Horner's syndrome resulting from a substantial mass lesion: an atypical manifestation of granulomatosis with polyangiitis

Sirs,

Granulomatosis with polyangiitis (GPA) is a rare autoimmune disease. It is a type of small-vessel vasculitis. In this case report, we present a complex and atypical manifestation.

A 50-year-old woman was referred due to recurrent episodes of sinusitis and now additional severe pain from the right side of the face. The pain was located to the nasal cavity, maxillary sinus and ear region. This was accompanied by sensitivity to light, loss of taste and smell, and altered sensibility in the teeth in the upper right side of the mouth. A weight loss of 3–5 kg occurred. Examinations were initiated at the ENT and Rheumatology departments. The ENT-examination revealed a perforation of the nasal septum. A septal biopsy was performed, and CT and MRI scans were ordered. The general clinical examination performed in the rheumatology clinic was normal, except for the observation of mild ptosis and a small pupil on the right eye, *i.e.* Horner's syndrome.

Biopsy showed vasculitis, necrosis and multinucleated giant cells. The MRI scan showed a 4x2, 5x2 cm mass in the right side of the nasopharynx (Fig. 1A). The mass was radiologically described to be extending into the retropharyngeal space and protruding into the carotid canal. Blood tests showed positive PR3-ANCA.

The biopsy result, blood tests and a compatible clinical picture pointed towards GPA as the most likely diagnosis. The mass could be related to this, but a biopsy was taken to ensure that malignancy could be ruled out. The result confirmed GPA. The observed Horner's syndrome was interpreted as possibly secondary to the mass lesion. After Horner's syndrome was discovered, an ophthalmologist was consulted, and the syndrome was verified. To exclude vascular pathology, CT angiography was done. The test was negative.

The patient was treated with prednisolone and cyclophosphamide, which was later converted to methotrexate. Today, disease control is achieved, and control MRI scans showed regression of the mass (Fig. 1B). Horner's syndrome is still present.

GPA is a rare diagnosis. A yearly incidence of 5–10 per million is reported (1). Treatment of the disease is lifesaving. There are no definitive diagnostic criteria (2). This case was complex, comprising the development from cold symptoms and sinusitis to the demonstration of a mass lesion and Horner's syndrome. It is possible that GPA may form space-occupying mass lesions in the form of granulomas (3), as it was seen here. There are examples of patients who have been diagnosed with GPA and a cancer at the same time (4), which underlies the principle that malignancy must always be ruled out whenever the risk of it appears. In this case, the biopsy from the mass confirmed GPA. The explanation for the observed Horner's syndrome is presumably related to the impact of the mass on the surrounding anatomy. From the superior part of the sympathetic trunk, the internal carotid nerve arises to form the sympathetic plexus, which lies around the internal carotid artery and subsequently creates connections to the orbit among other things. As mentioned, the mass was radiologically described to be protruding into the carotid canal. The carotid canal exactly contains the artery and the surrounding plexus, and it therefore seems possible that the impact at this point may have

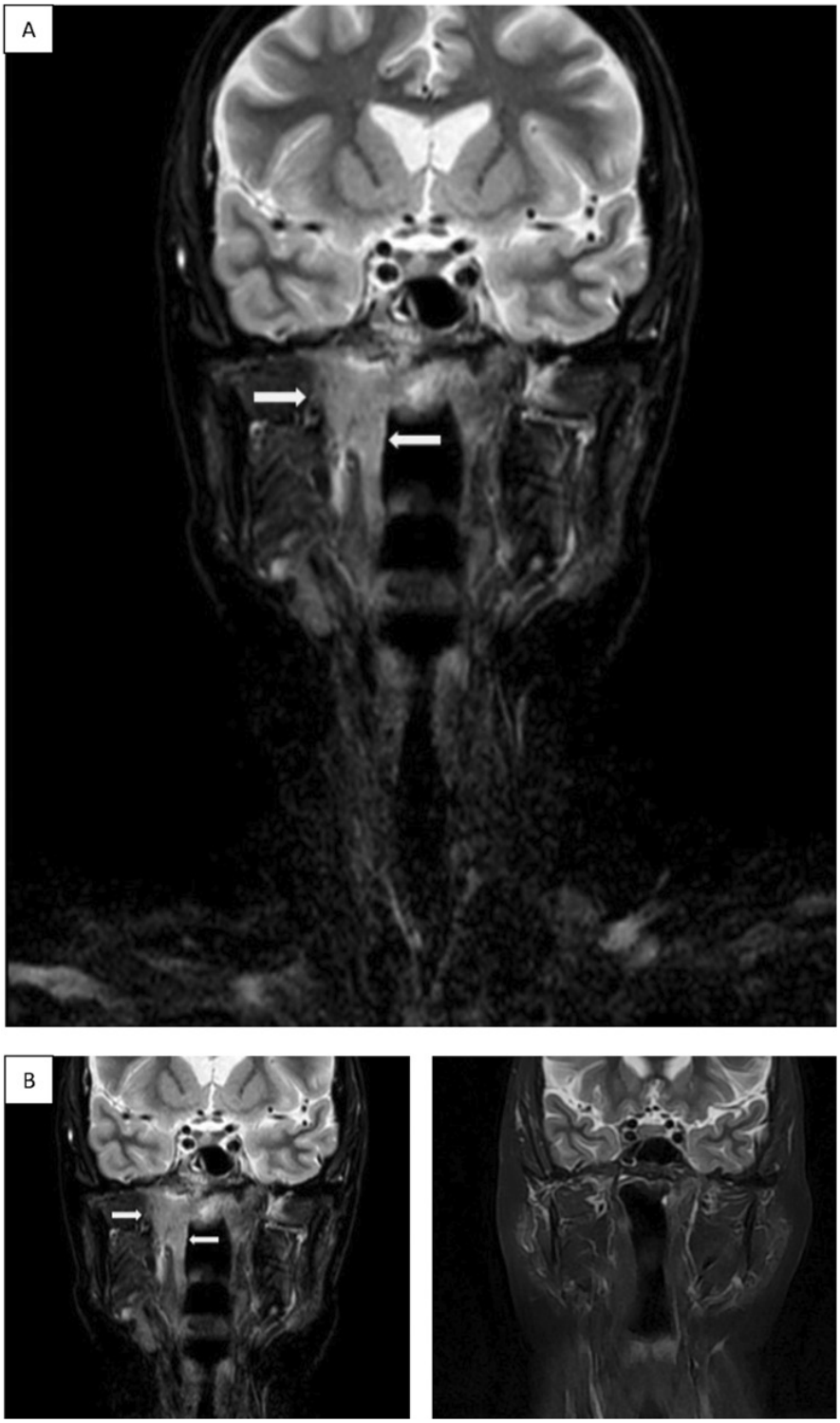

Fig. 1. MRi scans. **(A)** Initial MRi scan. **(B)** Comparison between the initial MRi scan and control MRi after treatment.

compromised the sympathetic innervation to the eye and thus resulted in the syndrome. Horner's syndrome based on GPA has been described as an extremely rare phenomenon (5). In this particular case, the syndrome presumably resulted from the occurrence of a substantial mass lesion.

In conclusion, this highlights the enormous variability that systemic diseases can manifest with, and the considerable differential diagnostic width that is tied to this group of diseases.

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Competing interests: none declared.

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