AN UNUSUAL ASSOCIATION OF COGAN'S AND CHURG STRAUSS SYNDROME

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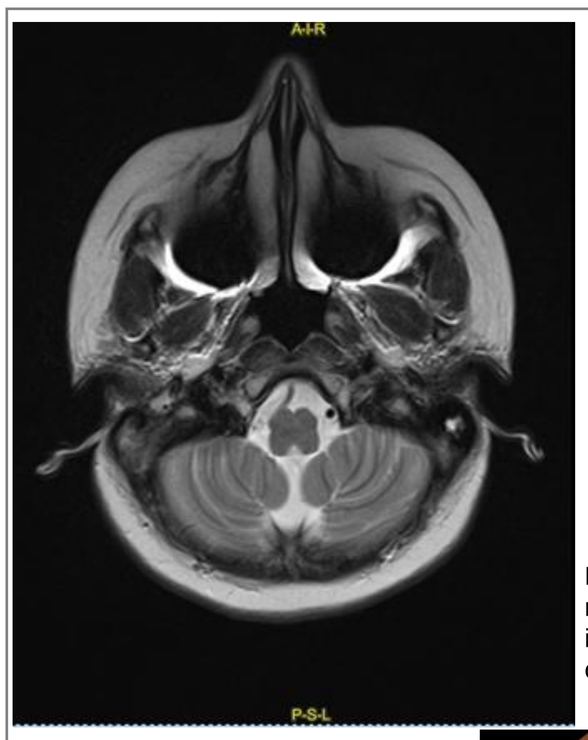
BACKGROUND: Cogan's syndrome (CS) is a rare systemic vasculitis of unknown origin, characterized by audiovestibular and ocular symptoms that may manifest simultaneously or sequentially. The diagnosis of CS is exclusively clinical, based on the Cogan or Haynes criteria. No specific diagnostic laboratory tests or imaging studies exist. To date, the association of CS with Churg Strauss Syndrome (CSS) was not reported.

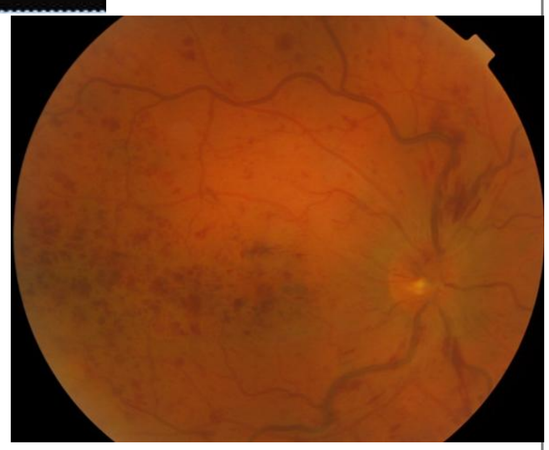
CASE REPORT: We report the case of a 34 year old woman with CSS and methotrexate therapy started two years earlier, who developed suddenly ocular and audiovestibular involvement.
Neurological examination revealed left visual impairment and severe bilateral hearing loss.
Brain MRI and Total Body PET not revealed signs of medium and large vessel vasculitis. Pure Tonal audiometry and Brainstem Auditory Evoked Responses showed serious neurosensory deafness and the fluorangiography showed a picture of central retinal vein occlusion.
Laboratory tests ruled out infective and coagulative disorders, CSF exam was normal.
Antibodies to connexin 26 and reovirus peptide resulted positive and a CS diagnosis was considered. High dose iv steroid therapy and monthly Cyclophosphamide was administered with improvement on visual loss and stabilization of the disease throughout the follow-up.
Instead for deafness a cochlear implant was performed with good results.

DISCUSSION: Diagnosis of CS is often missed or delayed due to its rarity, the non specific clinical signs at onset, and the lack of a confirmatory diagnostic test. Both CS and CSS are considered autoimmune vasculitis but cranial involvement is rare in CSS. In this case we may also suppose an overlap between these conditions.

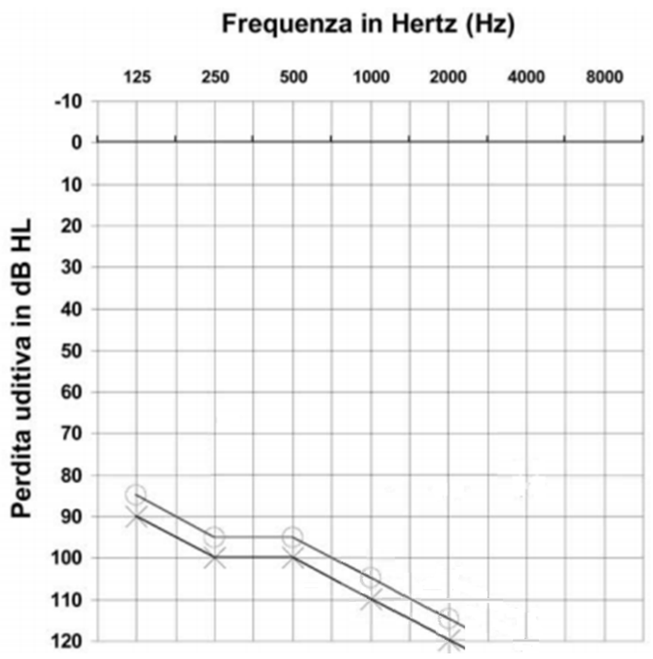
CONCLUSION: A correct and early diagnosis is important as an aggressive treatment should be started as soon as possible. In CS ocular symptoms are more responsive than the vestibuloauditory symptoms and in patients with profound deafness, who did not respond to immunosuppressive treatment a cochlear implant surgery should be performed as soon as possible to achieve the best result.



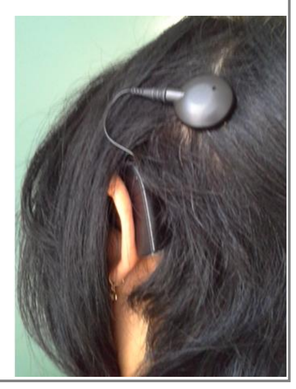
Brain MRI and MR angiography not revealed signs of vasculitic involvement neither any other kinds of pathologic signs



fluorangiography showing central retinal vein occlusion



Tonal audiometry showing symmetric bilateral hypoacusia of neurosensory type of deep entity



Bibliografia

Cogan syndrome—Pathogenesis, clinical variants and treatment approaches - A Kessel et al.

Cogan's Syndrome and Other Ocular Vasculitides
Gabriela M. Espinoza

Cogan's Syndrome—Clinical Guidelines and Novel Therapeutic Approaches - Oshrat E. Tayer-Shifman

Sudden hearing loss and Crohn disease: when Cogan syndrome must be suspected – G Tirelli, MD