associated with vasculitis (generalized granuloma annulare and basal cell carcinoma) [2]. Scalp necrosis results from multivessel occlusion of temporal, frontal, retro-auricular and occipital arteries, and it is present in severe forms of GCA [3]. It is considered a prognostic factor signaling higher risk of vision loss (present in up to 31.5% of patients) and higher mortality (ranging from 20.2% to 38% in previous case series) [2, 4–7]. Patients with GCA presenting with scalp necrosis need emergent treatment with high doses of glucocorticoids, which slow disease progression but are unable to avoid surgery is scalp necrosis is already established [2]. In such cases, surgical debridement, and in some instances, skin-grafts, may be necessary [2]. Nowadays ischemic cutaneous lesions in GCA are seldomly reported due to early diagnosis and prompt immunosuppressive treatment, with no previous reports of scalp necrosis recurrence after the onset of treatment [2].

Author contribution Miguel Miranda conceived and designed the analysis, collected the data, and wrote the paper. Rita Ramos Pinheiro and Cátia Carmona reviewed the paper. All authors discussed the results and contributed to the final manuscript.

Data availability Not applicable.

Declarations

Ethical approval This article presents a clinical case. This article is in accordance with the ethical standards of the institutional research committee and with the Helsinki declaration and its 2013 amendments.

Consent to participate Informed consent was obtained directly with the patient.

Conflict of interest The authors declare no competing interests.

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