IMAGES IN RHEUMATOLOGY

Orbital apex syndrome mimicking giant cell arteritis

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INTRODUCTION

Giant cell arteritis (GCA) is a large- and medium-sized vessel vasculitis that mainly affects older people[1]. It typically manifests as constitutional features, headache, vision disturbances, and elevated inflammatory markers. Around half the patients can present polymyalgia rheumatica (PMR)[1]. Treatment with high doses of glucocorticoids (GCs) should be initiated immediately to prevent ischaemic complications, such as visual loss. However, GCs can lead to significant toxicity[2], and other diseases may have a similar presentation to GCA. Therefore, a prompt and accurate diagnosis in suspected GCA is essential for successful management.

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CASE REPORT

We report a case of a 69-year-old woman who presented to the Emergency Department with a new onset of left hemicranial and retro-orbital headache, associated with diplopia and left eye amaurosis. Over the previous two months, she complained of weight loss (8% of total body weight) and PMR features. More recently, she reported pain below the right shoulder blade. She denied fever or jaw claudication. Her background history consisted of diabetes mellitus type 2 treated with insulin, and there was no history of smoking. On physical examination, cardiopulmonary auscultation was unremarkable, and no temporal artery abnormalities were found. Ophthalmic examination of the left eye revealed a lack of light perception, relative afferent pupillary defect, proptosis, horizontal recti muscles palsy, and hypoesthesia in the V1 and V2 territory of the trigeminal nerve. On fundoscopy, diabetic macular oedema was observed, and the optic disc showed no signs of ischaemia. Erythrocyte sediment rate was 50 mm/hr and C-reactive protein (CRP) 5.96 mg/dL. The thorax radiograph revealed a hypotransparent image, with an irregular oval shape, in the right inferior lobe (Figure 1a). Cranial computed tomography (CT) showed ischaemic

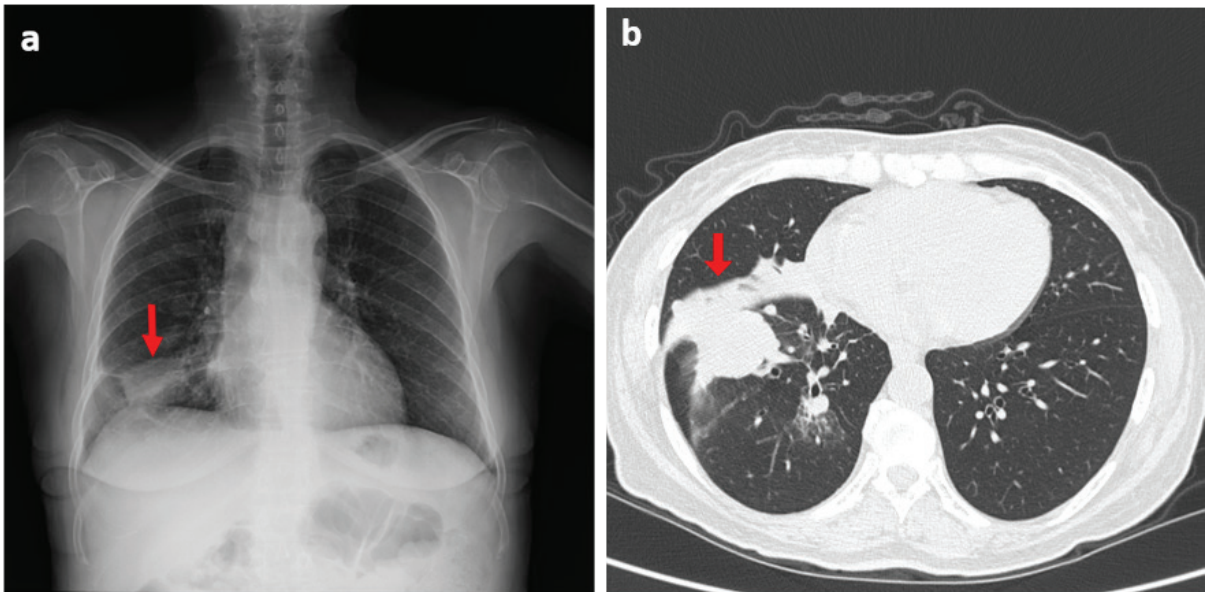

Figure 1. a) Chest posteroanterior radiograph showing a slightly irregular oval hypotransparent image in the inferior right lobe (arrow). b) Chest Computed Tomography showing a lung mass in the right inferior lobe (arrow).

