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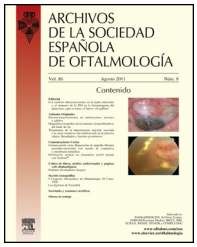

Short communication

# Papilledema secondary to a superior sagittal sinus thrombosis. Mantle cell lymphoma paraneoplastic syndrome☆

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ABSTRACT

*Clinical case:* A 46-year-old patient presented with visual loss in the left eye during the previous months. Ophthalmoscopic examination and magnetic resonance angiography found the presence of papilledema due to thrombosis in superior sagittal sinus. The examination findings revealed a mantle cell lymphoma.
*Discussion:* Cerebral venous thrombosis is an unusual cause of papilledema. This type of thrombosis may be secondary to hyper-viscosity within a context of a paraneoplastic syndrome.



## Papiledema secundario a trombosis del seno sagital superior. Cuadro paraneoplásico de un linfoma del manto

*Palabras clave:*
Angiorresonancia
Linfoma del manto
Papiledema secundario
Trombosis venosa cerebral
Seno sagital superior

RESUMEN

*Caso clínico:* Se presenta el caso de un varón de 46 años con disminución de la visión del ojo izquierdo de meses de evolución. Se diagnostica de papiledema por trombosis del seno sagital superior gracias a la angiorresonancia. En busca de la etiología de la trombosis se descubre un linfoma del manto.
*Discusión:* La trombosis venosa cerebral es una causa poco frecuente de papiledema. Puede deberse a cuadros de hiperviscosidad en el contexto de un síndrome paraneoplásico.



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## Introduction

Papilledema (PE) is a bilateral papillary edema secondary to intracranial hypertension. In contrast with other optic nerve conditions, it has very little repercussion on visual acuity (VA). The most frequent cause of PE is idiopathic intracranial hypertension which frequently expresses in obese women in reproductive age. Brain venous thrombosis is an infrequent cause of PE which must be taken into account in the presence of an atypical PE, i.e., a male with good VA.[1,2]

## Clinic case report

A 46-year-old male patient with diminished VA in the left eye (LE) with 2 months evolution, associated headache for several months with a diagnostic of bilateral sinusitis was presented. The VA of both eyes was 1. Funduscopy revealed edema of nervous fiber layer in both eyes, larger in LE, associated to papillary hemorrhage (Fig. 1a and b). The visual field exhibited increased blind spot (Fig. 2a and b). Blood pressure was normal.

The analysis produced the following data: hemogram with normal red series and platelets. Leukocytosis of 33,500 with large atypical lymphocytes exhibiting grainy, chromatin nuclei, some of them activated, with frequent nuclear shadows. Coagulation was normal. General and liver biochemistry normal. C-reactive protein 0.8 mg/dl supplements normal. Proteingram and immunoglobulin normal. Negative for infectious serologies. Elementary urine normal, excepting protein traces. Urgent axial computerized tomography showed occupation of maxillary sinuses compatible with sinusitis, absence of full capture at the level of the superior sagittal sinus (SSS) (empty Delta sign).

A lumbar puncture was performed with an opening pressure of 50 cm/$H_2O$. Biochemistry: glucose: 5.737 mg/dl, overall protein: 18.7 mg/dl, appearance of rock water. Cytology: cell count of zero cells/$mm^3$; subsequently, after the patient was admitted, humor infiltration was demonstrated with flow cytometry.

Cranial magnetic resonance (MR) and angio-MR in venous phase exhibited occupation of SSS compatible with thrombosis extending to the right transversal sinus (Fig. 3a and b).

The patient was admitted with a diagnostic of PE secondary to SSS thrombosis with possible septic etiology. Treatment was established with anticoagulants and antibiotic. The patient was referred to the Hematology Department for studying possible lymph-proliferative syndrome. The extension study revealed supra- and infra-diaphragmatic ganglion involvement with infiltration of peripheral blood, bone marrow and central nervous system. Medullogram revealed a high percentage of polymorphic lymphocytes (84%) and nuclei occupying almost the entire cell, some with 1 or 2 grooves (Fig. 4). The diagnostic was phenotype B-λ histological type non-Hodgkin mantle cell lymphoma Ann-Arbor stage IV-A with high-risk prognostic rating. Treatment was initiated with intrathecal chemotherapy with liposomal cytarabine in parallel with poly-QT of the Hyper-CVAD type. After achieving complete remission, an autogenous transplant was performed.

Two months later the edema with major residual papillary paleness in LE disappeared. In routine checkups, the ocular fundus and visual field do not exhibit new alterations and final VA of 1 is maintained in both eyes.

## Discussion

Thrombosis in sinuses and cerebral veins is a rare disease. These thromboses are mainly located in the SSS (62%) and the transversal sinus (40–45%). In up to 90% of cases the location of the thrombosis compromises more than one sinus, as in the present case.[3–5]

The most frequent causes of SSS thrombosis are pregnancy, puerperium and contraceptives. However, it is also associated to predisposing host factors such as hypercoagulability, congenital or acquired thrombophilia, immunomediated inflammatory diseases, neoplasia, infections, traumatisms, hyperviscosity syndromes, cardiac insufficiency and severe dehydration, among others.[6]

In the present case, thrombosis exhibited tumor etiology. Thrombosis in SSS is more frequently associated to metastasis followed by hematological (>14%) and lastly solid organ tumors.[7] The involvement could be due to local invasion by the tumor, a condition of hyperviscosity or a paraneoplastic condition.[3,7,8]

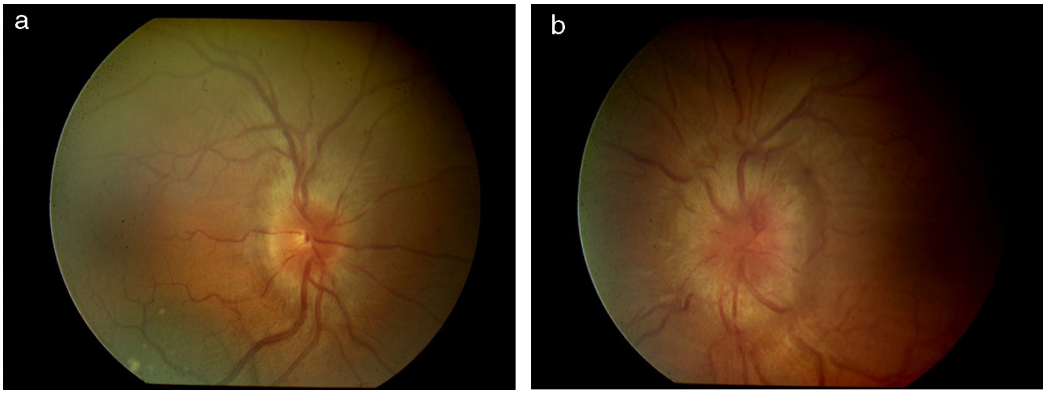


**Fig. 1 – (a) Right eye retinography. (b) Left eye retinography. Both eyes exhibit papillary edema with peripapillary hemorrhage in the left eye.**