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Clinical Case Reports

ENT Manifestations Revealing Wegener's Granulomatosis: Case Report

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Abstract:

Granulomatosis with polyangiitis, formerly known as Wegener granulomatosis, is a rare multisystem autoimmune disease of unknown etiology. It is one of the antineutrophil cytoplasmic antibody (ANCA) associated vasculitic disorders.

A 34-year-old male consulted in ENT department for bilateral purulent rhinorrhea, right hemicrania and dysphagia without fiver. He presented nasal saddle, complete destruction of nasal septum with otological and laryngological signs. These characteristic lesions pushed us to look for other systemic signs. We identify cranial nerve palsies and pulmonary damage. Nasal cavity biopsy with immunohistochemical study objectived nonspecific granulomatous inflammatory process. Tests for ANCA was positive. Typical ENT and pulmonary manifestations, histological results as well as a positive ANCA test allowed the diagnosis of Granulomatosis with polyangiitis. Treatment included mostly corticosteroid therapy (prednisone) with biotherapy (rituximab) and evolution was stationary without the occurrence of other complications.

In this manuscript, we try to describe this case in order to highlight the frequent ENT involvement in granulomatosis with polyangiitis and remind that it can reveal this disease.

Keywords : Granulomatosis with polyangiitis, ENT involvement, antineutrophil cytoplasmic antibody (ANCA)

Introduction:

Wegener's granulomatosis or Granulomatosis with polyangiitis (GPA) is a vasculitis which affects small and medium blood vessels. It is characterized by a pneumorenal syndrome often associated with otorhinolaryngological signs. Various diagnostic criteria have been proposed to diagnose GPA and distinguish the disease from other vasculitides including clinical, biological and histological signs. We report the case of a young man presenting GPA without renal involvement contrasting with rich otorhinolaryngological symptoms, and emphasizing that they can often reveal this disease.

Case report:

A 34-year-old male consulting in ENT department for bilateral purulent rhinorrhea, right hemicrania and dysphonia without fiver. Onset dates back to 6 months by purulent then crusty bilateral rhinitis

which was subsequently accompanied by hemicrania, feeling of fullness of right ear with ipsilateral hypoacusis. Finally, dysphonia, dysphagia and a chronic wet cough completed the symptomatology.

At admission, the patient presented nasal saddle (fig 1), right crude facial paralysis, complete destruction of nasal septum, middle and lower turbinates, with right secretory otitis media. The right tympanogram was flat with negative acoustic reflex associated with moderate conductive hearing loss. Clinical observation revealed also presence of the right side: Fixed hemilarynx (lesion of X), curtain sign (lesion of IX), ageusia of anterior 2/3 of the tongue (lesion of VII), drop of shoulder (lesion of XI) and tongue deviation (lesion of XII) (fig 2).

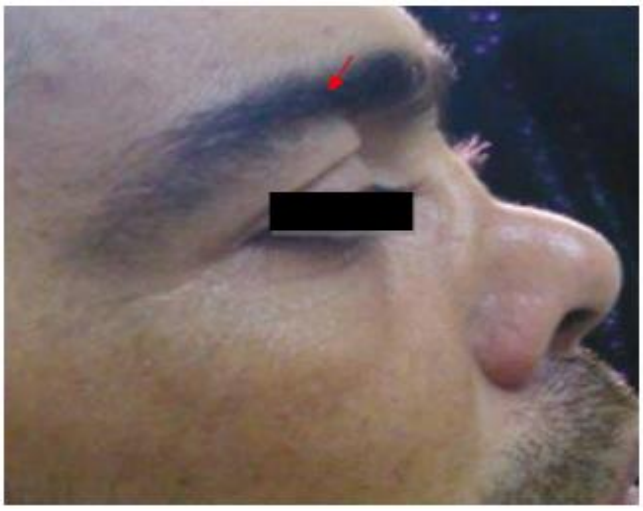

Figure 1: Typical nasal saddle of GPA (red arrow).

