

## Case report

## Clinical and pathological correlation of cotton wool spots in secondary angle closure glaucoma

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## ABSTRACT

**Purpose:** Cotton Wool Spots (CWS) are a commonly described retinal finding in the posterior segment associated with an extensive number of systemic diseases. The appearance of a CWS in the setting of glaucoma has rarely been reported and has not been correlated with pathology to localized loss of the nerve fiber layer previously. In this case report, we augment a previous report of an 18 year old female with a diagnosis of low grade ciliary body melanoma. This patient experienced eventual mechanical angle closure with a CWS appearing in the posterior pole in the setting of acute elevation of intraocular pressure (IOP). This eye underwent enucleation and pathology evaluation.

**Observations:** Fundus photography documented a CWS in the posterior segment during a period of acute elevation in IOP. Subsequently the eye was enucleated due to pain from refractory angle closure glaucoma secondary to low grade iris-ciliary body ring melanoma. The specific site of the prior CWS was studied with 1 $\mu$  Epon retinal step sections stained with a novel AgNO<sub>3</sub> solution. Light microscopy demonstrated a retinal nerve fiber layer scar and inner nuclear layer collapse in the prior location of the CWS. Light microscopy and transmission electron microscopy shortly after enucleation had demonstrated temporal quadrant laminar optic nerve (ON) retrograde axonal transport block.

**Conclusions and Importance:** Although not commonly associated with glaucoma, CWS can present in the setting of acute elevations of IOP and may be associated with loss of nerve fiber layer. This loss of nerve fiber layer can confound the ability to judge glaucoma progression based on nerve fiber layer thickness via optical coherence tomography and changes in disc contours. Patient care may benefit from care provider's awareness of this possible phenomenon in the setting of angle closure.

## 1. Introduction

This case report extended from the original report, to our knowledge, is the first to provide histopathological co-localization of retinal damage corresponding to a long faded CWS in angle closure glaucoma.<sup>1</sup> Cotton wool spots (CWS) are a well-described retinal finding occurring in association with many different well understood and idiopathic systemic retinal-vascular diseases, most commonly diabetes and hypertension.<sup>2</sup> Typical CWS appear as transient gray to white areas with indistinct borders in the retinal nerve fiber layer (RNFL) usually less than 1/3 disc diameter, often adjacent to small retinal vessels. CWS contain clusters of cytooid bodies which form due to focal obstruction of orthograde or retrograde axonal transport in retinal ganglion cell axons.<sup>2–4</sup> CWS usually fade over a few weeks following initial presentation and have only recently been associated with lasting retinal or disc damage.<sup>5–7</sup> Most CWS will appear in the posterior fundus without

any loss of visual acuity unless the central macula is involved. Traditional assumptions held that following the resolution of a CWS there were no pathological sequelae. Subsequent investigation with scanning laser ophthalmoscopy has shown localized loss of nerve fiber layer thickness corresponding with prior CWS<sup>5–7</sup>. Therefore, CWS are not as innocuous as previously assumed. The incidence of CWS has rarely been reported in association with glaucoma<sup>(3)</sup> and subsequent co-localized pathologic examination following resolution of CWS has not previously been reported.

## 2. Case report

An 18 year old female was diagnosed with unilateral secondary angle closure as a result of ring melanoma of the ciliary body OS, detailed in a previous report<sup>(1)</sup>. She was initially followed for iris abnormalities that were misdiagnosed initially as iridocorneal endothelial

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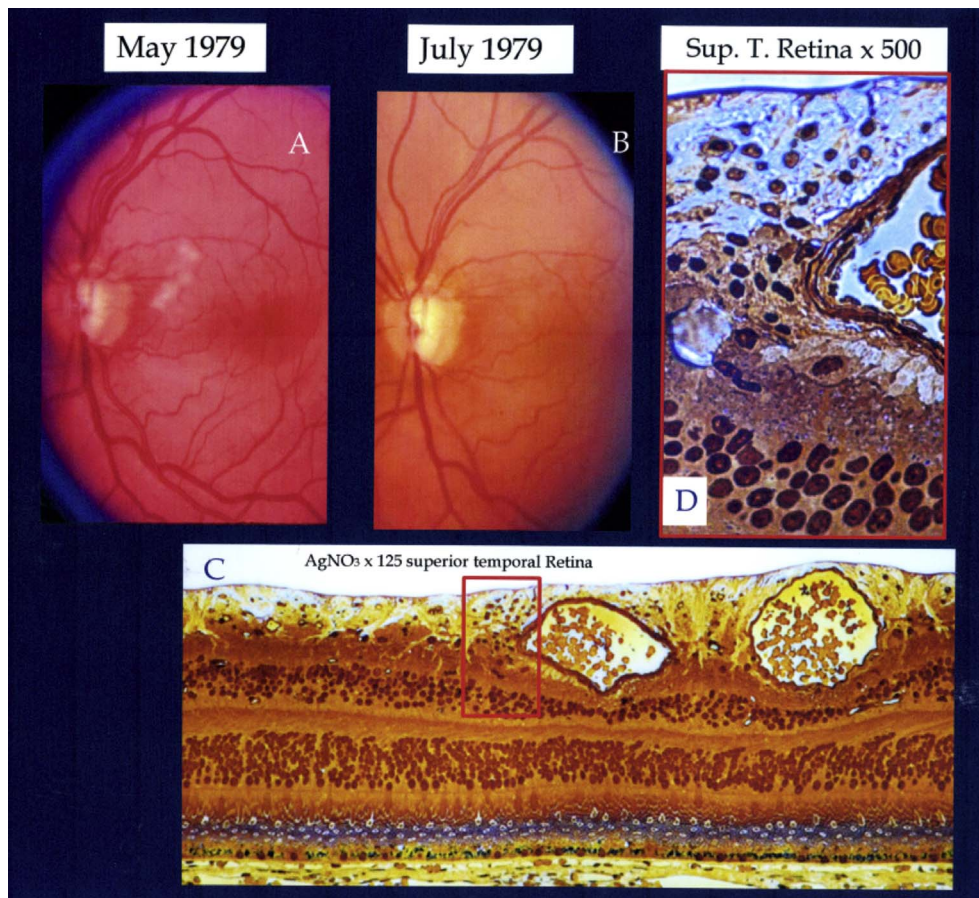


