



Wegener's granulomatosis of the paranasal sinuses with orbital and central nervous system involvement-diagnostic imaging

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Abstract

Wegener's granulomatosis (WG) is a systemic vasculitis that can affect any organic system, but primarily involves the upper and lower respiratory tracts and the kidneys. WG relatively frequently affects the nervous system (in 30-50%), usually in the form of peripheral or cranial neuropathy. Involvement of the brain is reported in a very small percentage of patients (2%-8%). Three major mechanisms have been described as the cause of central nervous system (CNS) disease in WG: contiguous invasion of granuloma from extracranial sites, remote intracranial granuloma and CNS vasculitis. CNS involvement caused by contiguous invasion of granuloma from extracranial sites is the rarest. We report the case of a 37-year-old man with WG, manifested as a pulmonary and paranasal sinuses disease, with orbital and CNS involvement, caused by contiguous invasion from the paranasal sinuses. In this report, the rich spectrum of findings achieved by computed tomography and magnetic resonance are demonstrated. The importance of computed tomography in bony destruction PNS findings, and the importance of MR imaging in evaluation of the direct intracranial spread from nasal, paranasal and orbital disease are also emphasized.

Introduction

Wegener's granulomatosis (WG) is a necrotizing granulomatous vasculitis of small and medium sized vessels and usually affects the upper and lower respiratory tract and kidneys (1, 2, 3). Upper respiratory tract features are typical at presentation, such as sinusitis and nasal obstruction. The orbits, heart, skin, and joints are frequently involved (4). Intraorbital WG involvement is usually accompanied by paranasal sinus disease (5). Cerebral and meningeal involvement is uncommon, occurring in only 2%-8% of patients (2, 4, 6).

We report a man with WG presented by paranasal disease, orbital involvement and direct intracranial

propagation, which is the rarest mechanism that causes CNS disease in WG.

Case report

A 37-year-old man came to the Department of Radiology with a four year history of haemoptysis and three month history of headache, diplopia, left ptosis, blocked left nostril with occasional elimination of yellow-green and haemorrhagic content in the form of drops from the nasal cavity, and computed tomography (CT) of the head and orbits was performed. He was treated twice for skin lesions, with the diagnosis Cellulitis et furunculosis and Suppurative Panniculitis, with no histopathological signs of granulomatous disease.

Unenhanced CT scans of the head revealed extensive bony destruction of the nasal septum, all of the turbinates, PNS walls, except the frontal sinus, right orbital roof, anterior and central skull base and mucosal thickening in the maxillary sinuses. The sphenoid sinus, the residual part of the cranial ethmoid complex and the retrobulbar part of the left orbit were almost totally filled with inflammatory mass (Fig. 1). The radiologist suspected PNS Wegener's granulomatosis with orbital and possible brain involvement. Chest radiography and CT scans of the thorax showed a left-sided cavitating lesion, 95 × 65 × 85 mm, with an air-fluid level in it, suggesting pulmonary WG.

The patient was admitted to our hospital. Labs on admission showed mild leukocytosis, mild anaemia and high values of C-reactive protein. The patient's neurological status suggested lesions of the left III, IV, V and VI cranial nerves.

Magnetic resonance (MR) of the head showed irregular infiltrative lesions with low signal intensity on T1-weighted images, and intermediate to high

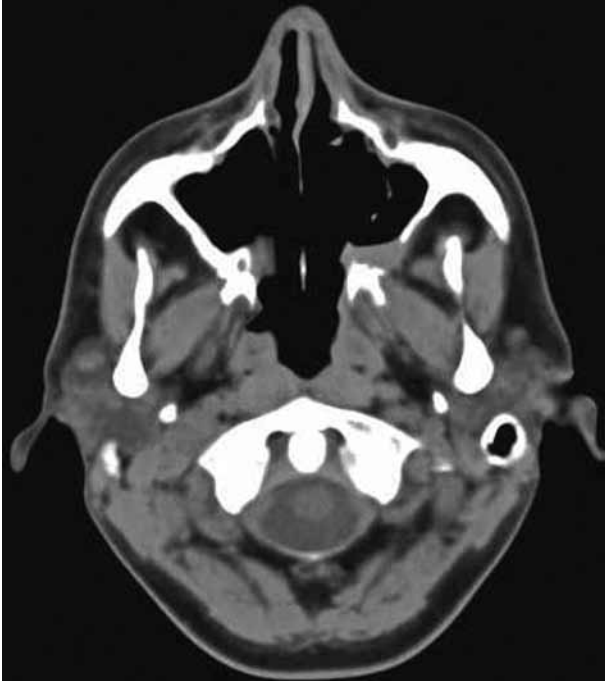


FIG. 1. — An axial CT image of the sinuses shows bony destruction of the nasal septum, turbinates and medial wall of the maxillary sinuses.