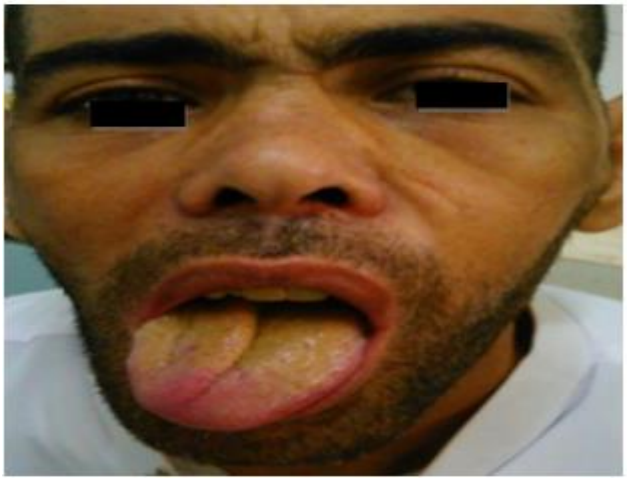

Figure 2 : Right Tongue deviation (XII cranial nerve's palsies)

Three categories of possible etiologies were considered: ENT infectious, neoplastic, and immune-mediated but the presence of nasal saddle was very suggestive of GPA.

Computed tomography (CT) of paranasal sinuses and skull showed a lysis of nasal septum and the intersinusonasal walls (fig 3), mucosa's inflammatory thickening of all paranasal sinuses (fig 4) with edema of right petrous bone and skull base's (fig 5).

RMI of paranasal sinuses and skull showed lysis of nasal septum and intersinusonasal walls with nasopharyngeal and auditory tube inflammatory edema (fig 6).

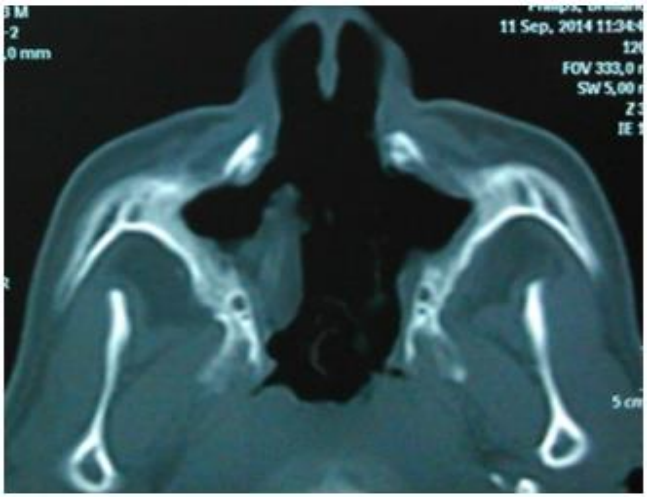

Fig 3: CT scan of paranasal sinuses and skull showing lysis of nasal septum and the intersinusonasal walls.

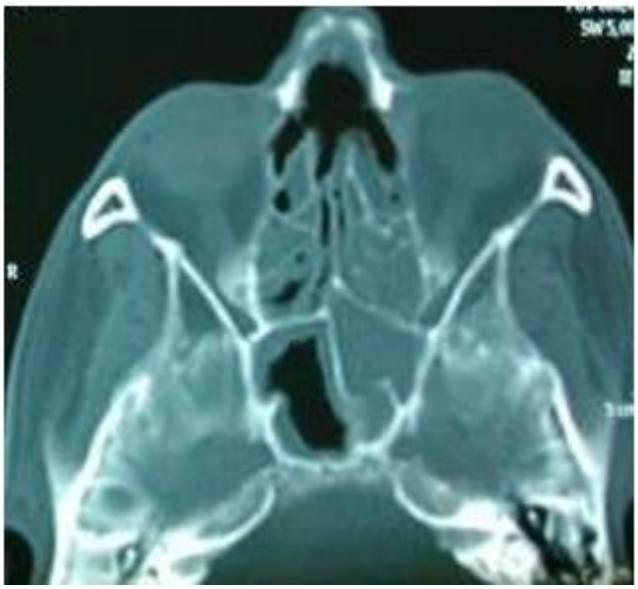

Fig 4: Sinuses CT scan showing mucosa's inflammatory thickening of ethmoidal and sphenoidal sinus.

