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Pathology Page

Wegener's granulomatosis that mimics nasopharyngeal cancer

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A 54-year-old female was diagnosed with systemic lupus erythematosus in a hospital elsewhere during 1994. She had been without regular medication control for 9 years when she was first admitted to our hospital after a seizure. She also had progressive general weakness and a poor appetite; she was suffering from body weight loss, a dull sensation with mild chest discomfort, dyspnea on exertion, dizziness, and a facial skin rash after exposure to sunlight. A laboratory examination showed hemoglobin 11.3 g/dL, white blood cell count 5500/mm^3, erythrocyte sedimentation rate 74 mm/h, blood urea nitrogen 14 mg/dL, serum creatinine 0.6 mg/dL, albumin 3.2 g/dL, and total protein 5.1 g/dL. C3 was 111 mg/dL (normal 90–120 mg/dL) and C4 33.2 mg/dL (normal, 20–40 mg/dL). As a result of the seizure, T1-weighted magnetic resonance imaging of the head with contrast enhancement was carried out, which revealed a strong and heterogeneous enhanced soft tissue lesion in the left nasopharynx; it had a perineural spread and extended into the carotid space, oropharynx, and pterygopalatine fossa (Fig. 1A). The woman underwent a tissue biopsy, and the tissue sample was found to have noncaseous granulomatous inflammation (Fig. 1B). Acid fast, Periodic Acid-Schiff (PAS), and Gomori Methenamine Silver (GMS) staining of the tissue sample were all negative. In addition, no abnormal atypical lymphoid cell infiltration was noted .A laboratory study showed the patient to be positive for cytoplasmic antinuclear antibodies (titer 1/1280). As a result of the elevated antineutrophil cytoplasmic antibody (c-ANCA) level and the histopathological findings, Wegener's granulomatosis (WG) was diagnosed. The woman was regularly followed up in the Rheumatologic Department and was treated with prednisolone. Follow-up brain magnetic resonance imaging showed that the abnormal mass had disappeared. Follow-up laboratory testing found that the c-ANCA titer had reduced and was < 1/20 at 4 years after the initial diagnosis. At a later time, the patient died of septic shock associated with multiple organ failure.

WG is a rare disease of uncertain cause. It is characterized by granulomatous inflammation with geographic basophilic palisading necrosis and can affect a variety of tissues, including blood vessels. WG primarily affects the upper respiratory tract, lungs, and kidneys. Recently, a new classification of WG has been proposed, and this adds a third form to the two forms described earlier. The three forms are as follows: (1) the classical generalized systemic or diffuse form that always involves the kidneys and causes necrotizing glomerulonephritis; (2) the localized or limited form that does not involve the upper respiratory tract or the kidneys; and (3) the third newly proposed form that is purely granulomatous (without evidence of vasculitis). Our case fits the third form with the WG masses seen on the magnetic resonance images being isointense to slightly hypointense relative to muscle on T1-weighted images, while contrast enhancement was able to reveal a strongly enhanced soft tissue mass.

The c-ANCAs are autoantibodies directed against a cytoplasmic antigen that is expressed by human neutrophils and monocytes. The target antigen of these antibodies is proteinase 3. A high level of c-ANCAs has both high specificity and sensitivity with respect to WG, and the c-ANCA level has been found to parallel disease activity, being present in the serum of 95% of

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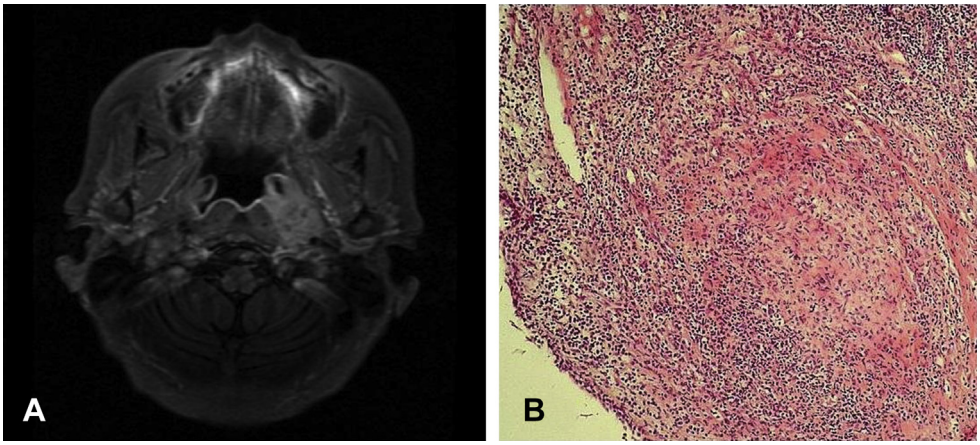

Fig. 1. (A). MRI T1WI with contrast enhancement. The axial section reveals a strong and heterogeneous enhanced soft tissue lesion within the left nasopharynx. (B) Histopathology shows granulomatous inflammation without vasculitis (hematoxylin and eosin stain, 100×). MRI = magnetic resonance imaging; T1WI = T1-weighted image.

patients with the active generalized form of the disease. As far as we know, in Taiwan, our case is the first case of WG that mimics a nasopharyngeal tumor.

Further reading

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