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Frederico Castelo Moura & Ida Fortini

To cite this article: Frederico Castelo Moura & Ida Fortini (2017): Optociliary Shunt Vessels: Role in Diagnosis and Treatment of Atypical Pseudotumor Cerebri, Neuro-Ophthalmology

To link to this article: <http://dx.doi.org/10.1080/01658107.2017.1304967>



Published online: 21 Apr 2017.



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PHOTO ESSAY

Optociliary Shunt Vessels: Role in Diagnosis and Treatment of Atypical Pseudotumor Cerebri

Frederico Castelo Moura ^a and Ida Fortini^b

^aDepartment of Ophthalmology, School of Medicine, University of São Paulo, São Paulo, Brazil;; ^bDepartment of Neurology, School of Medicine, University of São Paulo, São Paulo, Brazil

ABSTRACT

A 46-year-old man presented with severe visual loss and optic atrophy associated with optociliary shunt vessels. The diagnostic work-up revealed intracranial hypertension and cerebral venous sinus stenosis, with no evidence of previous thrombosis. In view of the severe visual dysfunction, both eyes were submitted to optic nerve sheath fenestration. After surgery, a regression of collateral vessels was observed in both eyes.

ARTICLE HISTORY

Received 7 February 2017
Revised 3 March 2017
Accepted 7 March 2017

KEYWORDS

Intracranial hypertension;
optic nerve sheath
fenestration; optociliary shunt
vessels; pseudotumor cerebri

Case report

A 46-year-old man complained of severe visual loss starting 2 years earlier, originally without headache or eye pain. The first manifestation was transient visual obscuration. This was followed a few months later by loss of the peripheral field, but with preservation of central vision. Six months before the first examination, severe headache set in, associated with reduced central vision. The patient's body mass index (BMI) was 26.5 kg/m².

Upon examination, visual acuity was no light perception in the right eye and hand movements in the temporal field of the left eye. The pupillary light reflex was poor in both eyes. No abnormality was seen on biomicroscopy, and the intraocular pressure was 12 mm Hg in both eyes. The fundus examination revealed atrophic optic nerve and optociliary shunt vessels in both eyes (Figure 1). Manual perimetry could not be performed due to severe visual loss.

Magnetic resonance imaging (MRI) revealed no space-occupying lesion, but flattening of the posterior sclera and partially empty sella were present (Figure 2). Magnetic resonance

angiography (MRA) showed transverse and sigmoid sinus narrowing to the left but no signs of thrombosis (Figure 2). The lumbar puncture opening pressure was 42 cm H₂O, with normal constituents.

Both eyes were submitted to optic nerve sheath fenestration via the medial transconjunctival approach. Five days after surgery, a regression of collateral vessels was observed in both eyes (Figure 1). Visual acuity in the left eye improved to counting fingers at 2 m. The right eye remained at no light perception.

The reported case (a male with BMI <30 kg/m²) illustrates the difficulty of diagnosing atypical pseudotumor cerebri. The absence of headache in the early stages and the presence of optic atrophy (rather than papilloedema) at the time of the first examination made diagnosis even more challenging. The presence of optociliary shunt vessels was the only finding suggesting intracranial hypertension as the cause of optic atrophy (after ruling out optic nerve sheath meningioma and central retinal vein occlusion).¹

Regression of papilloedema is a relevant clinical parameter of treatment efficacy in patients with pseudotumor cerebri. In post-papilloedema

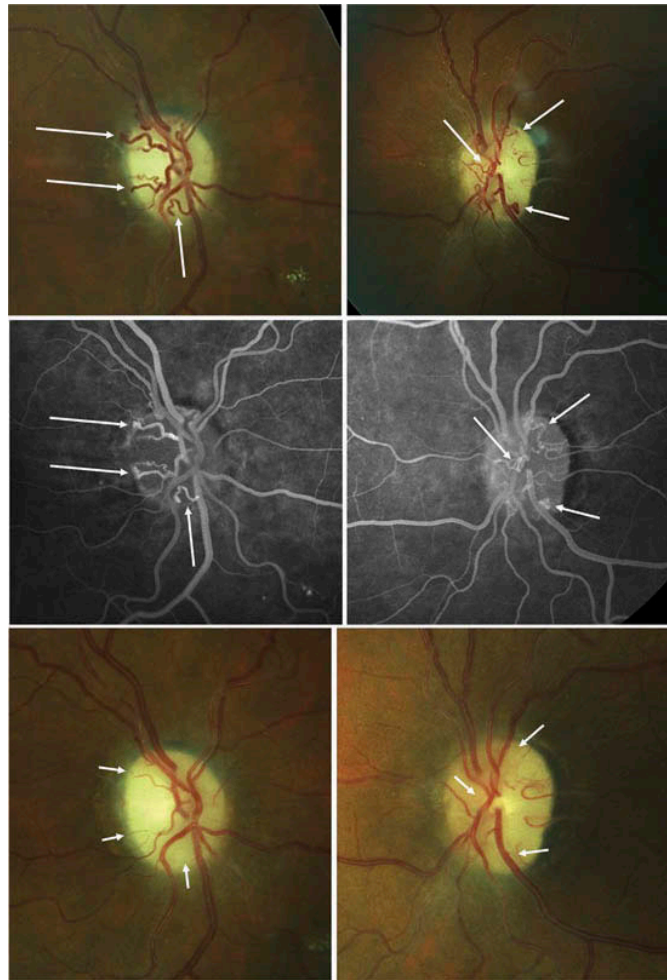


Figure 1. Fundus photographs (top row) and fluorescein angiography (middle row) of the optic nerve, showing optociliary shunt vessels (arrows) prior to optic nerve sheath fenestration (bottom row). After surgery, there was regression of optociliary shunt vessels.

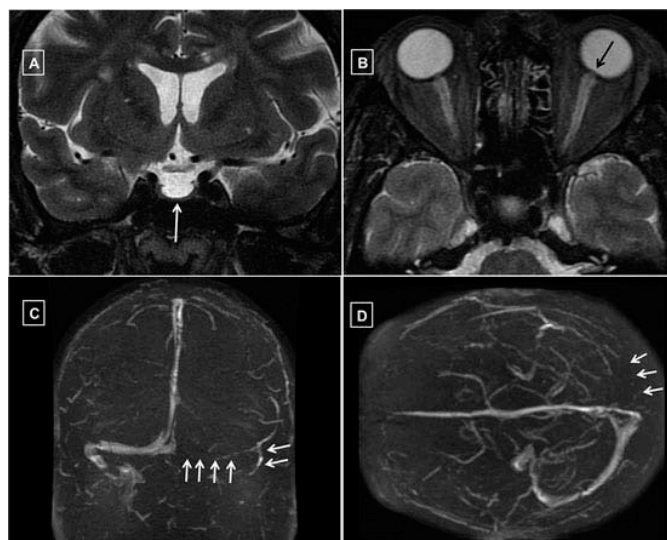


Figure 2. Top row: MRI showing empty sella turcica (a, arrow) and posterior scleral flattening (b, arrow). Bottom row: MRA showing narrowing of the left transverse and sigmoid sinus (c and d, arrows).

atrophy, no optic nerve oedema is observed due to retinal ganglion cell damage; thus, papilloedema is not a useful clinical parameter. The regression of optociliary shunt vessels documented in the present case served as a parameter of successful optic nerve sheath fenestration.²

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the article.

ORCID

Frederico Castelo Moura  <http://orcid.org/0000-0002-7949-6832>

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