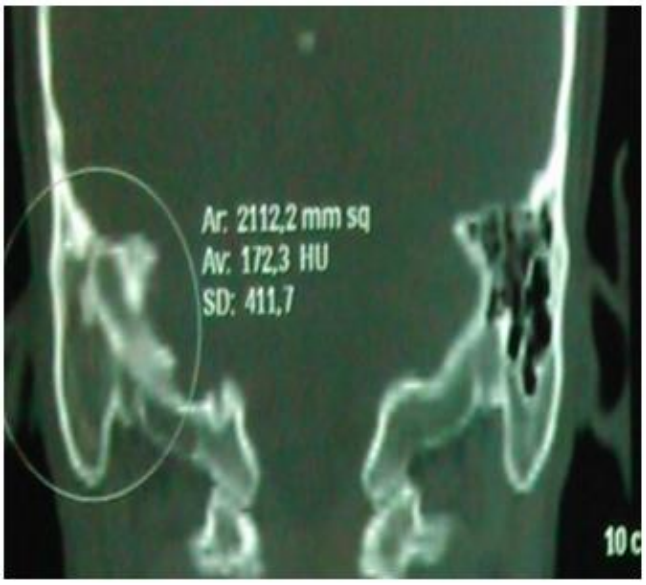

Fig 5: Skull CT scan objectiving edema of right petrous bone and skull base's.

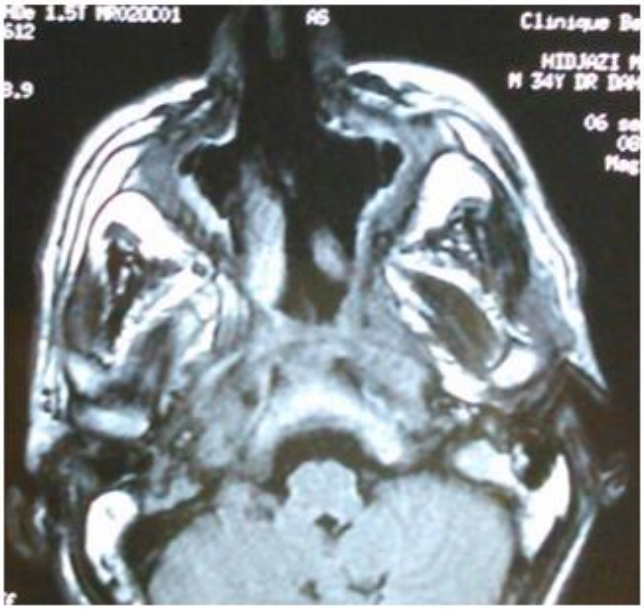

Fig 6: RMI of paranasal sinuses and skull showing lysis of nasal septum and intersinusonasal walls with nasopharyngeal and auditory tube inflammatory edema.

Nasal cavity biopsy with immunohistochemical study objectived nonspecific granulomatous inflammatory process with giant cells without of lymphomatous process.

All tested serologies were negative (human immunodeficiency virus (HIV), hepatitis B virus, hepatitis C virus). The patient's erythrocyte sedimentation rate and C-reactive protein level were elevated with neutrophilia. He had positive tests for anti-neutrophil-cytoplasmic-antibody (ANCA).

Brain MRI was normal. Lung radio and pulmonary CT showed nodular tissue lesion process in the left lower lobe.

Typical ENT and pulmonary manifestations (without renal involvement), histological results as well as a positive ANCA test allowed the diagnosis of GPA.

Treatment and follow-up were done in collaboration with the internal medicine department, it included corticosteroid therapy (prednisone), biotherapy (RITUXIMAB), anti-pneumococcal vaccination and preventive antibiotic therapy such as co-trimoxasole. Evolution at 2 years was stationary without the occurrence of other complications.

Discussion:

Granulomatosis with polyangiitis, formerly known as Wegener granulomatosis, is a rare multisystem autoimmune disease of unknown etiology. GPA is one of the ANCA associated vasculitis disorders. It is characterized by a pneumo-renal syndrome associated with otorhinolaryngologic manifestations. The skin, eyes, joints, nervous system and heart can also be affected [1].

