CLINICAL CASE - PHOTO ESSAY

MELANOCYTOMA OF THE OPTIC NERVE HEAD

VAN WINDEN M., AL -SABAY N*, SALU P**

CASE REPORT

A 42 year-old male of North-African origin consulted our outpatient clinic with complaints of presbyopia. On routine fundoscopy, he was found to have a dark, elevated lesion at and around the optic nerve in his left eye with an extension to the nasal chorioretina. We had the opportunity to follow this patient on about eight years. His best corrected visual acuity was 1.0 on both eyes and revealed to be unchanged during our follow-up. On biomicroscopy, we found a normal anterior segment in both eyes, without melanocytosis of the conjunctiva. Fundoscopy demonstrated a dark-brown to black lesion located in the optic disc and extending over the nasal margin involving the adjacent choroid. Colour (Figures 1a and 1b), green monochromatic fundus photographies and recordings were performed. Fluorescein angiography showed a typical important hypofluorescence in the area of the elevated mass (1). No changes have been observed on fundoscopy and fluoangiography since the beginning of our observation. A-Mode and B-Mode scan echography (Sonomed/Simovision - B 10 MHz) showed a mildly elevated and highly reflective prominent mass in the optic disc without intraneural extension (2-3).

Concerning the involvement of the adjacent chorioretina on the nasal side, we did not observe any choroïdal excavation which is well known to be a typical sign for a melanoma. Nor there was any sign of decline in reflectivity. Between 2006 and 2009, the echographic examination of the lesion showed neither growth nor change of the prominence (Figures 2a and

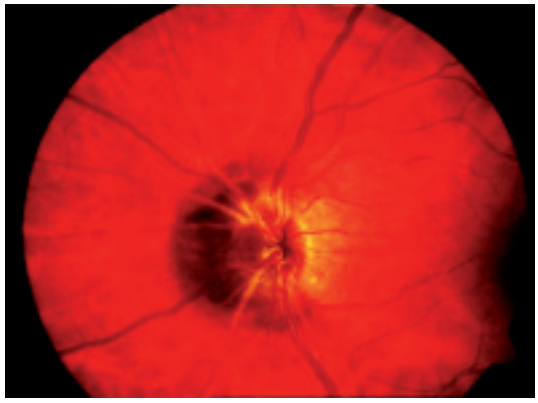

Fig. 1a: Color fundus photography of the left optic disc in 2002 showing a dark pigmented lesion.

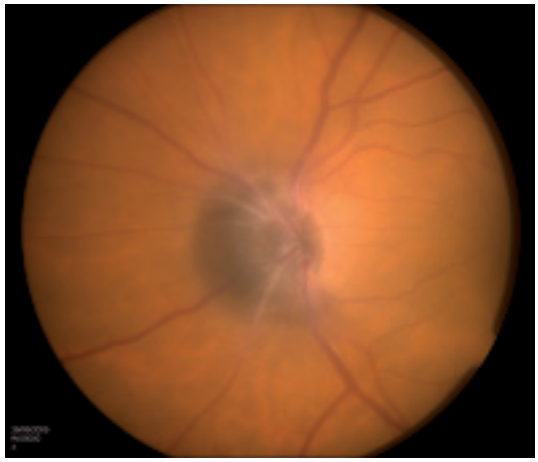

Fig. 1b: Color fundus photography of the left optic disc in 2009.

.....

* *Departement Oogheelkunde, UZ Brussel, Vrije Universiteit Brussel, VUB*

Submitted: 19-05-09
Accepted: 7-08-09

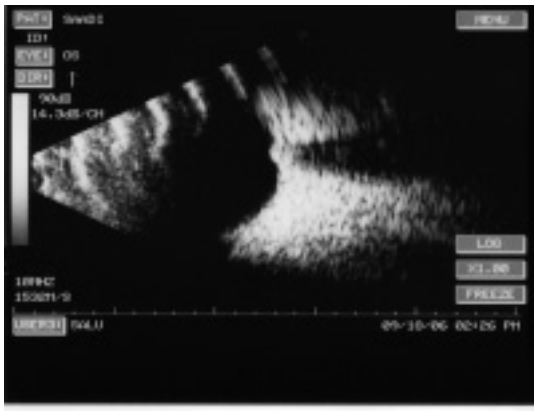

Fig. 2a: B-Mode scan echography with a probe of 10 MHZ showed the highly reflective prominence of the left optic disc, without any posterior extension the optic nerve in 2006

