

## An Unusual Presentation of Horton's Disease

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## Abstract

Horton's disease or temporal gigantho cellular arteritis is an inflammatory disease of the vessels, particularly affecting older female. We report the case of a 50-year-old patient with a classic clinical picture of anterior arteritic optic neuropathy with ischemia of the optic nerve, with a horizontal level and hypo-choroidal perfusion of the corresponding territory. The peculiarity in this patient is the negativity of the biological assessment and biopsy of the temporal artery.

**Keywords:** Anterior ischemic optic neuropathy, Choroidal ischemia, Blindness/Emergency

## Introduction

Arteritic acute anterior optic neuropathy is a diagnostic and therapeutic emergency because of the risk of bilateral and definitive blindness. It results from an occlusion by thrombosis in the territory of the ophthalmic artery, leading to massive ischemia in PCA vascularized territories. We report the case of an unusual presentation of Horton ischemic optic neuropathy whose clinical is typical but with a negative paraclinical assessment.

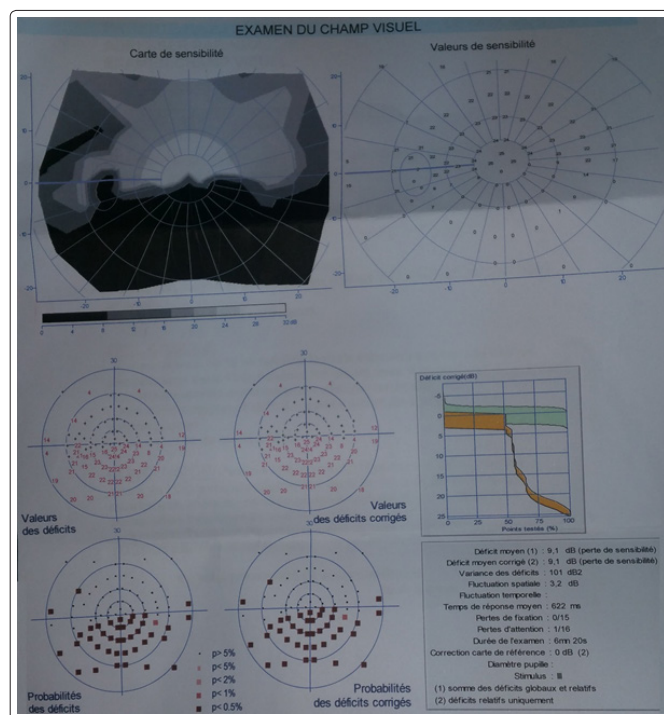
## Observation

A 50-year-old patient, with no particular pathological history, presented to ophthalmic emergencies for a sudden decrease in visual acuity of the left eye, with hyperesthesia of the scalp and intermittent claudications of the jaw. Ophthalmologic examination revealed corrected visual acuity at 7/10 in the left eye (10/10 OD) with relative afferent pupillary deficit. Examination of the visual field by confrontation revealed a lower altitudinal deficit. Examination of the fundus found a papillary edema stage 2 with flame-shaped haemorrhages from the inferior temporal arcade, with signs of choroidal ischemia in the upper choroidal territory (white deep lesions: elschnig spots).

The rest of the clinical examination shows induration in the regard of the temporal artery. Biological emergency assessment revealed a slight increase in CRP with normal sedimentation speed. The patient was placed on a solumedrol bolus for 3 days with adjuvant therapy with an oral relay (1mg/kg/day prednisone).

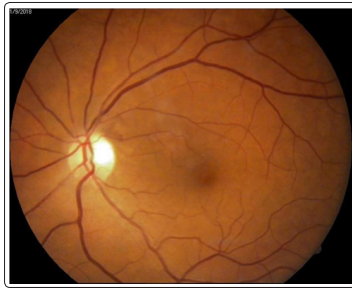
The evolution was marked by a recovery of the visual acuity (10/10Ac), with a regression of the papillary edema and an installation of a lower papillary pallor.

Automated static perimetry revealed a lower altitudinal deficit attached to the blind spot (Figure 1).