The disease was first described in the medical literature at the end of the 19th century in a clinical case. Then, in the 1930s, it was named by Friedrich Wegener, who described the clinical triad consisting of lung, kidney and upper respiratory tract compromise. Since 2011, Wegener granulomatosis is known as granulomatosis with polyangiitis [2].

The annual worldwide incidence of GPA is estimated to be 10-20 cases per one million. A higher incidence is noted in the colder regions. It occurs in patients between 45 and 60 years old of both genders and recent studies have shown no sex predilection [3,4].

Various diagnostic criteria have been proposed to diagnose GPA and distinguish the disease from other vasculitides.

The American College of Rheumatology criteria include: a) Urinary sediment showing red blood cell casts or more than five red blood cells per high power field, b) Abnormal findings on chest radiograph, c) Oral ulcer or nasal discharge, and d) Granulomatous inflammation on biopsy. The presence of two or more out of the above-mentioned four criteria was associated with a 92% specificity and 88% sensitivity. [5]

The ELK (E stands for ears, nose, and throat or upper respiratory tract, L for lung, and K for kidney) proposed by DeRemee utilizes ANCA to diagnose. Per these criteria, any typical manifestation involving the ELK along with positive ANCA or typical histopathological finding qualifies for a diagnosis of GPA [6]. We used these diagnostic criteria for our patient.

ENT signs are present in 80 to 100% of patients and are often revealing. There is crusted and ulcerated rhinitis (22%) with nasal obstruction, purulent and bloody rhinorrhea with destruction of nasal cartilage or bone resulting in saddle nose deformity. These signs were present in our patient. Sinus CT scanning is the radiographic test of choice to evaluate sinus disease and shows more or less severe damage. Both categories of deafness are typical of the disease. Conductive hearing loss due to auditory tube dysfunction secondary to nasopharyngeal disease is noted in many cases of GPA and is present in our patient. Sensorineural hearing loss and vestibular dysfunction are seen in some individuals. Middle ear involvement, including serous otitis media and mastoiditis, is also seen like this case report. Subglottic stenosis and bronchial stenosis are a potentially serious complication of GPA. ENT damage is associated with a better vital prognosis but with more relapses [1].

Pulmonary involvement is one of the cardinal features of GPA. It occurs in 45% of patients at presentation and 87% during the course of the disease. Cough, hemoptysis, and pleuritis are the most common pulmonary symptoms. The most common radiographic findings include pulmonary infiltrate (67%) and nodules (58%). These are present in our patient [7].

Renal damage is common (38 to 100% of cases) but can missed [8], case of our patient.

Nervous system involvement may occur in as many as 67% of patients and includes mononeuritis multiplex, sensorimotor polyneuropathy and cranial nerve palsies (observed in the reported case) [7].

In our patient, the clinical data discussed a specific multicentric lesion in particular tuberculosis, TNK lymphomatous process with pulmonary metastasis or other forms of ANCA associated vasculitis.

Neutrophilic leukocytosis, inflammatory anemia, thrombocytosis and elevated inflammatory proteins are the rule in diffuse forms of GPA and are noted in our patient. ANCA are present in more than 90% of cases (high diagnostic value) [9].

Treatment is based on the combination of corticosteroid therapy and immunosuppressive treatment (cyclophosphamide, rituximab, azathioprine, methotrexate…). Treatment is classified into two phases: the induction phase and the maintenance phase [10].

The average life expectancy for a patient with GPA without any treatment is 5 months, with a 1-year survival rate of less than 30%. In recent times more than 80% of patients who are treated are alive at least for eight to nine years. Pejorative forms are common (86%). The main cause of death is infection, especially in the first two years of treatment. One in five deaths is linked to kidney failure [11].

Conclusion:

GPA is a multisystemic disease that can affect virtually all organ systems. It remains a severe, chronic disease that readily recurs. The almost constant ENT manifestations of GPA must attract attention and seek other systemic signs. The ENT doctor is often the link between the patient and the internist doctor, guiding diagnosis for rapid and adequate management.

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