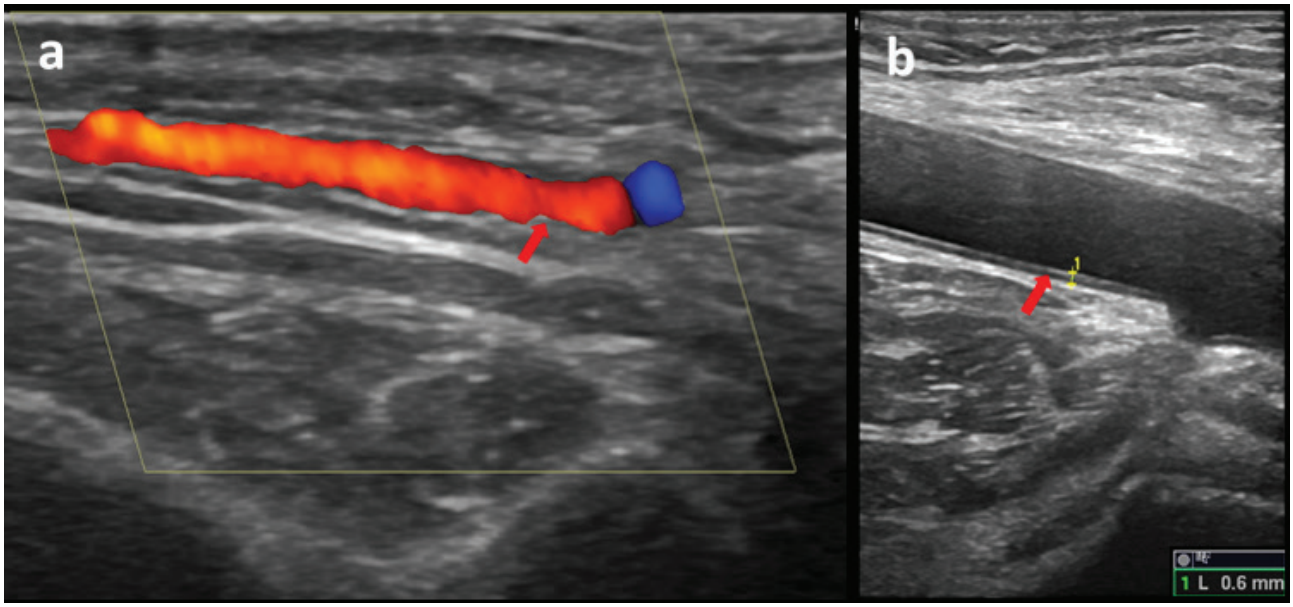

Figure 2. a) Colour Doppler ultrasound of the left common superficial temporal artery, in longitudinal view, showing a normal intima-media complex without the presence of halo sign (arrow). b) Ultrasound of the left axillary artery, in longitudinal view, showing a normal intima-media complex with 0.6 mm of thickness and without the presence of halo sign (arrow).

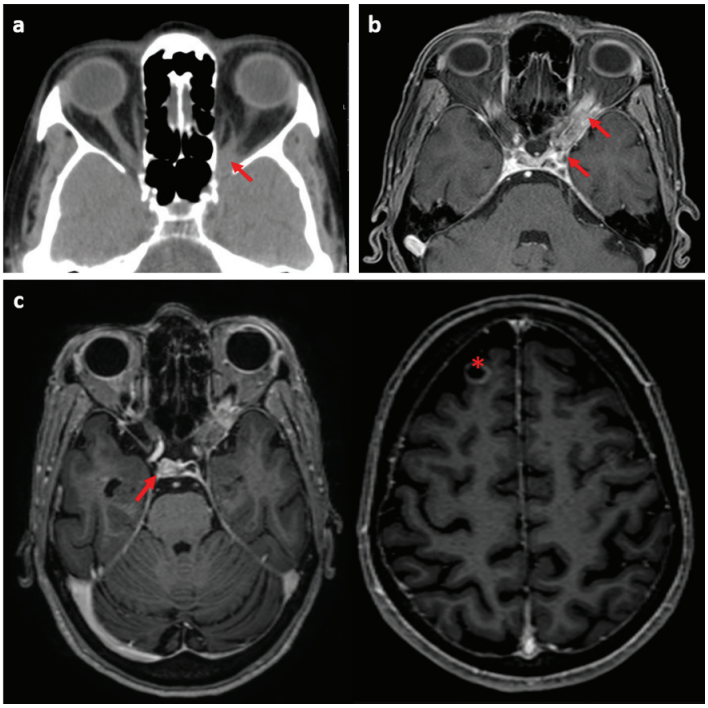

Figure 3. a) Axial CT scan of the orbits – Soft tissue component on the left orbital apex (arrow); there is fat stranding and ill-defined optic nerve and extraoculomotor muscles.
b) Orbits MR – Axial T1 with fat suppression after gadolinium: There is fat obliteration and contrast enhancing soft tissue in the left orbital apex, involving left optic canal and superior orbital fissure (arrow). There is also involvement of the left anterior cavernous sinus, optic nerve in the optic canal and superior rectus muscle, with mild left proptosis. c) Brain MR – Axial T1 post gadolinium – There is an osseous and dural lesion, with contrast enhancement after gadolinium, in the right dorsum sellae (arrow) and a cystic brain lesion in the right frontal convexity, with a ring enhancement pattern (asterisk).

microangiopathic leukoencephalopathy and the cerebrospinal fluid analysis was normal. Due to initial high suspicion of GCA, pulses of methylprednisolone 1g per day were initiated, leading to a reduction of CRP; however, the patient remained symptomatic. On the 3rd day of treatment, an ultrasound of the temporal and axillary arteries showed no signs of vasculitis (Figure 2).

A thoracic CT revealed a lung mass with multiple lung and adrenal nodules (Figure 1b). Histology confirmed lung adenocarcinoma. The patient also underwent cranial MRI showing fat obliteration of the left orbital apex with isointense tissue enhancement after gadolinium suggestive of orbital apex syndrome (Figures 3A, 3B), a rare condition characterized by optic neuropathy and

ophthalmoplegia due to the involvement of structures within or near the orbital apex[3]. A dural and a parenchymal lesion were also detected consistent with metastases (Figure 3c). Therefore, metastatic lung cancer was assumed as the cause of orbital apex syndrome. The patient stopped GCs with a quick tapering scheme. However, two months after diagnosis, she died due to acute cholecystitis complicated by renal failure.

CONCLUSION

Other diseases may mimic GCA. This patient presented an orbital apex syndrome secondary to lung adenocarcinoma manifesting with headache, visual disturbances, PMR, and high inflammatory markers. A fast track approach[4] with immediate ultrasound could have optimized clinical investigation and prevented unnecessary GC treatment.

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