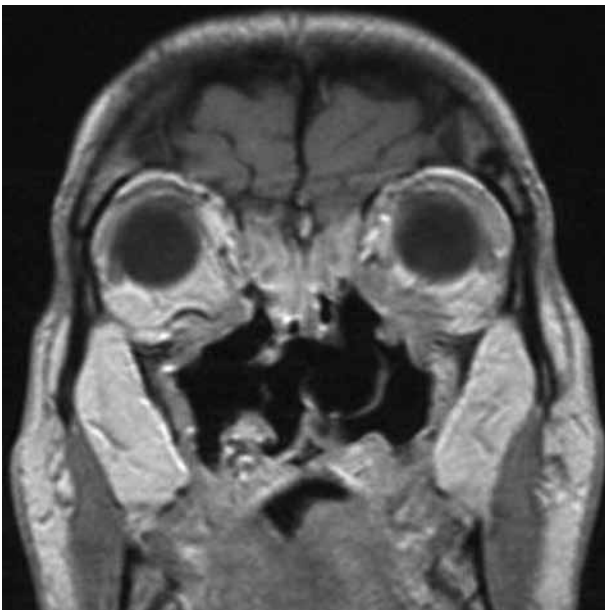


FIG. 2. — A coronal T1-weighted contrast-enhanced MR image shows destruction of the nasal septum and turbinates, with strongly mucosal contrast enhancement in the maxillary sinuses.

signals on T2-weighted images, with strongly mucosal contrast enhancement in the nasal cavity and maxillary sinuses (Fig. 2). A soft-tissue mass almost totally filled the sphenoid and ethmoid sinus. The medial orbital wall of the left orbit had been

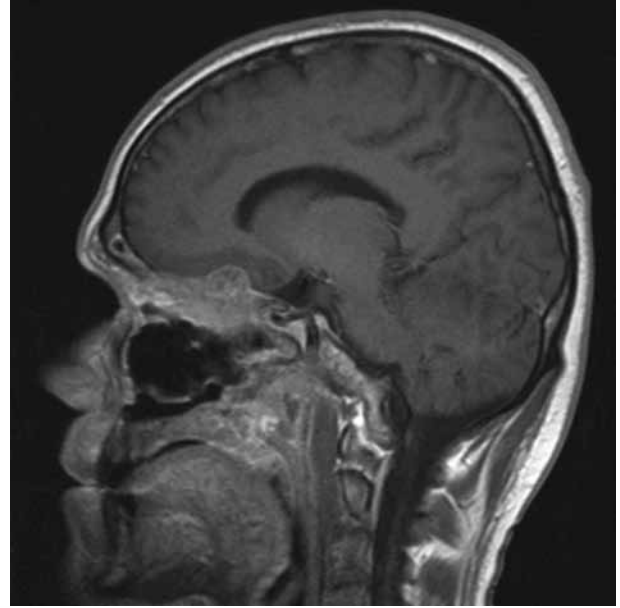


FIG. 3. — A sagittal T1-weighted contrast-enhanced MR image shows the intracranial spread of mass from the ethmoid sinus through the destroyed cribriform plate into the left frontal lobe.

destroyed by the soft-tissue mass which occupied the intra- and extraconal orbital spaces with initial propagation of the mass into the extraconal space of the right orbit.

MR scans also showed intracranial spread of the mass from the ethmoid sinus through the destroyed cribriform plate in the left frontal lobe as well as thickening and enhancement of the dura in the left temporal and both frontal lobes (Fig. 3, 4). MR exam corresponded to WG of the PNS, with orbital and brain involvement. The histopathological examination of the skin lesion from the forearm showed chronic pustular and granulomatous dermatitis, with minimal vascular wall damage (Fig. 5). Serum anti-neutrophil cytoplasmic antibodies were positive, according to the enzyme-linked immunosorbent assay, which confirmed WG.

After corticosteroids and cytotoxic agent treatment, six months later, control MR scans revealed complete regression of the inflammatory mass in the left frontal lobe, with persisted enhancement of the dura of the anterior skull base. Inflammatory mass in the left orbit, maxillary and ethmoid sinuses were in good, incomplete regression.

