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P0134 - IMMUNOGLOBULIN G4- RELATED DISEASE WITH HYPERTROPHIC PACHYMENINGITIS CAUSING OPTIC NERVE INVOLVEMENT

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Resumen

Objectives: The purpose of this paper is to report a case of IgG4/HP cause his low frequency broad differential diagnostic and mimics pathologies and give to know appropriate criteria for diagnosis and accurate treatment.

Methods: We report a 34 year old man, who presented with a history of eight months of gradual visual loss predominantly right eye without other neurological symptoms. No constitutional or systemic symptoms were presented. He had no past medical History. At neurologic exam the patient was alert, oriented, conserved mental superior functions, Marcus gunn right pupil, right amaurosis, bundle left vision, severe reduction in campimetry.

Results: The patient was treated with surgical resection, by left frontotemporal approach, both optic nerves were explored during surgery, finding tumefactive lesion implanted in meningeal layers of anterior clinoid, sellar tuberculum and minor wings of sphenoid comprising both optic nerves and ipsilateral third cranial nerve, the mass was resected, bilateral optic nerves kept free of compression. After surgery visual acuity do not get worse, not postoperative hormonal disruptions was presented and any complication was described.

Conclusions: IgG4 RHP presented with optic nerve involvement is a pathology very infrequent but potentially treatable that require the physician suspicion and pathologist accuracy to correct diagnosis. Because the differential diagnosis is broad, whitout pathognomonic features and the clinical presentation is very which difficult recognition. Imaging can guide but is necessary complementary studies including serologic test, histopathologic studies and CFS sample and much times multidisciplinary approach. The treatment is based on medical management with corticosteroid therapy as first line treatment which is usually effective and other therapies as second line as rituximab another biologic therapy.