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CASE BASED REVIEW

Cranial neuropathies in granulomatosis with polyangiitis (Wegener's): a case-based review

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Abstract The purpose of this case-based review is to highlight cranial nerve involvement in granulomatosis with polyangiitis (Wegener's). In this disease, cranial nerve involvement may be less frequent than other neurological manifestations, but often goes unrecognized by physicians as a sign of the disease, and its prevalence and importance is likely underestimated. Awareness of this aspect of the disease is necessary to make the proper diagnosis rapidly, as it can be a major feature of a patient's presentation. We also briefly discuss the known pathogenic mechanisms, which could be important when selecting the best therapeutic option.

Keywords ANCA-associated vasculitis · Cranial neuropathy · Cyclophosphamide · Granulomatosis with polyangiitis (Wegener's) · Neurological manifestations · Pachymeningitis · Rituximab

Introduction

Classical granulomatosis with polyangiitis (GPA; Wegener's) is an inflammatory necrotizing granulomatous vasculitis that typically involves the upper respiratory tract, lungs, and kidneys. GPA can also present with less typical symptoms and various clinical manifestations, mimicking many other disorders [1, 2]. While neurological involvement is common, occurring in between one-quarter to half of all cases, cranial nerve involvement appears less frequent, described in less than 10 % of cases [3].

Here, we review cranial nerve involvement in GPA in light of the case of a woman suffering from rapidly progressive GPA, in which cranial nerve involvement was the prominent feature, along with dysarthria, dysphagia, and tinnitus. We also briefly discuss pathogenic mechanisms, whose consideration could be important when selecting the best therapeutic option.

Methods

We describe in detail a case of GPA that was complicated by possible comorbidities and incidental discoveries, with particular attention to the cranial neurological manifestations. We conducted a literature search using PubMed for English, French, German, and Spanish language articles published between 1965 and April 2013 using the keywords "Wegener," "neurologic manifestations," "cranial neuropathy," and "granulomatosis polyangiitis." Bibliographies of articles relevant to neurological manifestations were reviewed for additional articles, and all articles pertaining to cranial manifestations of GPA were retained.

Case report

A 73-year-old diabetic woman was initially diagnosed with seropositive rheumatoid arthritis (RA) by her general practitioner. She presented with symmetrical synovitis involving the proximal interphalangeal and metacarpal phalangeal joints, wrists, elbows, and shoulders, with an elevated sedimentation rate (81 mm/h) and high C-reactive protein (CRP) (127 mg/l), and strongly positive for rheumatoid factor and anti-cyclic citrullinated peptide (CCP) antibodies. Low-dose corticoids

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(≤10 mg/day) were given orally with a good initial clinical response.

However, the patient rapidly developed intermittent claudication of the lower limbs. Although she had a history of diabetes, hypertension, and a femoropopliteal bypass 3 years prior for arteriopathy in the lower limbs, after angiological investigation, a vascular etiology was considered unlikely by the general practitioner. Likewise, no evidence of spinal stenosis was found clinically or by lumbar spine magnetic resonance imaging (MRI). In the presence of a persistent inflammatory syndrome and claudication, with pain in her calves and buttocks after walking 200 m, as well as paraesthesia of both foot plants at rest, she was admitted to our rheumatology ward for investigation.

At admission, she further complained of asthenia, night sweats, and a 5-kg weight loss over the previous 6 months, but denied any fever. She mentioned recent nocturnal left frontotemporal headaches, associated with some tinnitus and hearing loss on the same side. On admission, general and neurological examinations were normal for her age, with the exception of an aortic heart murmur, reduced pallesthesia in the ankles, and persistent synovitis of the proximal interphalangeal and metacarpal phalangeal joints. ENT examination revealed erosive nasal crusting and sensorineural hearing loss on the left side.