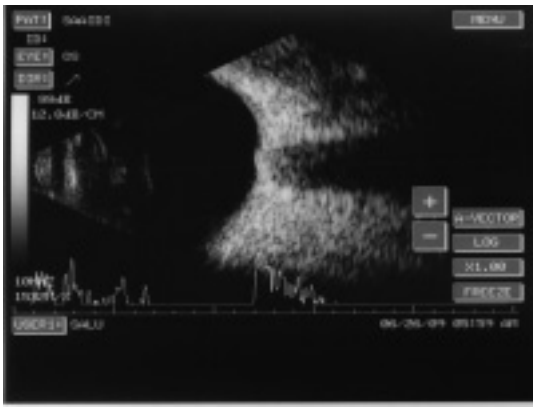

Fig. 2b: A-Mode and B-Mode scan echography in axial scan prominence of the left optic disc staying stationary in: 2009. The A-scan shows sound attenuation in the optic nerve.

2 b).The OCT (Stratus type III OCT) displayed a dome-shaped lesion with a gradual slope at the tumour margin. It demonstrated an important anterior extension of the lesion with a single band of hyperreflectivity (Fig.3). Dense posterior shadowing obscured all optical details of the optic nerve, as previously mentioned by Chaudhary et al in 2006 (1) and Shields et al in 2008 (4). This empty space behind the high reflectance is due to the blockage of the light-transmission through the deeply pigmented tumour. No adjacent retinal oedema was observed. On the basis of the OCT data, the retinal nerve fibre layer (RNFL) showed an important axonal fibre loss at the site of the lesion on the affected eye. This pathological thinning is the result of an important axonal loss on the nasal quadrant with a superior and inferior extension due to a chronic compression of the optic nerve head.

From the start of our observation, the Humphrey Visual field examination (VF) (Central 30-2 threshold test, stimulus III) had revealed an enlarged blind spot expanding superiorly and inferiorly as well as a concentric shrinking nasally (Figures 4a and 4b). On Goldmann kinetic perimetry (isopter V-4 and III-3) testing, we saw a similar abnormality, principally with III-3, which revealed a similarly enlarged blind

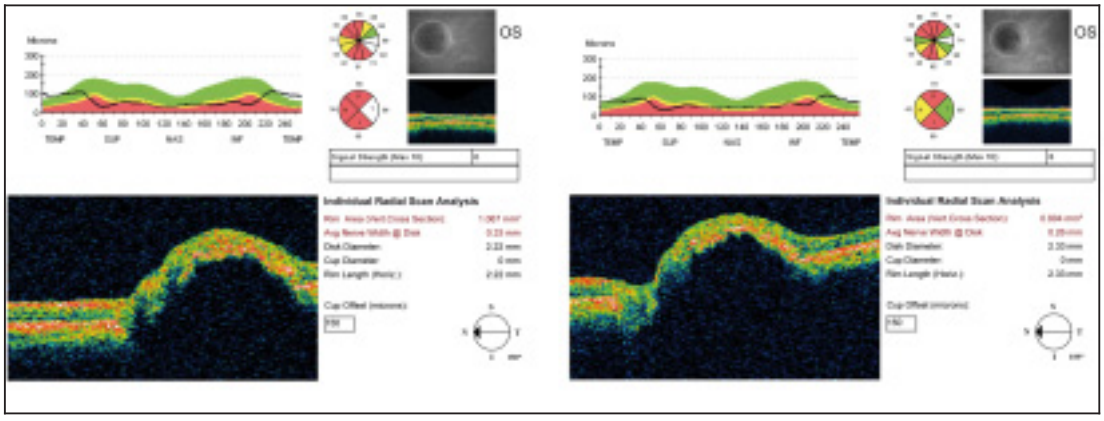

Fig. 3: The OCT (Stratus type III OCT) of the left optic nerve head in 2006 (on the left side) compared with the scan performed in 2009 (on the right side) showing a high reflectance layer with dense posterior shadowing.

