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Imaging diagnosis of orbital granulomatosiswithpolyangiitis

Abstract :
Wegener'sgranulomatosis (WG) or granulomatosiswithpolyangiitisis a vasculardiseasewichiscausingnecrotizinggranulomatousvasculitisthatmainly affect the upper and the lowerrespiratory tract, and the kidney. The ophtalmologic manifestations are not veryusual ;they are rarely the first symptomesoccuring the WG disease but cancomplicateit.
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 presentedophtalmicsymptosessuch as exophtalmy, ICH syndrome and bilateral ptosis and the diagnostic of orbital wegenerwascalled up based on CT and MRI wherewecouldsee orbital pseudo infammatory's masses and alsohypertrophy of sinus'swall and pachymengitis.
Key words : *Wegenerdisease ; Granulomatosis ; polyangiitis ; Vascularitis ; ocularinvolvement ; pseudotumor ; CT ; MRI*

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I. Introduction :

Wegener'sgranulomatosisalsocalledgranulomatosiswithpolyangiiteisa systémicdisordercharacterized by necrotizinggranulomatousvasculitisthatwasdiscribedin 1936 for the first time, itsphysiopathologyisunknown. It affects manely the ENT region, the lungand kidneys.
ocular manifestations in WG patients are not the first symptom, itdoes go alongwith a systemic changesbut itcancomplicatesymptomes and spoil life qulaity of patients.

II. Case Presentation :

35 y man whoisknownwith a history of convulsions in the childhoodthatnever been explored, hewasadmitted to the ER for a statusepilepticus, the first examination, alsothebiological and the radiologicalanalysisdidn'tshowanyabnormality. The patient was sent home withasymptomatictreatment.
01 monthlater, he was back in the ER with an ICH syndrome, horizontal diplopia and ptosis. The clinicalexmaination has shown a drowzy patient, febrileat 38° withaslightbilateralexophtalmyleftmost (figure 1) with a ptosis in the sameside and ophtalmoplegia.

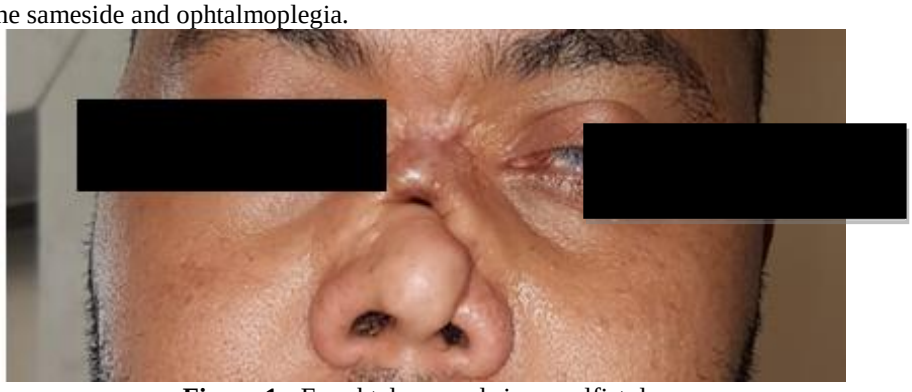

Figure 1 : Exophtalmos and sinonasalfistula

lumbarpuncturewaspracticedwith pressure measurementthatshowed a 36cm H2O (normal pressure isarround 12-15 cm of water), then a cytochemical and bacteriologicalstudy of the CSF was normal and the patient wasgoing on meningitis dose of antibiotic.
Ophtalmologicexaminationdidn'tshowanysign of vasculitis
Biopsy of nasal cavitywaswithoutanycharacteristics
Radiological exploration with a CT and an MRI after the diagnosis of WG wasretainedbased on biologicalproofsfound 02 intra orbitals masses thatcouldbe compatible withgranuloma, also a sinalwallthickening and pachymeningitis (figure 2-3).