Her erythrocyte sedimentation rate was still elevated at 83 mm/h and CRP at 116 mg/l. She was anemic with a hemoglobin level of 103 g/l, but leucocyte and platelet counts were normal, as were renal (creatinine at 37 μmol/l) and thyroid functions and urine sediment. Elevated titers were confirmed for both rheumatoid factor (74 U/l, $N<20$) and anti-CCP antibodies (160 U/l, $N<20$).

Chest X-rays revealed multiple pulmonary opacities, some of which were excavated. A thoracoabdominal CT scan confirmed the presence of multiple excavated pulmonary nodules, up to 10 cm in diameter, with no mediastinal adenopathies (Fig. 1a). To complicate the case further, the scan also revealed bilateral heterogeneous renal masses (up to 8 cm on the right side; Fig. 1b) as well as bilateral adrenal nodules 2–3 cm in diameter. However, there was no observed liver or spleen enlargement, no other abnormal masses, nor bowel or retroperitoneal involvement.

Based on these findings, there were a large number of potential diagnoses. A metastatic neoplasia was of course suspected. Because of a past history of left *Aspergillus fumigatus* sphenoiditis with osteitis and secondary optic neuropathy in a patient on oral corticoid therapy, she was initially treated intravenously by voriconazol for potential invasive disseminated aspergillosis. Further microbial analysis of the bronchoalveolar lavage and blood analysis allowed us to exclude this diagnosis, as well as other fungal, bacterial or tuberculous infections. Rheumatic pulmonary nodules appeared very unlikely. In contrast, suspected GPA was quickly

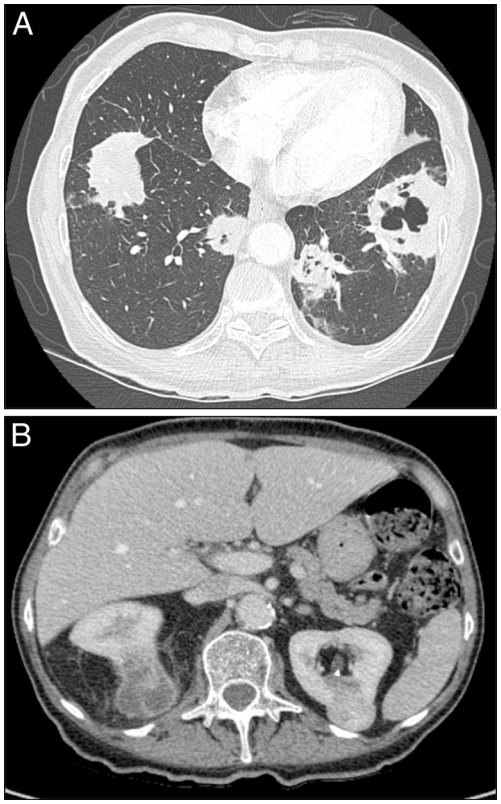

Fig. 1 Thoracoabdominal CT scan showing numerous bilateral excavated peripheral pulmonary masses (**a**), bilateral heterogeneous renal masses (**b**), and bilateral adrenal masses (not shown)

confirmed by elevated anti-PR3-specific anti-neutrophil cytoplasmic antibodies (cANCA) (1/160) and histological evidence of a necrotizing granulomatous vasculitis on the pulmonary transthoracic biopsy. Cerebral CT scan showed pansinusitis without any cerebral abnormalities, in particular, no involvement of the mastoids or auditory canal. A mononeuropathy multiplex of the lower limbs was confirmed by electroneuromyography. Further workup failed to show any kidney involvement with a normal 24-h urine analysis and sediment, while the biopsies performed on both renal and adrenal masses indicated bilateral renal oncocytomas, a benign renal tumor, and benign nodular adrenal hyperplasia—incidental discoveries and a good example of Saint's Triad in an older woman [4].

