DOES INCOMPLETE IRVAN SYNDROME EXIST?

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INTRODUCTION

Idiopathic retinal vasculitis, aneurysms and neuroretinitis (IRVAN) syndrome is a rare clinical entity characterized by BILATERAL RETINAL ARTERITIS, numerous ANEURYSMAL DILATATIONS of retinal and optic nerve head arterioles, NEURORETINITIS and UVEITIS. The diagnosis is based on some clinical features such as anterior uveitis, vitritis, multiple aneurysmal dilatations, perivenous sheathing, disc oedema, macular star and intraretinal lipid deposition. Fluorescein angiography may demonstrate the presence of peripheral capillary nonperfusion. Visual loss is caused by exudative maculopathy and neovascular sequelae of retinal ischemia. Laser photoagulation may be beneficial if extensive ischemia.

CLINICAL CASE

A 21-year-old women presented with blurred vision for 10 days in her left eye (LE). Visual acuity (VA) was 0.9 in right eye (RE) and less than 0.1 in LE. Anterior segment examination revealed the presence of tyndall ++, with no further abnormalities in both eyes. Fundoscopy examination showed small areas of vasculitis in periphery of RE, and mild vitritis, perivenous sheathing, disc oedema, intraretinal hard exudates, macular scar, and retinal diffuse oedema in LE. Fluorescein angiography showed focal venous leakage along with areas of capillary nonperfusion in periphery of both eyes, and diffuse retinal and optic nerve leakage in LE.

The WORK-UP included: complete blood analysis with autoimmune tests, evoked potentials, VDRL, FTA-ABS, urinalysis, tuberculin skin testing, infectious serology, lumbar puncture, MRI, and chest radiograph. No abnormalities were found in all these complementary exams.

SLOW-TAPERING OF SYSTEMIC AND TOPICAL STEROIDS WAS STARTED.

ONE YEAR LATER:
- VA remained unchanged.
- Anterior segment showed no signs of inflammation.
- Fundoscopy revealed absence of activity in both eyes, and the presence of an atrophic optic nerve in LE.

CONCLUSIONS

This case might represent an incomplete form of IRVAN syndrome, without aneurysms, or an atypical form of idiopathic vasculitis with papillitis and great hard exudation. Anyway, careful long-term follow-up is recommended for this kind of patients, as oral steroids can be ineffective and visual function can be severely deteriorated.

- There are no cases described in peer-reviewed literature about incomplete IRVAN syndrome. However, the optic disc oedema and the extensive intraretinal lipid deposition in our patient resembled it.
- However, the absense of aneurysms make it fairly impossible, as it is one of the major criteria considered for IRVAN syndrome until now.
- Another option would be the existence of an ATYPICAL FORM OF IDIOPATHIC VASCULITIS WITH PAPILLITIS AND GREAT HARD EXUDATION.
- Anyway, this kind of clinical presentation deserves careful examination, as oral steroids can be ineffective and visual function can be severely deteriorated.

REFERENCES