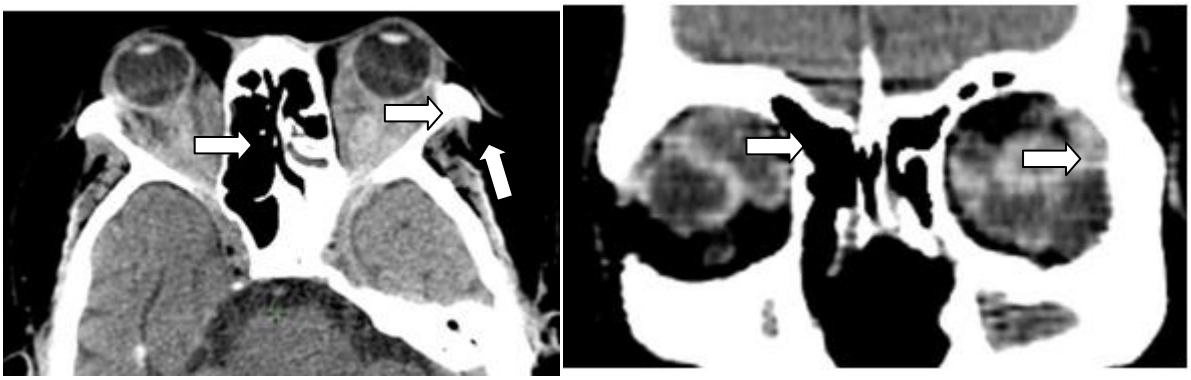

Figure 2 : enhanced CT-scan showing : 02 intraconic and retrocular massessurronding the optic nerve and repressing the oculomotormusclesthatisheterogenouslyenhanced

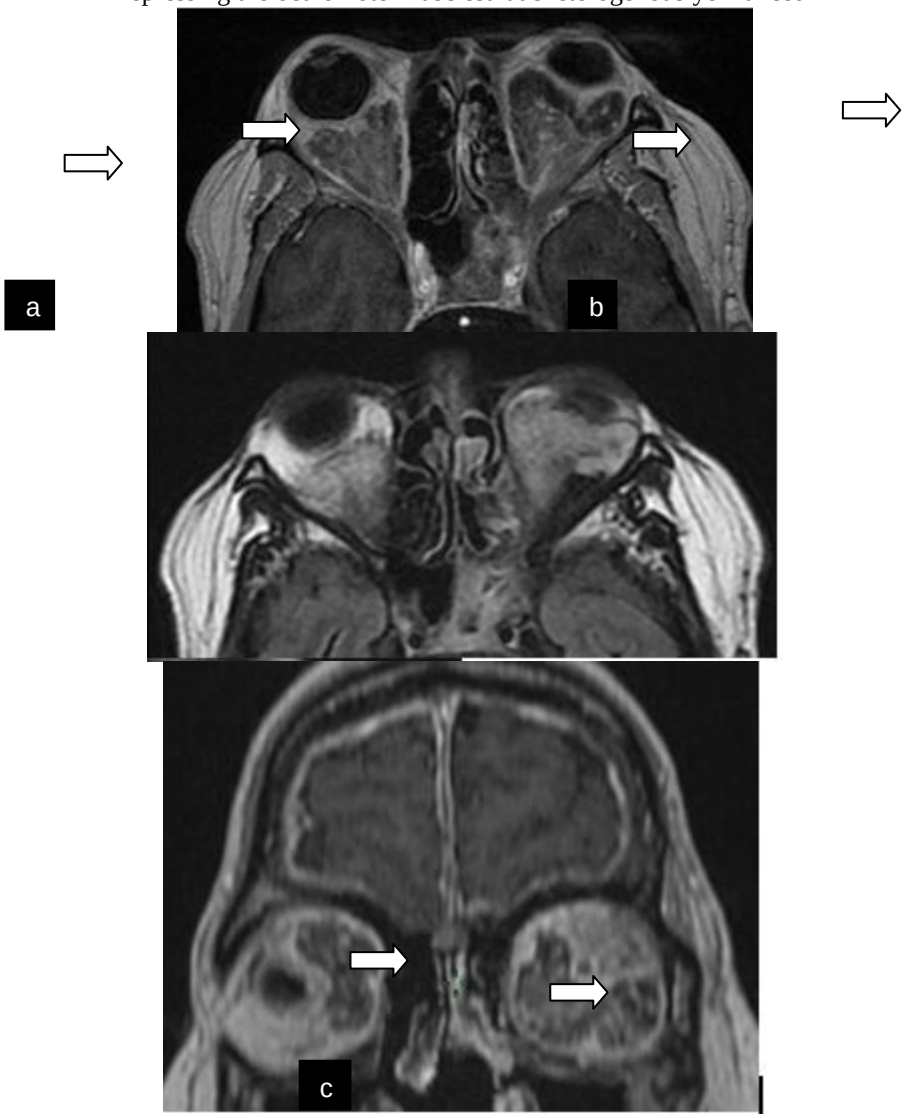

Figure 3 : T2 and T1 weighted images MRI :bilateralretrorbital masses that are showinghyerintense signal T2 and hypointense signal T1 withheterogeneousencancement