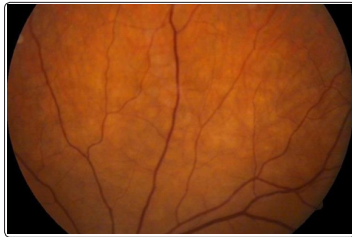


**Figure 1:** Automated visual field 30-2 revealing a lower altitudinal deficit with horizontal level attached to the blind spot

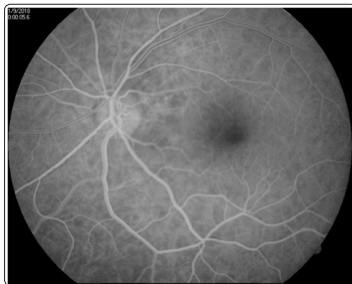
Fluorescein angiography shows sectorial ischemia of the optic nerve (lower papillary pallor) (Figure 2), with hypo choroidal perfusion areas at the upper choroidal territory (Figure 3 and 4) with the presence of pinpoint (Figure 5). Moreover, the rest of the quadrants were without particularity (Figure 6).



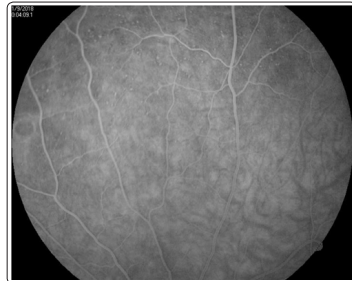
**Figure 2:** Superior papillary pallor with horizontal level, with haemorrhage at the beginning of the superior temporal arcade in resorption after bolus of corticotherapy



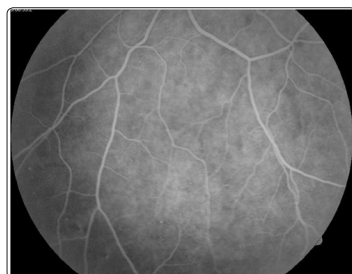
**Figure 3:** Elschnig spots in upper choroidal territory showing choroidal hypofusion



**Figure 4:** Upper sector ischemia of the optic nerve with choroidal filling delay with hypo areas choroidal perfusion



**Figure 5:** Upper choroidal ischemia with diffuse pinpoint



**Figure 6:** Normal perfusion of the lower quadrant

The patient received 10 days later a biopsy of the left superficial temporal artery (Figure 7), whose pathological result was negative.



**Figure 7:** Biopsy of the left superficial temporal artery

### Discussion

Anterior ischemic optic neuropathy (AION) secondary to Horton's disease is due to acute ischemia of the optic nerve head by occlusion of the posterior ciliary arteries [1].

In front of clinical suggestive signs, inflammatory biological examination can guide the etiological diagnosis especially the pair SS and CRP which is increased (Note that the CRP is more sensitive) as well as the palpation of the temporal pulse [2].

Automated perimetry must be systematically in the acute phase, and allows to objectify a fascicular scotoma corresponding to all fiber vascularized by the short PCA. This scotoma is characterized by its altitudinal character and attached to the spot of Mariotte [2].

Fluorescein angiography allows visualization of a massive choroidal filling defect in the territory of the injured short PCA [3].

The diagnosis is histological through the temporal artery biopsy (>2cm) which can be negative in 5 to 15% of cases, but which must not in any case delay the therapeutic management, rather it allows to justify treatment with long-term corticosteroid [4]. The risk of negative biopsy is relatively low (<15%) even after 4 weeks of corticosteroid therapy [5].

Untreated, bilateralization is about 90% after 1 week of evolution. The treatment is based on the urgent systemic administration of 3 boluses of solumedrol and an oral relay of prednisolone (1mg/kg/day) [6]. The decrease doses and duration are judged on the regression of clinical and biological signs (SS, CRP).

This treatment is usually continued for at least 2 years with the measures associated with any prolonged corticosteroid treatment [7].

### Conclusion

The NOIA following Horton's disease is a diagnostic and therapeutic emergency because of the increased risk of bilateralisation.

The goal of treatment is the perfect control of the disease, mainly because of a high risk (8-18%) of amaurosis relapse during steroid decay. The low-dose aspirin would have a preventive effect on visual impairment and cerebrovascular disease Horton [8].

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