Discussion

WG usually affects the upper and lower respiratory tracts. The most common imaging findings of

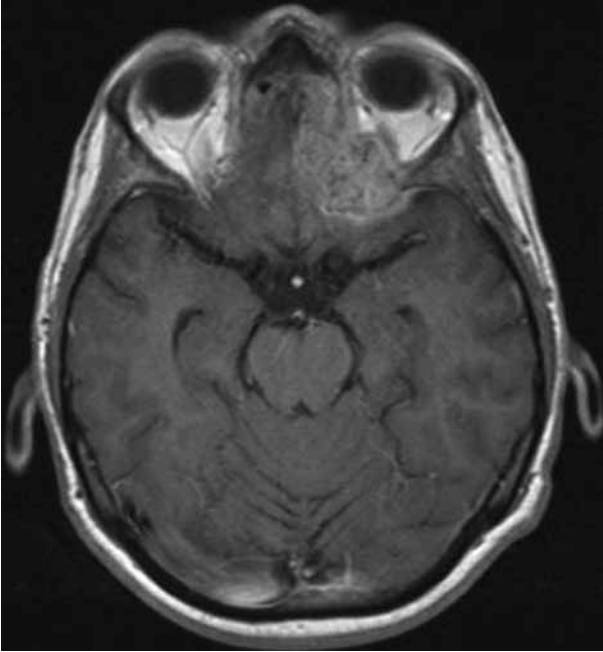


FIG. 4. — An axial T1-weighted contrast-enhanced MR image shows the inflammatory mass in the straight and orbital gyrus of the frontal lobe and in the left orbit.

the lung are discrete focal opacities that vary in size and appearance from diffuse consolidation to nodular masses (3).

Symptoms in the head and neck region are observed in up to 95% of patients with WG (1). A limited form of the WG may occur, with a more benign course, in which only the sinonasal tract is affected. The initial disease may present as a chronic, nonspecific inflammatory process of the nose and sinuses. Usually the nasal septum is first affected. The process becomes diffuse, and septal ulceration and perforations may result in a “saddle nose” deformity. Secondly, bacterial infections complicate the clinical and imaging pictures (7).

While mucosal abnormalities of the nose and PNS have been well characterized and the range from granulomatous lesions to diffuse mucosal thickening, abnormalities of the underlying bones of the PNS in this disease have not been well described (8, 9). Lohrmann *et al.* described pathologies of sinonasal CT in 28 patients with WG and the following CT findings were observed: (a) bony destruction, (b) sclerosing osteitis, (c) bony thickening and (d) mucosal thickening (10). In our case, bony structure destruction of the nasal cavity and sinuses are dominant, with diffuse mucosal thickening of the maxillary sinuses, and with an inflammatory mass in the sphenoid and ethmoid sinuses.

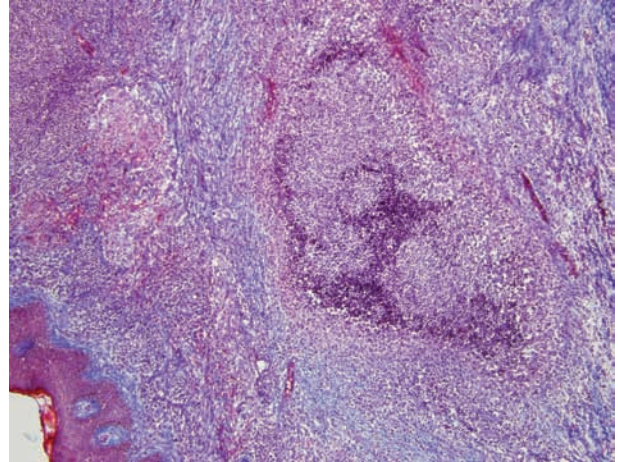


FIG. 5. — Histopathological image. Infiltration of polymorphonuclear cells in dermis and small epithelioid granuloma (Masson's Trichrome stain).

CNS involvement is an uncommon manifestation of WG, reported in 2%-8% of patients (4, 6, 11). Three major mechanisms have been described as the cause of CNS disease in WG: contiguous invasion of granuloma from extracranial sites, remote intracranial granuloma, and CNS vasculitis (11). Provensale and Allen reviewed CT or MR studies of 15 patients with WG (12). Abnormal CNS findings were reported in seven patients, but none of them had extension from sites external to the CNS extension as was revealed in our patient.

Intraorbital WG involvement is ultimately seen in 50% cases of generalized WG (4). We found left orbital involvement with combined intra- and extraconal location and initial right orbital involvement.

Conclusion

Direct intracranial propagation WG from the paranasal sinuses is the rarest mechanism described as the cause of CNS disease in WG. MR imaging is the most important diagnostic imaging for evaluation of direct intracranial spread from nasal, paranasal and orbital disease, while CT imaging has a very important role in findings of bony destruction in PNS.

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