The patient wasgoingundercorticotherapythen immunosuppressive, and finalychemotherapyafterseveral relapses. After 4 years of chemotherapywithalkylating agent withoutanyclinicalimprovement the patient wasbreaking out and sohewasconverted to palliativetreatment.

Discussion :

Granulomatosiswithpolyangiitis (WG) is a rare chronicand idiopathicautoimmunediseasethatis responsable for necrotisis of medium and smallbloodvessel'swalls.

It affects preferentiallythe upper and lowerairwaymicrovesselsoit has a preditivetropism for the lung (95%), paranasalsinus(90%), naso-pharynx and the kedney (95%) wichinvolvesprognosis for survival[1]. Ocularinvolvmentis rare but increases in frequencywith time.[2].
The averageage of onsetis 40-50years, but candevelopatanyagefrom 9-70 withoutany sexe dominance. [3-4].
The diagnosisisbased on a body of clinical and biological arguments and canbeconfirmedonly by histology.
Cliniclythe diseasepresentationis the same of a simple sinusitis or chnronicrhinitiswith possible polyposis. Ocularsymptoms are rarely the fristones to show up and couldbeexplaned by the proximity of the eye to paransal sinus ; thosesymptomescanbe : exophtalmy, pain, ptosis, eyelidoedema, eye mouvement limitation. Any segment of theeyecanbeaffected. The anteriorcompartment and the eyeball are the mostlyinvolved part and i twill clinicly shows up as : dacryosistitis, conjunctivitis, keratitis, anteriornecrtizingscleritis.[2]
Biologicly, Serumantineutrophilcytoplasmicantibody (ANCA) canbemeasured ineach patient, and it has a highsensitivity and specificity for diagnosis but itisn'tpathognomonic for WG.
Histopathologically, the biopsie wichispractised in nasal mucosa or directly in the orbital masses shows three types of lesions : ishemicnecrosis, giantcellgranulomatosis ans vasculitis of small and medium vessels. [4]However the biopsies could not be conclusive and shows just a non specificinflammatory type.
Radiologically, the CT scann and the MRI are the key to explain the ophtalmologicsymptoms of the patient in firts place, thenitmaysuggest the diagnosis of WG anditalsomayappreciate the extension of the disease and the therapeuticresponse of the patient.
The CT scan shows intra-orbital and extra-ocular masses, thatare agressive of the neighbour structures, such as adjacent bone destruction or thikning. However, the CT can not make an affirmative diagnosisbetween the granulomatosispsuedo-tumor and otherprimotive or secondarytumor of the eye.

The MRI, will shows unilateral or bilateral intra-orbital masses that has the same signal Comparedwith the ocular
muscles, lesionsgenerally showslightlylower T1WI and higherT2WI signals, and are hmogenouslyenhancedwith indistinct boundaries. [5]the MRI willalso show the granulomatosis extension to the surrounding structures (cavernous sinus, meninges, orbital fissure and optic foramen)

Imaging willalsofound a tipcalinflammatorygranulomatousthickeningof the sinalwallbecloudedbone destruction orthickening.

The medicaltreatementisessentiallybased on immunosuppréssives in particularcorticoides and lymphotoxic agents such as cyclophosphamides. Whenfacedattreatementdeficiency or failuresomesurgicalmethodsremains essential such as endoscopicdisobliteration, laryngoplasty or tracheotomy.. [4]

III. Conclusion :

Granulomatosiswithpolyangiitisor Wegener'sgranulomatosis (WG) islethalsystemicdisease, that orbital manifestations remainextremely rare, however the diagnosisshouldbehighlysuspectedeach time that a destruction of paranasalsinalboneisassociated to homogeneous masses. EarlyDiagnosis and treatementcanimprovesignificantly the pronognosis and the quality of live of the patients.

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