DOI 10.5935/0034-7280.20150051

# Retro-orbital tumor suggestive of optic nerve sheath meningocele

## *Tumoração retroorbitária sugestiva de meningocele da bainha do nervo óptico*

Michelle Rodrigues Gonçalves Dias Chaves[1], Isabella Bezerra Wanderley de Queiroga [2], Mario Augusto Pereira Dias Chaves [1,3,4], Fernando Melo Gadelha [1,2] , Debora Apolônio Vieira [2]


### *ABSTRACT*

*Meningocele of the optic nerve sheath is an extremely rare condition with a few cases reported in literature. Image studies reveal tubular-cystic enlargement of the optic nerve although with the same thickness. Symptoms are often related to the involvement of the optic nerve, leading from slow to accelerated decreasing of the visual acuity. The early surgical treatment is the decompression of the optic nerve sheath, which it could provide improvement of visual function. We are presenting a case report of a patient who showed clinical and radiological signs of this rare pathological condition. Male patient attended at service complaining of proptosis of right eye (OD) since birth, in progress during the last months associated to stabbing pain. Best corrected visual acuity (BCVA) of OD was movements at 50 cm far; OS showed no abnormalities. Nuclear Magnetic Resonance of the OD showed expansive formation with cystic aspect, defined boundaries, located in an intraconal situation on the right orbit cavity and in a closing anatomical relationship to the optic nerve, inducing compression, deformity and anterior displacement of this eye besides presenting signal similar to spine liquor in all sequences obtained. The first hypothesis was meningocele of right optic nerve sheath. Then, patient was referred for surgical decompression.*


***Keywords**: Meningocele/diagnosis; Exophthalmia; Optic nerve/pathology; Neoplasms; Case reports*


### RESUMO

Meningocele da bainha do nervo óptico é uma condição extremamente rara, com poucos casos relatados na literatura. Exames de imagem revelam alargamento tubular-cístico do nervo óptico, com espessamento do mesmo. Os sintomas são muitas vezes relacionados com o comprometimento do nervo, ocasionando diminuição de lenta a acelerada da acuidade visual. O tratamento cirúrgico precoce por meio da descompressão da bainha do nervo óptico pode proporcionar melhora da função visual. Apresenta-se um caso de paciente com as características clínicas e radiológicas desta condição patológica rara. Paciente masculino, atendido no serviço com queixa de proptose do olho direito (OD) desde nascimento, com progressão nos últimos meses associada à dor. Melhor acuidade visual corrigida de conta dedos a 50 cm do OD. Olho esquerdo sem anormalidades. Ressonância Magnética de OD demonstrou formação expansiva cística de limites definidos em situação intraconal em órbita direita, em íntima relação com nervo óptico, determinando compressão, deformidade e deslocamento anterior do bulbo ocular, além de apresentar sinal semelhante ao líquor em todas as sequências obtidas. Suscitou-se hipótese diagnóstica de meningocele da bainha do nervo óptico direito e o paciente foi encaminhado para cirurgia descompressiva.


**Descritores**: Meningocele/diagnóstico; Nervo óptico/patologia; Neoplasias; Exoftalmia; Relatos de casos

[1]Hospital Visão, João Pessoa (PB), Brasil.
[2]Centro de Referência Oftalmológica do Hospital Universitário Lauro Wanderley, Universidade Federal da Paraíba, João Pessoa (PB), Brasil.
[3]Centro de Estudos Oftalmológicos Renato Ambrósio (CEORA) do Hospital Oftalmológico de Brasília (HOB), Brasília (DF), Brasil.
[4]Fundação Altino Ventura (FAV), Recife (PE), Brasil

**The authors declare no conflicts of interest**

Received for publication 09/07/2012 - Accepted for publication 31/03/2013

## Introduction

Optic nerve sheath meningocele (ONAM) is an extremely rare condition, with few cases reported in the literature, being it defined as a sheath swelling of the optic nerve by spinal fluid. There is an expansion of the cerebrospinal fluid space around the optic nerve without associated inflammation, cerebral or orbital neoplasia. Imaging exams show a tubular-cystic enlargement of the optic nerve / sheath-optical complex with thickening of the optic nerve. The differential diagnosis includes tumors of the optic nerve as meningioma, vascular hamartoma, glioma, neurofibromatosis, Von Hippel-Lindau desease, hemangioendothelioma, or skull-orbital fracture[1].

The symptoms are often related to nerve impairment, classically causing headaches or progressive, slow or fast visual decline[2].

This paper aims at reporting the case of a patient whose clinical data and imaging exams are highly suggestive of optic nerve sheath meningocele, and stressing the main features of this rare disease.

