Intravitreal ranibizumab injection for choroidal neovascularization in Strümpell-Lorrain Syndrome

Néovascularisation choroïdienne associée au syndrome de Strümpell-Lorrain traitée par ranibizumab

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Summary Strümpell-Lorrain syndrome, or hereditary spastic paraplegia is a genetic disease of the central nervous system affecting the spinal cord and cerebellum. It represents a clinically heterogeneous group of neurodegenerative disorders characterized by progressive spasticity and hyperreflexia of the lower limbs. Ocular abnormalities include keratitis, macular pigmentary abnormalities, juxtafoveolar retinal telangiectasis and choroidal neovascularization. We report the first case of choroidal neovascularization associated with Strümpell-Lorrain syndrome treated successfully with intravitreal ranibizumab injection.

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MOTS CLÉS
Antib-VEGF; Choroidal neovascularization; Hereditary spastic paraplegia; Strümpell-Lorrain syndrome

Résumé La maladie de Strümpell-Lorrain ou paraplégie spastique familiale est une maladie génétique du système nerveux, touchant la moelle épinière et le cervelet. Les anomalies oculaires associées à ce syndrome sont les kératites, les atteintes maculaires comme les anomalies pigmentaires, les télangiectases juxtafovéolaires et les néovascularisations choroïdiennes. Nous rapportons ici le premier cas de néovascularisation choroïdienne, associée au syndrome de Strümpell-Lorrain, traitée par injection intravitréenne de ranibizumab.

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Strümpell-Lorrain syndrome, also called hereditary spastic paraplegia (HSP) and familial spastic paraplegia, is a group of clinically and genetically diverse disorders [1]. This disease is estimated to affect 9.6 individuals in 100,000. HSP may be inherited as an autosomal dominant, autosomal recessive or X-linked recessive trait. Multiple recessive and dominant forms exist. Ocular abnormalities, including macular changes, pigmentations and retinal degeneration have been described already in the older literature [2]. More recently, juxtafoveal retinal telangiectasis and subretinal neovascularization have been reported as an unusual association of familial spastic paraplegia [3]. Here, we report a patient with Strümpell-Lorrain syndrome and subretinal neovascularization, successfully treated by intravitreal anti-vascular endothelial growth factor (VEGF).

A 19-year-old woman was referred to our department for sudden decrease of vision in his left eye (LE). The patient and her father were affected with Strümpell-Lorrain syndrome due to SPG3A mutation [4]. Best-corrected visual acuity (BCVA) was 20/20 in the right eye (RE) and 20/200 in the LE. Fundus biomicroscopy revealed normal findings in the RE, and a raised lesion in the LE, located in the temporal perifovea, with pigmentary changes and subretinal hemorrhages (Fig. 1). Fluorescein angiography (FA) and indocyanine angiography (ICGA) showed normal findings in the RE, and demonstrated intense hyperfluorescence due to leaking subretinal neovascularization in the LE (Fig. 1). Interestingly, FA and ICGA, together with spectral-domain optical coherence tomography (SD-OCT) revealed the presence of a retinal-choroidal anastomosis within the neovascular membrane (Fig. 1).

Intravitreal ranibizumab injection (0.5 mg/0.05 mL) was administered to treat the neovascularization. At month-1, BCVA increased to 20/20, and FA, ICGA and SD-OCT revealed the CNV closure and absence of intraretinal and subretinal fluid. A total of three intravitreal injections. Visual gain and CNV closure were maintained at month-12 follow-up visit (Fig. 2).

To our knowledge, the association of subretinal neovascularization with Strümpell-Lorrain syndrome is rare. To date there is only one report to document the development of subretinal neovascularization in patients with Strümpell-Lorrain syndrome, but no treatment was provided [3]. Intravitreal injections of ranibizumab, has shown to improve visual acuity safely in eyes with neovascular age-related macular degeneration (AMD) [5,6]. Subretinal neovascularization associated with Strümpell-Lorrain syndrome may be due to different pathogenic mechanism compared with AMD. In fact, juxtafoveal retinal telangiectasis has been reported to precede the development of subretinal neovascularization in Strümpell-Lorrain syndrome [3]. Despite these differences in pathogenesis, anti-VEGF therapy appeared effective in our case of subretinal neovascularization associated with Strümpell-Lorrain. Based on these findings, intravitreal ranibizumab could be considered as a treatment option for subretinal neovascularization associated with Strümpell-Lorrain syndrome.

**Figure 1.** Fundus color of the left eye showing a raised lesion in the temporal perifovea with pigmentary changes and subretinal hemorrhages at baseline (A). Fluorescein angiography (B) and indocyanine angiography (C) intense hyperfluorescence due to leaking subretinal neovascularization. Spectral-domain optical coherence tomography showing intraretinal and subretinal fluid, and revealing the presence of a retinal-choroidal anastomosis within the neovascular membrane (D, arrow).
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Figure 2. Fundus color of the left eye showing a fibrotic lesion in the temporal perifovea with pigmentary changes (A), at month-12 follow-up visit, after three intravitreal injections of ranibizumab. Fluorescein angiography (B) and indocyanine angiography (C) showing staining of the fibrotic lesion and absence of late leakage. Spectral-domain optical coherence tomography (D) showing absence of intraretinal and subretinal fluid.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References