Imaging diagnosis of orbital granulomatosis with polyangiitis

Abstract:
Wegener’s granulomatosis (WG) or granulomatosis with polyangiitis is a vasculardisease with necrotizing granulomatous vasculitis that mainly affect the upper and the lower respiratory tract, and the kidney. The ophthalmologic manifestations are not very usual; they are rarely the first symptoms occurring the WG disease but can complicate it.
Report cas of 35 y man who is known as a carrier of WG thing that was confirmed by clinical and biological tests especialy ANCA, then the patient presented ophthalmic symptoms such as exophtalm, ICH syndrome and bilateral ptosis and the diagnostic of orbital wegener was called up based on CT and MRI where we could see orbital pseudo inflammatory masses and also hypertrophy of sinus’s wall and pachymeningitis.

Key words: Wegener disease; Granulomatosis; polyangiitis; Vascularitis; ocular involvement; pseudotumor; CT; MRI

I. Introduction:
Wegener’s granulomatosis is also called granulomatosis with polyangiite sis a systemic disorder characterized by necrotizing granulomatous vasculitis that was described in 1936 for the first time, its pathophysiology is unknown. It affects mainly the ENT region, the lung and kidneys. Ocular manifestations in WG patients are not the first symptom, it does go along with a systemic change but it can complicate symptoms and spoil life quality of patients.

II. Case Presentation:
35 y man who is known with a history of convulsions in the childhood that never been explored, he was admitted to the ER for a status epilepticus, the first examination, also the biological and radiological analysis didn’t show any abnormality. The patient was sent home with asymptomatic treatment.
01 month later, he was back in the ER with an ICH syndrome, horizontal diplopia and ptosis. The clinical examination has shown a drowsy patient, febrile at 38° with slight bilateral exophtalmos and ptosis in the same side and ophthaloplegia.

Figure 1: Exophtalmos and sinonasal fistula

Lumbarpuncture was practiced with pressure measurement that showed a 36cm H2O (normal pressure is around 12-15 cm of water), then a cytotoxic and bacteriological study of the CSF was normal and the patient was going on meningitis dose of antibiotic.
Ophthalmologic examination didn’t show any sign of vasculitis
Biopsy of nasal cavity was without any characteristics
Radiological exploration with a CT and an MRI after the diagnosis of WG was retained based on biological proofs found 02 intraorbitals masses that could be compatible with granuloma, also a sinus wall thickening and pachymeningitis (figure 2-3).
Figure 2: enhanced CT-scan showing: 02 intraconic and retrocular massessurrounding the optic nerve and repressing the oculomotormusclesthatisheterogenouslyenhanced.

Figure 3: T2 and T1 weighted images MRI: bilateralretroorbital masses that are showinghyerintense signal T2 and hypointense signal T1 withheterogeneousencancement.

The patient wasgoingundercorticotherapythen immunosuppressive, and finallychemotherapyafterseveral relapses. After 4 years of chemotherapywithalkylating agent withoutanyclinicalimprovement the patient wasbreaking out and sohe wasconverted to palliative treatment.

**Discussion:**

Granulomatosis withpolyangiitis (WG) is a rare chronic and idiopathicautoimmunediseasethatis responsible for necrotosis of medium and smallbloodvessel’swalls.

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It affects preferentially the upper and lower airway microvessels so it has a predilection for the lung (95%), paranasal sinus (90%), naso-pharynx and the kidney (95%) which involves prognosis for survival [1]. Ocular involvements are rare but increases in frequency with time [2].

The average age of onset is 40-50 years, but can develop at any age from 9-70 without any sex dominance [3-4].

Biologically, Serum antineutrophil cytoplasmic antibody (ANCA) can be measured in each patient, and it has a high sensitivity and specificity for diagnosis but isn’t pathognomonic for WG.

Histopathologically, the biopsy which is practised in nasal mucosa or directly in the orbital masses shows three types of lesions: ischemic necrosis, giant cell granulomatosis and vasculitis of small and medium vessels [4]. However, the biopsies could not be conclusive and shows just a nonspecific inflammatory type.

Radiologically, the CT scan and the MRI are the key to explain the ophthalmologic symptoms of the patient in first place, then it may suggest the diagnosis of WG and it may appreciate the extension of the disease and the therapeutic response of the patient. The CT scan shows intraorbital and extraocular masses, that are aggressive of the neighboring structures, such as adjacent bone destruction or thickening. However, the CT can not make an affirmative diagnosis between the granulomatosis pseudo-tumor and other primotive or secondary tumor of the eye.

The MRI, will shows unilateral or bilateral intraorbital masses that has the same signal compared with the ocular muscles, lesions generally show slightly lower T1WI and higher T2WI signals, and are homogenously enhanced with indistinct boundaries [5].

Imaging will also found a typical inflammatory granulomatous thickening of the sinus wall beclouded bone destruction or thickening.

The medical treatment is essentially based on immunosuppressives in particular, corticoids and lymphotoxic agents such as cyclophosphamides. When faced at treatment deficiency or failures, some surgical methods remain essential such as endoscopic disobliteration, laryngoplasty or tracheotomy [4].

### III. Conclusion:

Granulomatosis with polyangiitis or Wegener’s granulomatosis (WG) is a lethal systemic disease, that orbital manifestations remain extremely rare, however the diagnosis should be highly suspected in time that a destruction of paranasal sinus bone is associated to homogeneous masses. Early diagnosis and treatment can improve significantly the prognosis and the quality of life of the patients.

### Bibliographie:

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