

## SHORT COMMUNICATION

Case report

# Central retinal vein occlusion and sudden deafness: a possible common pathogenesis

A. GLACET-BERNARD<sup>1</sup>, W. ROQUET<sup>1</sup>, A. COSTE<sup>2</sup>, R. PEYNÈGRE<sup>2</sup>, G. COSCAS<sup>1</sup>, G. SOUBRANE<sup>1</sup>

<sup>1</sup>University Eye Clinic of Créteil, University of Paris XII, Créteil

<sup>2</sup>Department of Otorhinolaryngology and Head and Neck Surgery, Intercommunal and Henri Mondor Hospitals, University of Paris XII, Créteil - France

**CASE REPORT.** *A 31-year-old woman complained of blurred vision in the right eye due to a well-perfused central retinal vein occlusion (CRVO) and two months later, of sudden deafness (SD) in her right ear. Her visual acuity and hearing recovered almost completely within a few months. Medical evaluation disclosed the combination of slight coagulation abnormalities (moderate decrease in protein S, slightly elevated lipoprotein (a)), and elevated fibrinogen, with plasma hyperviscosity.*

**DISCUSSION.** *The occurrence of CRVO then SD suggests that the same underlying conditions can be considered as risk factors for both diseases and shows up some similarities in the pathogenesis of these acute impairments of microvascular blood flow in the retina and the cochlea. (Eur J Ophthalmol 2001; 11: 197-9)*

**KEY WORDS.** *Retinal vein occlusion, Sudden deafness, Viscosity, Microcirculation, Thrombosis*

Accepted: September 11, 2000

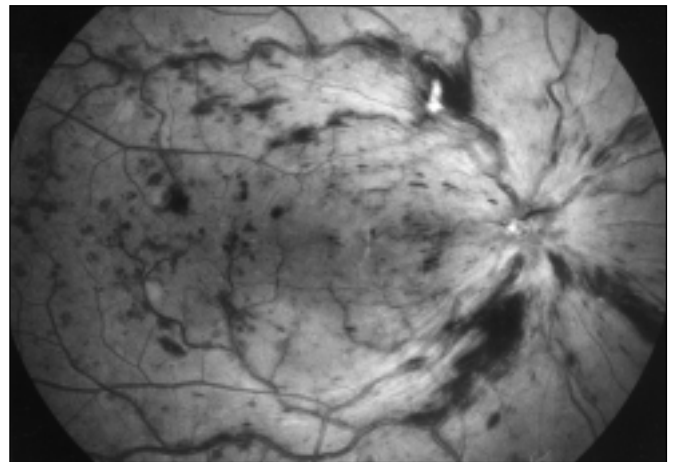
## INTRODUCTION

We report the case of a young woman who presented central retinal vein occlusion (CRVO) followed by sudden deafness (SD), to illustrate some similarities in the pathogenesis of these acute impairments of microvascular blood flow in the retina and the cochlea.

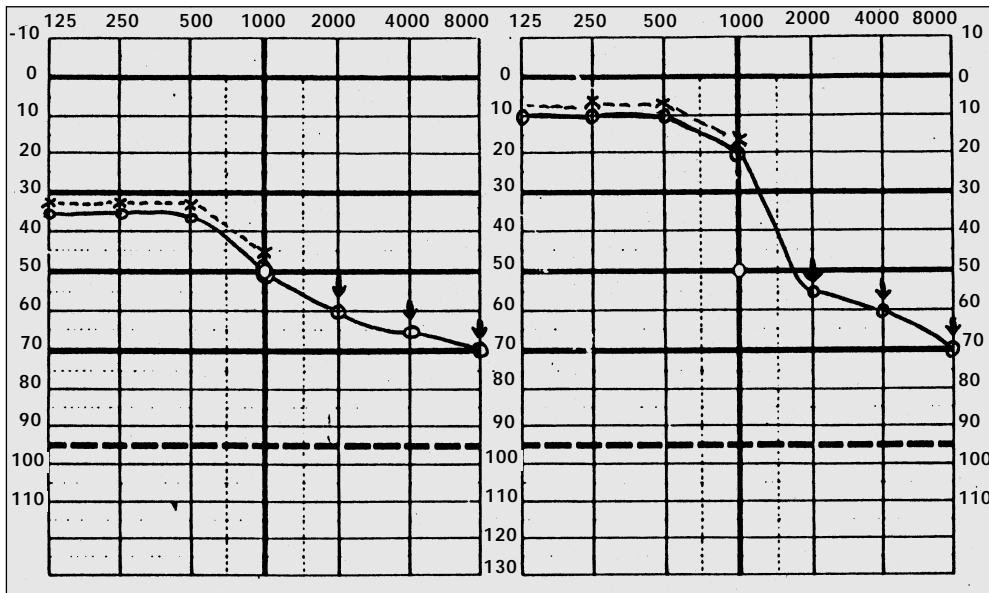
## Case report

A 31-year-old Caucasian woman complained of a two-week history of blurred vision in her right eye. She has had an uncomplicated delivery two weeks before. She had not wanted to breast feed her child and had been given oral contraceptives ten days before (combination of 30 µg ethinyl estradiol and progesterone). The patient had had three normal pregnancies, with three healthy children. She had been taking oral contraceptives for ten years. She has neither a medical nor a family history.