## Case Report

Male patient, 28, attended at the Lauro Wanderley University Hospital complaining of congenital exophthalmos in the right eye (RE) in progress during the last months associated with pain in sporadic twinge. Background: 28 weeks of pre-term, delayed neuro-psycho-motor development. Inspection: Proptosis of the RE (Figure 1). Visual acuity (VA): RE: Counting fingers at 50 cm and 20/20 (1.0) LE (-4.50 D -1.00D 170º). Bio: RE: Cortical cataract 2+/4; LE with no abnormalities. Fundoscopy: RE: Posterior pole choroidal with significant optic atrophy and wide area of atrophy of the Retinal Pigment Epithelium associated to the hyperpigmentation area adjacent to the optical disc. LE: Excavation/cup relationship increased, characteristic macular color, vascular arcades of usual conformity and applied retina. IOP: RE: impracticable, LE: 12mmHg.

RE ultrasound showed cystic, retro orbital lesion in the optic nerve topography, with other exams being asked. Computed tomography (CT) showed the ventricular system with normal dimensions, morphology and topography, basal cisterns of normal aspect, brain structures with normal radiographic density, observing fusiform thickening of the right optic nerve associated to ipsilateral exophthalmos, with no changes in the optical channel or the intracranial route of the optic nerve. Nuclear magnetic resonance (NMR) in the RE showed expansive formation of cystic aspect with defined limits and thin walls located in intrachoanal situation in the right orbit, with close anatomical relation with the topography of the optic nerve, determining anterior compression, deformity and displacement of the ipsilateral eye bulb, besides showing a signal similar to CSF in all sequences obtained, with size of 3.4 x 3.2 x 3.1 cm (Figures 2, 3 and 4).

There was no significant anomalous impregnation of the paramagnetic contrast (GADOLINIUM) in the walls or within the lesion. The extraocular muscles were displaced by said formation, determining compression, deformity and proptosis of the RE eye bulb. Being it very characteristic, the first diagnostic hypothesis raised was rioght optic nerve sheath meningocele.

The patient was then referred to decompression surgery, returning eight months after surgery with no complaints of pain or headache, however reporting persistence of low visual acuity in the RE. The physical examination of the RE showed significant improvement in ocular proptosis, but with persistence of corrected visual acuity of hand movements, esotrope, direct photomotor reflex and consensual absent; onset of conjunctival and corneal lesion, superior temporal quadrant of the cornea, extending until close to the visual axis (Figure 5). The pathology report showed that the withdrawal piece was a "typical fibroadipous hyaline tissue without epithelial lining, compatible with benign cystic lesion wall."

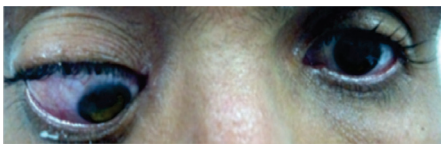

**Figure 1:** Patient in initial eye exam during inspection, demonstrating important ocular proptosis in the RE

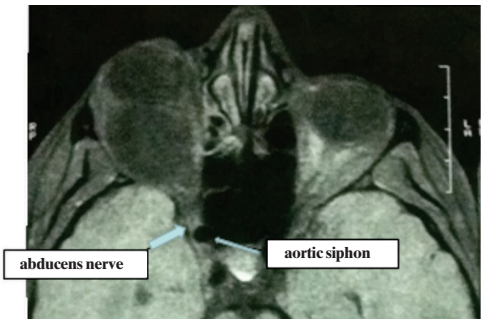


**Figure 2:** Pre-contrast axial section weighted in T1 showing isosignal of mass lesion, leading to proptosis of the RE

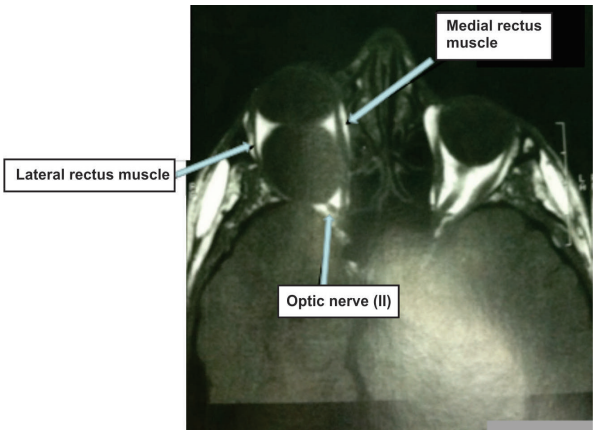


**Figure 3:** . Axial MRI, heavy sequence in T1 with fat saturation. Lesion with low signal after the use of paramagnetic contrast, highlighting significant dilation of the optic nerve sheath, with consequent proptosis of the RE.