While hospitalized, the patient developed sudden dysarthria with paresis of the left hypoglossal nerve. An angio-MRI allowed us to exclude acute bulbar ischemia but revealed an infiltrative left retropharyngeal mass encasing the internal carotid artery and hypoglossal nerve (Fig. 2). No involvement of the mastoids or auditory canals was observed. Concomitantly, her frontotemporal headaches were worsening, were unresponsive to opioids, and only weakly responded to NSAIDs. A normal lumbar puncture at this time ruled out any significant meningeal involvement. She was started on high doses of corticosteroids (1 mg/kg) with pulse intravenous

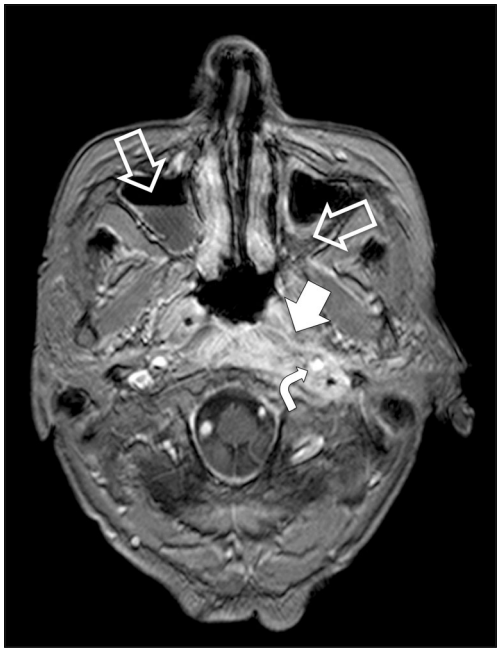

Fig. 2 MRI (T1 gadolinium-enhanced sequences) revealing a diffuse infiltrative lesion of the left retropharynx (*plain arrow*) encasing the internal carotid artery (*curved arrow*). Bilateral maxillary sinusitis are indicated by the *open arrows*

cyclophosphamide, a treatment that rapidly improved her dysarthria, headaches, left otalgia, and tinnitus, as well as her general condition. The inflammatory syndrome began to improve moderately, with an erythrocyte sedimentation rate (ESR) of 66 mm/h and CRP of 62 mg/l, and the cANCA became negative. She was discharged on tapering orally administered corticoids and intravenously administered cyclophosphamide every 3 weeks.

However, she was readmitted 3 weeks later for worsening dysarthria, sudden dysphagia to solids, recurrent aspiration of liquids, and again severe nocturnal frontotemporal headaches, but on the right side this time. Physical and laryngoscopy examinations indicated bilateral paresis of nerve XII, as well as sensory loss in nerve IX and X territories. A new angio-MRI was performed, which revealed a significant decrease in the size of the known infiltrative retropharyngeal mass and no clear explanation for this worsening neurological presentation, in particular no sign of ischemic or vascular involvement. In addition, there were no signs on MRI of meningeal involvement, but a new lumbar puncture showed mild hyperproteinorachia at 569 mg/l (normal range 150–460), but no pleocytosis, findings compatible with some meningeal involvement. Without any other overt manifestations, but with worsening major cranial involvement, no clear improvement of the inflammatory syndrome (ESR 63 mm/h and CRP 47 mg/l), and in the absence of any other potential etiology but GPA, she was switched to daily orally administered cyclophosphamide and high-dose intravenously administered corticosteroid (three pulses plus 1 mg/kg). The paresis of nerve XII slowly improved under this regimen and logopedic therapy. She was discharged with a persistent inflammatory syndrome (ESR 66 mm/h and CRP 167 mg/l) and cranial neuropathies, but with all other disease manifestations well-controlled.

Despite ongoing immunosuppressive treatment with high-dose corticosteroids (1 mg/kg) and orally given cyclophosphamide (2 mg/kg) and in the absence of any other symptoms, we could not control the inflammatory syndrome (ESR 79 mm/h and CRP 93 mg/l) nor the cranial manifestations; she experienced recurrence of the right temporal headaches, worsening hearing loss on both sides, and a new right facial paresis. In the absence of any clear response and with progressing cranial neuropathies, without any room to increase the cyclophosphamide (leucopenia), she was treated with a first course of 2×1 g of rituximab at a 15 day interval and corticosteroids, which resulted in a very slow improvement of her inflammatory syndrome and cranial neuropathy over the subsequent few months. The cranial nerves remained the major concern and handicap throughout the course of the disease.