Fig. 1. Fundus Photo OS during and after resolution of cotton wool spots and correlating histopathology from superior temporal macula. Relevant findings include a collection of glial cells in the nerve fiber layer and partial collapse of the middle nuclear layer. AgNO<sub>3</sub>, Epon 1 $\mu$  section X 27.5 & 125.

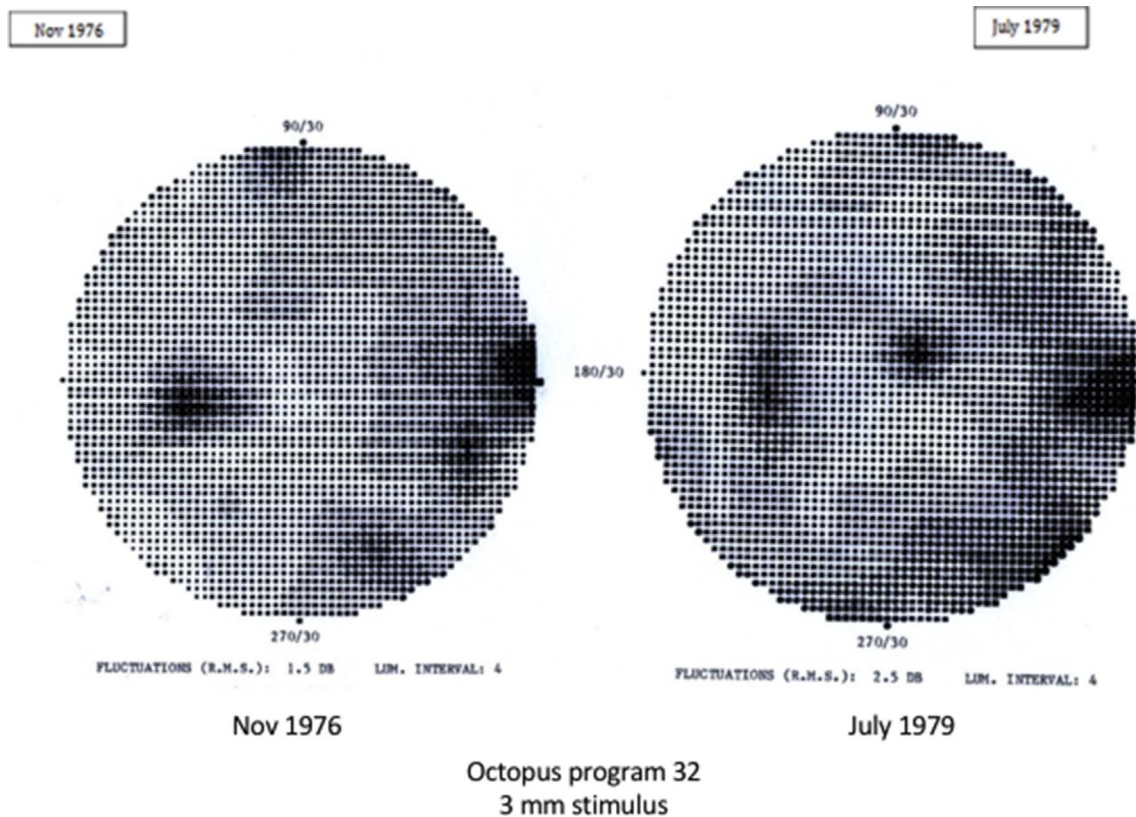


Fig. 2. An Octopus VF in Nov 1976 OS revealed an early nasal step clearly progressed by August 1979.

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