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Eagle's syndrome mimicking thyroid-associated orbitopathy

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Thyroid-associated orbitopathy (TAO), also known as Graves' orbitopathy, is a potentially sight-threatening autoimmune ocular disease. TAO is usually associated with hyperthyroidism. However, TAO can present independently of Graves' disease, endocrinologic manifestations, or elevated thyroid receptor antibodies (TRAbs). In the latter case, TAO requires differentiation from other intra- and extraocular pathologies mimicking its course. This paper presents a patient with Eagle's syndrome manifesting as TAO.

A 70-year-old woman was admitted to the hospital for diagnostics and treatment of bilateral exophthalmos. The patient underwent a thyroidectomy due to nodular goitre 20 years previously and had been treated with levothyroxine since then. Eye symptoms (bilateral exophthalmos [left eye > right eye], conjunctival redness, eyelid oedema, photophobia, lacrimation, retro-orbital pain, and intermittent diplopia) started 5 months before admission. The patient also complained of left-sided facial pain. She had previously been treated with oral methylprednisolone without any improvement. Subsequently, an orbital MR scan revealed symmetrically slightly thickened and swollen extraocular muscles. Despite undetectable TRAbs, the TAO diagnosis was made after consultation with an ophthalmologist, with a Clinical Activity Score (CAS) of 5/7 points. The patient received intravenous pulses of methylprednisolone with a total dose of 1125 mg, with no evidence of any clinical improvement.

Meanwhile, on a head CT scan, significant hypertrophy of a left styloid process was visualized. The patient was diagnosed with Eagle's syndrome and underwent styloidectomy. After the procedure, retrobulbar pain and diplopia subsided.

Eagle's syndrome is a rare and poorly understood condition characterized by an elongated styloid process or a calcified stylohyoid ligament. An elongated process can be found in 4–7.3% of the population, but when the calcified stylohyoid ligament is included, the prevalence increases [1]. It occurs more often in females than in males (2:1 ratio), and the morbidity increases from age 50 years [2]. Eagle's syndrome symptoms are related to the anatomical structures involved and include temporomandibular disorders, ear pathologies, and facial pain. Pain is the most common manifestation, which usually occurs on the side of an elongated styloid process [3]. Sometimes it can also radiate to the ear and jaw, presenting as otalgia or temporomandibular joint pain [3]. Other very common symptoms are dysphagia, foreign body sensation in the throat, and pain when yawning and turning the head. Furthermore, Eagle's syndrome can also be associated with vascular symptoms related to carotid artery compression like transient ischaemic attacks or even ischaemic stroke, and ocular manifestations such as Horner's syndrome, ischaemic neuropathy, or diplopia [4]. Additionally, the elongated styloid process can compress the jugular vein in certain head positions, causing reduced venous outflow with symptoms of swelling of the periorbital tissues, and venous stasis in the conjunctival and retinal vessels mimicking the symptoms of orbitopathy. A few cases of internal jugular vein stenosis (IJVS) associated with the elongated styloid process have been described in the literature [4]. IJVS can present with many non-specific symptoms, including eye symptoms such as diplopia, eye bloating, blurred vision, and visual field defect [4]. According to Li et al., the lack of smooth muscles and elastic fibres



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in veins makes them vulnerable to compression from external structures. It may be one of the most important pathomechanisms of IJVS [4]. It may also link Eagle's syndrome to the TAO-mimicking eye symptoms presented by the patient.

The Eagle's syndrome treatment may be conservative and surgical, depending on the patient's preferences and the severity of symptoms. In conservative management first-line oral analgesics, such as non-steroid anti-inflammatory drugs (NSAIDs), are used, which may be subsequently combined with other medications: anticonvulsants, antidepressants, and local injections of steroids or anaesthetics [5]. However, the literature rather supports surgical treatment because it provides better outcomes in terms of symptomatic relief in patients [3].

Eagle's syndrome is often difficult to diagnose due to the wide array of symptoms. This article presents the first example in the literature of symptoms usually attributed to TAO as a manifestation of Eagle's syndrome. It stresses the need to re-evaluate the cause of eye symptoms in a patient with a history of thyroid disease, particularly if they do not respond to standard treatment.

Conflicting interests

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Author contributions

All authors listed have contributed sufficiently to the project by data acquisition and manuscript writing to be included as authors, and all those who are qualified to be authors are listed in the author byline.

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Data and code availability statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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