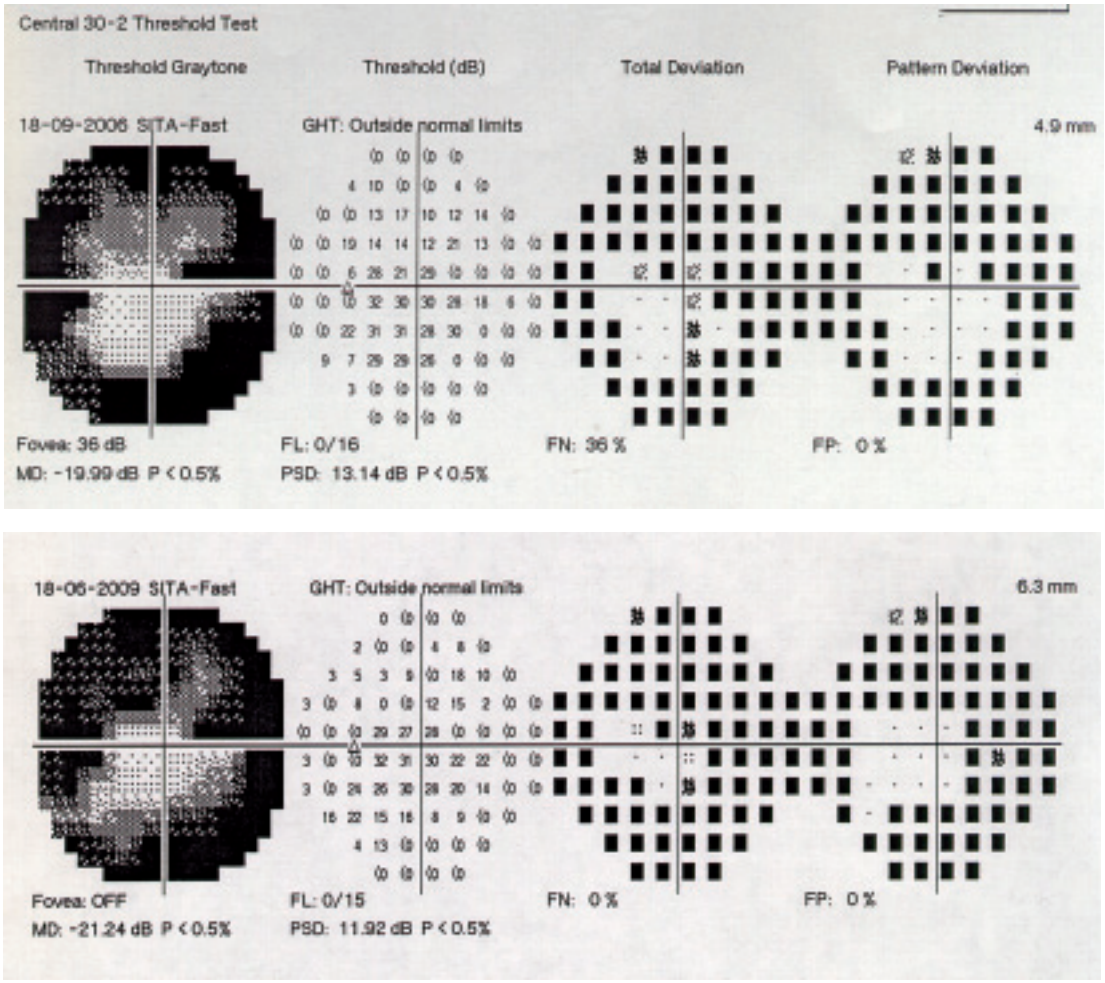

Fig. 4a and 4b: Humphrey visual field 2006 and 2009 showed an enlarged blind spot expanding superiorly and inferiorly as well as a concentric shrinking.

spot and a peripheral concentric limitation of the nasal field (Figures 5a and 5b). No further damage was noticed during our observation. The OCT indicated a damage of the RNFL nasally, which was translated into an important loss of the nasal visual field. The limits of the temporal visual field were preserved, except for the enlarged blind spot.