## Discussion

According to Garrity et al. [2] the pathology in question arises from the perineural subarachnoid space of the optic nerve, and should be differentiated from congenital cerebrospinal fluid accumulation (CSF) in the perioptic subarachnoid space. In such case, the lesion arises from an abnormal CSF flow through a diverticulum in the arachnoid membrane. They also report the cases of 13 patients with MBNO, with blurred vision and headache being the main symptoms.

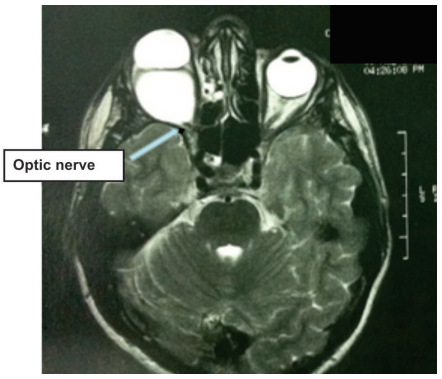


**Figure 4:** Axial section, heavy sequence in T2 with fat saturation. Lesion with high signal, highlighting significant dilation of the optic nerve sheath.

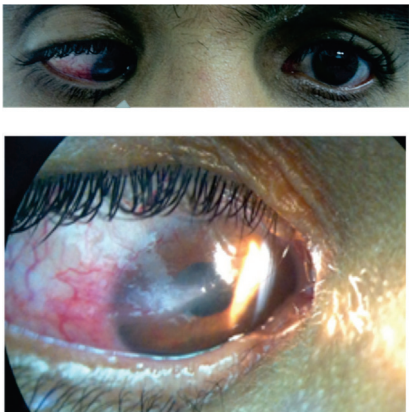

**Figure 5:** Patient in physical exam after surgery, in inspection to a greater lower increase, showing significant reduction in the proptosis of the RE

Lunardi et al. [1] reviewed the existing literature, finding only about 31 cases of MBNO throughout literature, and they describe the disease as primary cysts of the optic nerve sheath without apical mass or malformation of the skull-orbital junction. The symptoms are often related to the involvement of the optic nerve, with a slow or rapid decrease in visual acuity. Additional tests such as CT and MRI revealed a tubular-cystic enlargement of the optic nerve sheath, with thickening of the same. The authors suggest that early surgical intervention by means of the optic nerve sheath decompression enables an improvement of visual function with minimal morbidity in patients with a rapid decrease of visual acuity within 3-6 months.

Shanmuganathan et al. reported the case of an adult, 59, with symmetrical bilateral proptosis and complaint of progressive loss of visual acuity associated to retro-orbital pain and subsequently to cystoid macular edema (CME) in the RE. MRI showed bilateral optic nerve sheath dilation and enlargement, but with normal-sized optic nerve. They describe that EMC can be caused by a tractional meningocele force on the eyeball, and that the role of increased intracranial pressure (ICP) is not clear yet in the etiology of the disease, as only some of the reported cases had increased ICP, measured by lumbar puncture. They also concluded that early surgical decompression of the optic nerve sheath should always be considered in cases of progressive visual loss.

Mesa-Gutiérrez et al. reported the case of a patient, 53, with MBNO associated to intracranial hypertension, but with visual acuity corrected to 20/20 (1.0), with +2.00D in the initial exam. The diagnosis was confirmed only with imaging studies. The MRI identified a dilation of the optic nerve sheath caused by a fluid with characteristics consistent with cerebrospinal fluid. There were no changes in the optic nerves, as well as there was also no evidence of brain or orbit tumors. The lumbar puncture was performed with opening pressure of 22 mmHg, and the chemical analysis of cerebrospinal fluid was normal. After neurosurgical evaluation, the patient was only clinically treated with acetazolamide for three months, remaining stable over the 24 months of follow-up. In all follow-up examinations, the nerve function was normal, with a visual acuity of 20/20 without optical correction after treatment. The authors conclude that surgery should be reserved to severe cases with pain, considerable proptosis, or rapid and progressive decrease in visual acuity.

Spooler et al. [5] describe a case of a child with multiple congenital anomalies, including unilateral MBNO with rapid expansion and displacement of the lateral orbit, resulting in severe cosmetic deformity and complete blindness in his left eye, which was treated with surgical decompression. The authors also rectify that surgical decompression is the standard treatment, with improvement or progression interruption in most cases.

## Conclusion

The present case reports the presence of a retro-orbital lesion in intrachoanal situation and intimately anatomically related to the nerve, strongly suggestive of optic nerve sheath meningocele. The diagnosis can be given by the NMR, and even if pending histological examination, the investigative conduct is consistent, besides the indication of decompressive surgery in selected cases, exceptionally with rapid development and imminent functional and anatomical impairment of the surrounding structures[4]. In conclusion, the importance of remembering that entity is demonstrated by this case report, despite the rarity of the disease.

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