Discussion

The emphasis put on the classical triad in GPA [1, 5] has perhaps minimized the importance of the involvement of other organs, in particular neurological involvement [3]. Several studies have demonstrated that neurological involvement is common, with a prevalence ranging from 22 to 34 % [5–8], or even up to 50 % of cases [9, 10].

Peripheral neuropathy is the most common of these neurological manifestations, occurring in 11 to 44 % of cases [6–11]. The classical mononeuropathy multiplex represents 8 to 20 % of cases; distal symmetrical polyneuropathy is as frequent, occurring in 2 to 24 % of cases, while unclassified peripheral neuropathies represent the remaining cases. Central nervous manifestations are much less frequent, with 4 % of cases involving cerebrovascular events (intracranial and subarachnoid hemorrhages, subdural hematoma, venous thrombosis, and stroke syndrome), 3 % involving seizures, and some cases of encephalopathy [8].

Granulomatous involvement of the meninges (pachymeningitis) is another very rarely reported manifestation of GPA. There are 48 cases reported in the literature, often manifesting with headaches (72 %), encephalopathy (10 %), or seizures (15 %) [12]. Headaches are typically severe, and do not resolve with analgesics, but do respond to corticosteroid administration [3, 13]. MRI with gadolinium enhancement demonstrates diffuse or focal thickening of the dura in three-quarters of cases and involvement of the leptomeninges in the other quarter [12]. Seventy percent of patients with pachymeningitis also have abnormal CSF at lumbar puncture, with pleocytosis, proteinorachia, or elevated pressure [12].

Cranial neuropathies, as seen in the present case, are another neurological manifestation of GPA. They appear to be uncommon, with an estimated prevalence between 2 and 10 %, depending on the study [7–10]. Inclusion of oculomotor or optic neuropathies due to granulomatous infiltration and pseudotumor of the orbit in some older studies may explain some of the higher prevalences reported, for example, in one study where external ophthalmoplegia was present in 5 % of patients [8].

Apart from these cases, other cranial neuropathies seldom appear, and as in our case, their true pathogenesis is often unclear [13], but is likely related to either a local granulomatous or vasculitic process. Granulomatous pachymeningitis can be complicated by cranial neuropathies in 32 % of cases [12, 14], usually with multiple nerve involvement, in particular in cases where the base of the skull is involved [15–17]. However, pachymeningitis alone is rare and does not explain all cases either, and certainly not ours. Isolated or multiple cranial neuropathies without pachymeningitis have been documented, but are still rarely reported (Table 1). The pathogenic mechanisms of these cranial neuropathies in the absence of pachymeningitis remain unclear, and they are usually attributed to a vasculitic process.

Both types of processes were probably at play in our patient's symptoms. Our patient initially presented with dysarthria caused by unilateral hypoglossal (XII) paresis, soon followed by dysarthria and dysphagia associated with bilateral XII paresis, and a loss of sensitivity of cranial nerves IX and X. The MRI demonstrated an infiltrative retropharyngeal mass for which no biopsy was performed. However, similar clinical and radiological findings causing secondary cranial neuropathies (V, VII, IX, X, XI, and XII) have been previously reported in the literature [16, 20, 25], and granulomatous infiltration of the retropharynx is probable. Nevertheless, while this infiltrating mass of the retropharynx could explain the paresis of nerve XII on the left side and potentially the headaches, it could certainly not be responsible for the right side's symptoms, especially given that the second MRI showed a reduction in the size of the mass. Likewise, the retropharyngeal mass could not explain the tinnitus and hearing loss secondary to cranial nerve VIII involvement. Cases of nerve VIII involvement related to otomastoiditis have been described [21, 27]; however, this abnormality was not seen on cerebral CT nor MRI in our case. Pachymeningitis cannot be definitively ruled out in our patient, but two MRIs failed to show any thickening or enhancement of the meninges on T2 sequences or after gadolinium enhancement. As has been suggested for other cases [13], ischemic injury due to an occult vasculitic process might be one of the pathogenic mechanisms.