Magnetic Resonance Imaging (MRI) study of the optic nerve, performed in 2006, did not show a hyperintense infiltration in the central portion on T_1-weighted images (5). We could conclude that there was no evidence of retrobulbar tumoral invasion of the optic nerve.

Concerning the tests showing any abnormality such as visual field, echography and OCT, we remarkably did not observe any further deterioration during our follow-up. On the basis of these findings, we could substantiate that, in our patient, the melanocytoma existed before the start of our observation, that it remained stable and did not evolve into a melanoma.

DISCUSSION

Melanocytoma of the optic nerve head is a rare, distinctive, deeply pigmented benign tumour, frequently (in 54 % of the cases) involving the adjacent retina and choroids (2-6). Histologi-

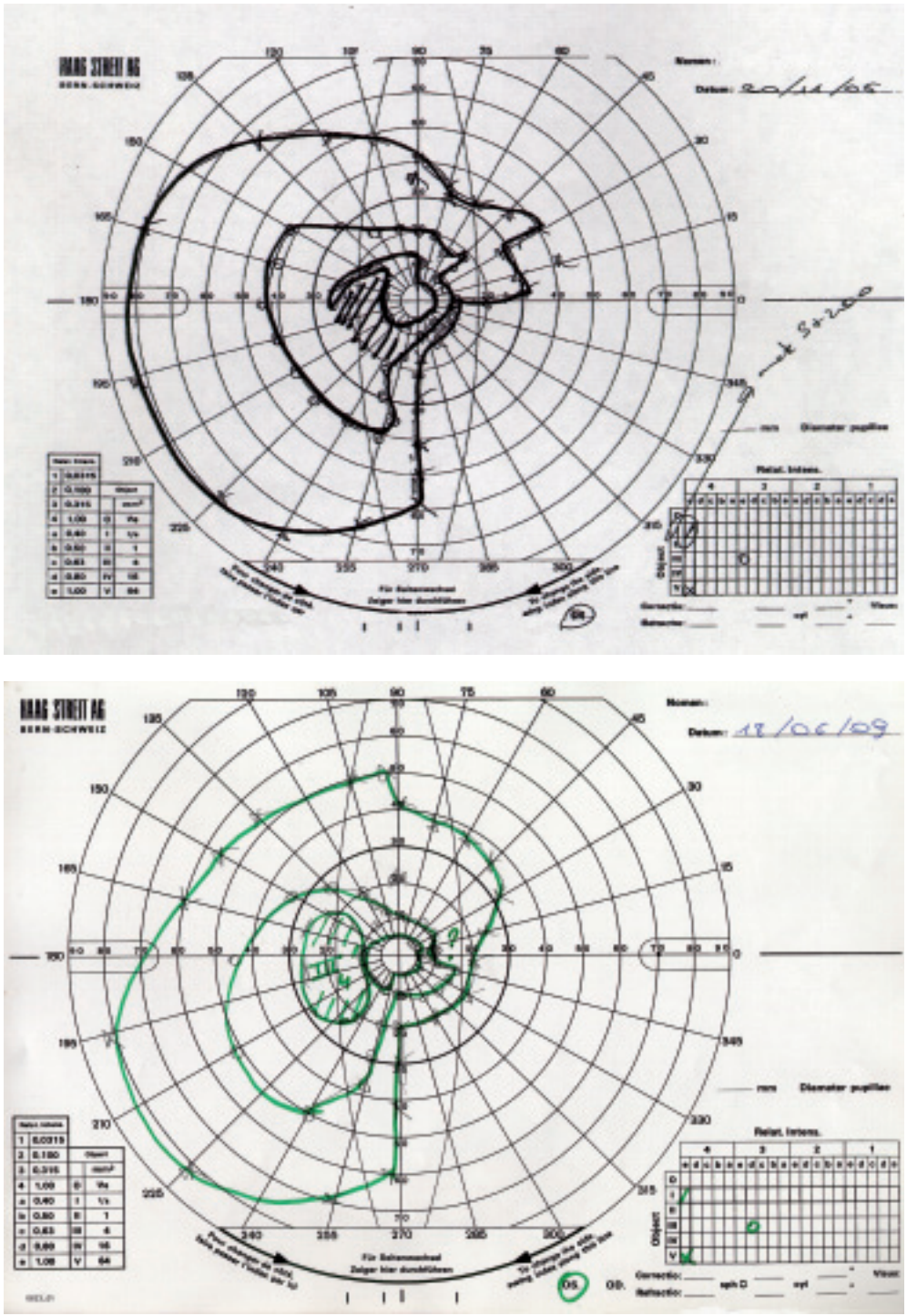

Fig. 5a and 5b: Goldmann visual field 2006 and 2009 showing an enlarged blind spot and a peripheral concentric limitation of the nasal field.

cally, a melanocytoma is a magnocellular naevus with round to oval cells which occurs more commonly in dard-skinned individuals (7). Only 1 to 2 % of the cases show a tranformation to malignity (6). A yearly follow-up is indicated with an examination consisting in visual acuity, visual field, fundoscopic examination and photography and if possible echography.

Any abnormality or change in the aspect of the findings or any decrease in the visual acuity can be the first sign of evolution to malignity. It is most important to keep in mind the criteria for transformation to a malign melanoma during follow-up, as described by Shields et al (6). Growth, extensive involvement of the optic nerve and severe visual loss are suggestive for this malignant transformation. Changes in visual acuity in patients with melanocytoma can vary from mostly non-significant visual impairment to slight visual loss caused by retinal and foveal exudation due to neuroretinitis. On