Short communication

Optic nerve infarction in Sneddon’s syndrome ☆, ☆

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A B S T R A C T

Clinical case: We report a case of a 26 year old woman with Raynaud’s phenomenon and livedo reticularis 2 years onset, who presented with visual field defects accompanied by hypertension, hematuria, and headache. The examination revealed multiple retinal and optic nerve head infarcts in both eyes.

Discussion: Sneddon’s syndrome is a vasculitis that produces livedo reticularis, neurological symptoms, and less frequently myocardial, renal and retinal infarction. After reviewing the literature, this is the first case described of Sneddon’s syndrome presenting with optic nerve infarction.

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R E S U M E N

Caso clínico: Se expone el caso de una mujer de 26 años con fenómeno de Raynaud y livedo reticularis de 2 años de evolución que se presentó con alteraciones del campo visual acompañadas de hipertensión arterial, hematuria y cefalea. En la exploración se objetivaron múltiples infartos retinianos y de cabeza del nervio óptico bilateral.

Discusión: El síndrome de Sneddon es una vasculitis que cursa con livedo reticularis, clínica neurológica y, menos frecuentemente, con infartos de miocardio, renales y retinianos. Revisando la literatura podemos decir que se trata del primer caso de síndrome de Sneddon descrito que se inició con infartos del nervio óptico.

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Clinic case

Female, 26, with history of Raynaud phenomenon and livedo reticularis (LR) with 2 years evolution, without ophthalmological antecedents (Fig. 1).

The patient visited the practice with arterial hypertension (244/108 mmHg), deteriorated kidney function, sensitive-motor disorder of upper limbs and scotoma in the visual field of the left eye (LE). Magnetic resonance revealed microinfarcts in the deep territory of the right medial brain artery. Kidney biopsy exhibited thrombotic microangiopathy, with no alterations in skin biopsy. The first suspicion was cerebral vasculitis of immunological origin, and accordingly bolus of methylprednisolone, clopidogrel and atorvastatin were prescribed (Fig. 2).

Since admission, the patient referred nasal scotoma in LE. Ophthalmological examination revealed visual acuity of 1.0 in both eyes (BE), afferent pupil defect in LE. Confrontation campimetry with loss of the temporal-superior and nasal-inferior hemifields in LE, with right eye (RE) being normal. Ishihara RE 19/20 and LE 9/20. Normal extrinsic ocular motility. Anterior segment was normal and posterior pole exhibited multiple microinfarcts in the retina and the optic nerve head in BE. Analytics produced normal hemogram, biochemistry and immunological tests (Fig. 3).

In the presence of LR and brain, retinal and optic nerve infarcts and after discarding other immunological causes, the patient was diagnosed with Sneddon syndrome (SS), prescribing treatment with acenocoumarol, enalapril and nitroglycerin patches.

Four months after admission and with patient stability allowing, visual field examination was performed which evidenced temporal and nasal-superior loss in RE and...

Fig. 1 – Images of livedo reticularis of right foot and left hand.

Fig. 2 – Nuclear magnetic resonance showing multiple small hypertense lesions corresponding to right acute-subacute infarcts in deep territory of ACM (arrow).
Fig. 3 – (A) RE: well-defined and not over-elevated papilla. Inferior and retinal cotton-like exudates. (B) LE: papilla with blurred edges, slightly over-elevated. Multiple cotton-like exudates in retina and optic nerve head. Lipidic exudates and intraretinal hemorrhages in the papillomacular bundle.

concentric loss in LE. Papillary optic coherence tomography (OCT, Topcon 3D-OCT 2000 Co. Tokyo, Japan) revealed nasal-superior atrophy in RE and inferior in LE (Figs. 4 and 5).

One year later, the patient referred stability, visual acuity of 0.6 in RE and 0.9 in LE. Afferent pupil defect persisted in LE, normal Ishihara in RE and 10/20 in LE, and bilateral papillary paleness. The visual field exhibited several focal defects in RE and concentric defect in LE, while OCT revealed persistence of the supero-nasal alteration in RE and inferior and superior in LE (Figs. 4–6).

Fig. 4 – Papillary paleness in BE one year after admission. RE, LE respectively.
Discussion

SS is vasculitis of small and middle sized arteries which mainly affects the skin and the central nervous system.\(^1,2\)
It can also affect other organs such as kidneys, heart, eyes and the peripheral nervous system.\(^3\) The incidence of SS is of 4/1,000,000 inhabitants/year. It mainly affects women between 25 and 35. It exhibits 10% mortality and significant morbidity. The literature describes cases of recessive autosomic inheritance without identifying any involved gene.\(^4\)

The mortality index is of 9.5% with a mean follow-up of 6 years.

SS histology is characterized by proliferation and migration of smooth muscle cells, colonizing the intimal subendothelial space with narrowing of vascular patency and formation of intravascular thrombi.\(^5\)

LR are irregular lesions of cyanotic appearance which are not influenced by temperature. They appear with diminished blood flow derived from arterial obstruction or local of vasoconstriction of skin venules. It can precede in up to 10 years the appearance of neurological clinic. Said clinic includes cephalia, vertigo, brain infarcts and cognitive deterioration due to multiple brain infarcts in late stages. Cardiovascular clinic could comprise arterial hypertension, acute myocardium infarct and mitral valve alterations.

In a review of the literature, the authors have found ophthalmological clinic describing central retina artery obstructions,\(^6,7\) one case of retinal capillary obstruction with associated neovascularization,\(^8\) visual field alterations of the hemianopsia type\(^9\) and central retinal vein obstruction.\(^10\)
In the patient referred herein, the authors found retinal and optic nerve capillary infarcts, which makes it the first described case of optic nerve head infarcts in SS.

SS is diagnosed with the presence of LR associated to neurological clinic, in all cases discarding other causes of vasculitis.\(^1\) The association of compatible histological study is not essential due to the difficulty in obtaining good quality samples. Laboratory analyses are generally normal.

Differential diagnostic must include systemic erythematous lupus, antiphospholipid syndrome, cryoglobulinemia, nodous panarteritis, livedoid vasculitis, cholesterol funnels and atherosclerosis, among others.

There is no effective treatment. The use of glucocorticoids or immunosuppressants has not demonstrated beneficial effects for preventing cardiovascular events. Recommended treatments include anticoagulation, antiaggregation, strict control of arterial pressure and cholesterol levels in plasma. In addition it is recommended to avoid smoking, oral contraceptives and overweight.

SS is a rare and difficult to manage entity that should be included in differential diagnostic in the presence of retinal vascular alterations in young patients, mainly women, with
a history of LR and neurological alterations, in all cases after discarding other immunological causes.

Conflicts of interests

No conflict of interests was declared by the authors.

References