Persistent inflammation and the better response to rituximab than cyclophosphamide in our case could be interpreted as an indirect sign of a vasculitic process [28], as rituximab appears to be preferentially effective for vasculitic manifestations of the disease. However, a major ongoing vasculitic process that would explain the persistence and progression of the isolated cranial nerve involvement is inconsistent with the clear improvement of the peripheral neuropathy and all other manifestations. The better response to rituximab than overt immunosuppression could also suggest an antibody-induced mechanism in a patient with multiple autoantibodies. We did not specifically test for the various paraneoplastic-associated autoantibodies, but the type of neurological involvement presented by our patient would be atypical for such a pathology [29, 30], and very unlikely with a neoplastic-free follow-up of almost 5 years. However, we can certainly not exclude another autoantibody-driven manifestation in a patient who seems prone to develop autoantibodies, with not only high titers of cANCA but also anti-CCP antibodies and rheumatoid factors. These antibodies could suggest the co-occurrence of both GPA and RA, which is rare, as only ten patients have been reported to date [31–34]. Yet, again, such neurological manifestations would not be typical of RA. In GPA, although arthralgias are more common than frank

Table 1 Reported cases of specific cranial nerve involvement in Wegener's granulomatosis not associated with pachymeningitis

Cranial nerve	Total reported cases[a]	Number of patients [ref]
I	1	1 [6]
II	32	10 [8], 1 [7], 10 [6], 7 [10], 4[b] [9]
III	14	2 [8], 9 [6], 2 [9], 1[b] [18]
IV	11	2 [8], 7[b] [6]
V	18	1 [16], 1 [19], 1 [20], 4 [8], 2 [7], 7 [6], 1 [9], 1 [18]
VI	12	8 [8], 8[b] [6], 2 [13]
VII	32	1 [16], 7 [8], 6 [7], 8 [6], 4 [10], 1 [9], 1 [18], 3 [21], 1 [22]
VIII	25	Present case, 2 [8], 1 [7], 8 [6], 2 [18], 7 [21], 3 [23], 1 [22]
IX	8	Present case, 1 [16], 1 [19], 1 [20], 1 [7], 3 [6]
X	9	Present case, 1 [16], 1 [19], 1 [24], 1 [20], 3 [6], 1 [13]
XI	7	1 [19], 1 [24], 1 [20], 3 [6], 1 [13]
XII	9	Present case, 1 [25], 1 [8], 1 [26], 1 [19], 1 [20], 1 [7], 1 [6], 1 [13]

[a] Cranial nerve involvement can be isolated or multiple in a single patient

[b] Secondary to cranial neuropathy and/or granulomatous infiltration of the orbit (pseudotumor), unspecified in older reviews

arthritis [1, 5, 9], it is common to have a presentation of symmetrical polyarthritis of the small and large joints and a false-positive test result for rheumatoid factor, leading to the incorrect diagnosis of RA [5].

Our case serves as a reminder that while the GPA triad typically involves the upper respiratory tract, lungs, and kidneys, it can also affect the nervous system. In particular, there can be severe and debilitating cranial nerve involvement, the prevalence of which is likely underestimated. While the pathogenesis can be explained by either the direct effect of local granulomatous lesions, pachymeningitis, or vasculitic processes, it often remains difficult to determine the etiology, as in the present case. However, identifying the etiology remains essential, as it can guide therapeutic choices, as in our case, where a lasting response was only obtained after the introduction of rituximab. Finally, cranial nerve involvement certainly deserves closer attention, as it can be an initial and unique mode of presentation [14, 21–23, 27, 35, 36] that can be very debilitating.

Disclosures None.

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