the other hand, severe visual loss is frequently related to central vein occlusion due to compression and spontaneous tumour necrosis. This can be considered as an important sign of malignant transformation. It is also important to emphasize that a mild growth or a subtle enlargement of the lesion over several years should not be considered as a sign of malignant transformation since it occurs in 10 to 15 % of the cases (6). When a dubious change occurs, MRI is indicated to investigate any malign infiltration into the optic nerve. Treatment is only requested when the rare event of malignant transformation occurs. Once the diagnostis of malignancy has been made, no other treatment but enucleation is available.

REFERENCES

(1) CHAUDHARY R., ARORA R., METHA D.K. et al. – Optical Coherence Tomography Study of Optic Disc Melanocytoma - Case Report. *Ophthalmic Surg Lasers Imaging 2006; 37(1):58-61.*

(2) SZABO A., DEGI R., KOLOZSVARI L. – Echographic examination of melanocytoma of the right optic nerve head. Ophthalmic Echography - Proceedings of the 20th SIDUO Congress, Budapest, Hungary 2004: 274-276.

(3) BYRNE S.F., GRENN R.L. – Ultrasound of the eye and orbit. Second edition Mosby 2002; 115-122, 151-152.

(4) SHIELDS C.L., PEREZ B. BENAVIDES R., MATERIN M.A., SHIELDS J.A. – Optical coherence tomography of optic disc melanocytoma in 15 cases. *Retina 2008 ; 28:441-446*

(5) DE POTTER P., SHIELDS C.L., EAGLE R.C. et al. – Malignant Melanoma of the Optic Nerve. Arch ophthalmol 1996 ; 114 :608-612.

(6) SHIELDS J.A., DEMERCI H., MASHAYEKHI A. et al – Melanocytoma of the optic disc . A review. *Survey of Ophthalmology 2006;51:93-104.*

(7) KANSKI J.J. – Clinical Ophthalmology. A systemic approach. 6th edition. Butterworth-Heinemann Elsevier. 2007:540.

.....

Correspondence and reprints:
Dr. M. Van Winden
UZ Brussel, Vrije Universiteit Brussel
Departement Oogheelkunde
Laarbeeklaan 101, 1090 Brussel
Belgium.
E-mail: mariannevanwinden@telenet.be