At presentation, visual acuity was 20/80 RE and 20/20 LE. Anterior segments and intraocular pressures were normal. Ophthalmoscopic examination revealed



**Fig. 1** - *Fundus photograph of the right eye showing a recent CRVO with venous tortuosity, hemorrhages in all quadrants and a cotton-wool spot on the superotemporal vessels.*



**Fig. 2** - Two months later, when the patient presented sudden hearing loss in her right ear, pure tone audiogram shows a perceptible hearing loss affecting both low and high frequencies (on the left). One week later, the patient had achieved almost complete recovery except for the higher frequencies (right).

a CRVO with flame-shaped hemorrhages in all quadrants, numerous cotton-wool spots at the posterior pole and disc and macular edema. Fluorescein angiography showed a well-perfused CRVO (Fig. 1). Left eye was normal. The patient was given a regimen of troxerutin (7 g daily) and prednisone (30 mg daily). Oral contraceptives were discontinued.

Thorough systemic evaluation including coagulation and immunological investigation revealed an elevated fibrinogen level (5.25 g/L), with plasma hyperviscosity, protein S activity at the lower limit of normal (76%) in relation to the recent pregnancy and oral contraceptives, slightly elevated lipoprotein (a) (0.33 g/L, normal < 0.3 g/L) and antinuclear antibodies at a non-significant titer of 1:50. Blood cell count, protein C, antithrombin III, activated protein C resistance and homocysteinemia were normal, as were the protein electrophoretic profile, creatinine, glucose and lipids. Anticardiolipin antibodies and lupus anticoagulant were negative, as were circulating immune complexes. Echocardiography and carotid ultrasound examination were normal.

Two months after the onset of CRVO, the patient complained of sudden hearing loss in her right ear, associated with vertigo. Pure tone audiometry showed a perceptible hearing loss involving both low and high frequencies (Fig. 2). MRI excluded an acoustic neurinoma. The patient was given intravenous nicergoline for three days, with carbogen inhalation

and oral prednisolone (60 mg/day). Over the following weeks, the patient almost completely recovered her hearing except in the higher frequencies. Visual acuity progressively recovered to 20/25 and fundus appearance returned to normal over the next ten months.

## DISCUSSION

The successive occurrence of CRVO and SD suggests that the same underlying conditions may be considered as risk factors for both diseases and points to some similarities. Like for CRVO, the pathogenesis of SD has not yet been totally elucidated (except in cases with an acoustic neurinoma), but an acute impairment of microvascular perfusion is thought to play a major role, involving abnormalities of plasma and blood viscosity, coagulation disorders and/or immune mediated processes (1-3). In our patient, hormonal changes due to the recent pregnancy and to the oral contraceptives may be taken into account for the vascular impairment.

Although no definite guidelines are available, similar treatment protocols are used in both diseases, including vasodilators, hemodilution, anticoagulants and steroid therapy. Cochlear and retinal blood flow show some features in common, such as a terminal microcirculation with a tight junction endothelium, and an internal pressure (of aqueous or endolymph), which

may result in limited adaptation to hemodynamic variations. Although retinal and cochlear arterial occlusion has been already reported in the rare SICRET or Susac's syndrome (4), the association of CRVO and SD has not yet been described.

In conclusion, this case report sheds light on the similarities between the anatomical and pathogenic features of CRVO and SD, suggesting that these two diseases may be explained by similar factors, and that the incidence of their simultaneous occurrence is probably underestimated.

Reprint requests to:  
Agnès Glacet-Bernard, MD  
Service d'Ophtalmologie  
40, avenue de Verdun  
94010 Créteil, France  
e-mail: agnes.glacet@chicreteil.fr

---

## REFERENCES

1. Mosnier I, Bouccara D, Sterkers O. Sudden hearing loss in 1997: etiopathogenic hypothesis, management, prognostic factors and treatment. *Ann Otolaryngol Chir Cervicofac* 1997; 114: 251-66.
2. Williamson TH. Central retinal vein occlusion: what's the story? *Br J Ophthalmol* 1997; 81: 698-704.
3. The Eye Disease Case-Control Study Group. Risk factors for central retinal vein occlusion. *Arch Ophthalmol* 1996; 114: 545-54.
4. Susac OJ. "Susac's syndrome". The triad of microangiopathy of brain and retina with hearing loss in young women. *Neurology* 1994; 44: 591-3.