Documented growth of a halo choroidal nevus

Croissance documentée d'un naevus choroïdien avec halo

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A 54-year old man with no past medical history except hyperactivity disorder presented with a 12-week history of right blurred vision. On examination his right visual acuity was reduced to 6/9 compared to 6/5 in the left eye. In the right fundus there was a halo choroidal nevus abutting the fovea at the posterior pole (Fig.1A) measuring 4.5 by 5 mm in diameter with a 1.6 mm thickness (sclera not included) and no evidence of hollowness on B scan ultrasound (Fig. 2G). Subretinal fluid was present tracking down into the subfoveal space on OCT (optical coherence tomography) (Fig. 2A) Fundus autofluorescence (FAF) indicated lack of lipofuscin (Fig. 2C). Examination of the left eye showed an iris nevus with no suspicious features. Fluorescein angiogram of the right fundus showed diffuse punctuate slow leakage from retinal pigment epithelium degeneration overlying the choroidal nevus (Fig. 2D). Photodynamic therapy (PDT) was therefore performed for symptomatic relief of a presumed leaky naevus. Two months after PDT visual acuity was 6/5 in both eyes, the subretinal fluid had resolved on OCT (Fig. 2B) and retinal pigment epithelium changes were observed (Fig 1B).

Thirty months later, he presented again with right visual loss. Visual acuity was 6/6 in the right eye and 6/5 in the left eye. The halo choroidal indeterminate melanocytic lesion now measured 6.9 by 9.2 mm in diameter and was elevated to 1.8 mm with an acoustic hollowness on B scan ultrasound (Fig. 2H). There was an increase in the size of the lesion on fundus examination with an increase in the pigmented central area corresponding to the nevus and marked increase of the surrounding halo and orange pigment overlying the lesion (Fig. 1C). On the OCT, subretinal fluid and increase of the size is observed (Fig 2E-F). Based on the documented growth since the first visit the clinical diagnosis of small choroidal melanoma was made and treatment consisted of brachytherapy with Ruthenium 106 plaque. Baseline liver ultrasound scan was within normal limits.

Discussion

Halo nevus can be found in the skin, also called Sutton's nevus or leukoderma acquisitum centrifugum, or in the choroid. Clinically these lesions have a pigmented central area surrounded by a yellow depigmented halo, but the reverse is also possible [1]. The histology of choroidal halo nevi has not been reported, because these benign lesions are not an indication for fine needle aspiration biopsy (FNAB) or enucleation. However, in the cutaneous halo naevi, the halo corresponds to an inflammatory infiltrate made by lymphocytes and histiocytes with decreased to absent melanocytes and melanin. This is in favour of an immune-mediated destruction of melanocytes [2]. The presence of halo-shaped depigmentation correlates with the onset of cutaneous nevus regression [2] but complete involution can take several years [3].

Mean age at diagnosis of halo choroidal nevi is 52 years [1], whereas halo cutaneous nevi are more common in children and young adults [2]. The prevalence was found to be 8% in middle-aged adults and only 2% in young patients [4]. The absence of a halo around a choroidal nevus has been associated with an increased risk of transformation into melanoma [5]. However, the presence of the halo is not a guarantee that malignant transformation will not take place. In the literature just few cases with documented growth of a halo choroidal nevus into melanoma have been reported [1,6]. In one patient growth occurred outside the halo 4 years after the initial diagnosis of nevus in a 60-year-old woman [6]. In a series of 150 patients with halo choroidal nevi growth into melanoma was detected in only 4 cases at a mean interval of 44 months; at presentation the mean tumour dimensions had been 6.8 mm in diameter and 2.2 mm in thickness [1]. Growth is more likely to occur if the halo choroidal

nevus is thicker, closer to the optic disc, acoustically hollow on ultrasonography, symptomatic and the presence of subretinal fluid and/or orange pigment [1]. These lesions should therefore be monitored long term (3 monthly to yearly follow-up, depending on the degree of suspicion) and the presence of a halo should not be interpreted as a sign of that is always benign.

In cutaneous naevi, changes in halo diameter are not always associated with malignant transformation [7], but a halo may rarely be associated with skin melanoma [8,9], especially when the lesions develop outside childhood or adolescence [10]. The significance of halo growth in choroidal naevi is not well understood. Close observation or treatment are therefore both options for management in this situation. In this case, as there was growth over a relatively short period of both the pigmented and depigmented parts of the choroidal lesion, definitive treatment for a small melanoma was performed. Primary transpupillary thermotherapy (TTT) and PDT should be avoided in the treatment of choroidal indeterminate melanocytic lesions with documented growth because both have been associated with a significant percentage of further lesion growth after treatment [11, 12]. Radiotherapy, either with proton beam or radioactive plaque, should be the primary treatment of choice.

Conflict of Interest Disclosures

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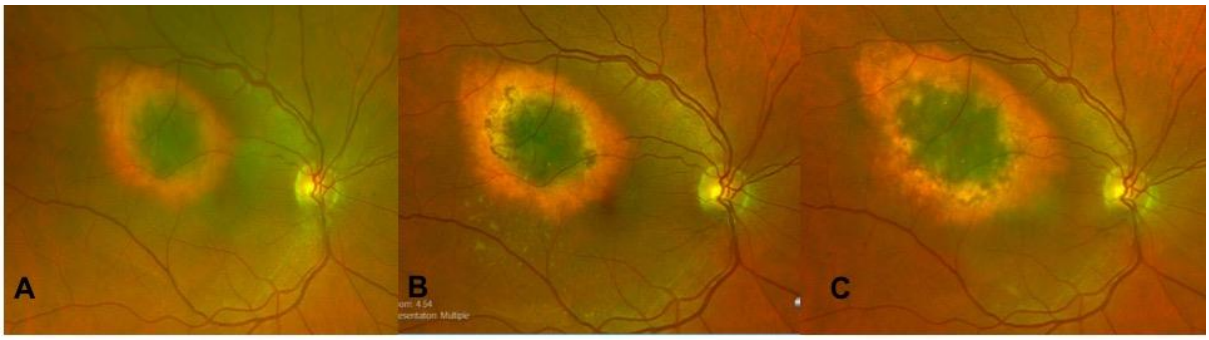
FIGURES

Figure 1

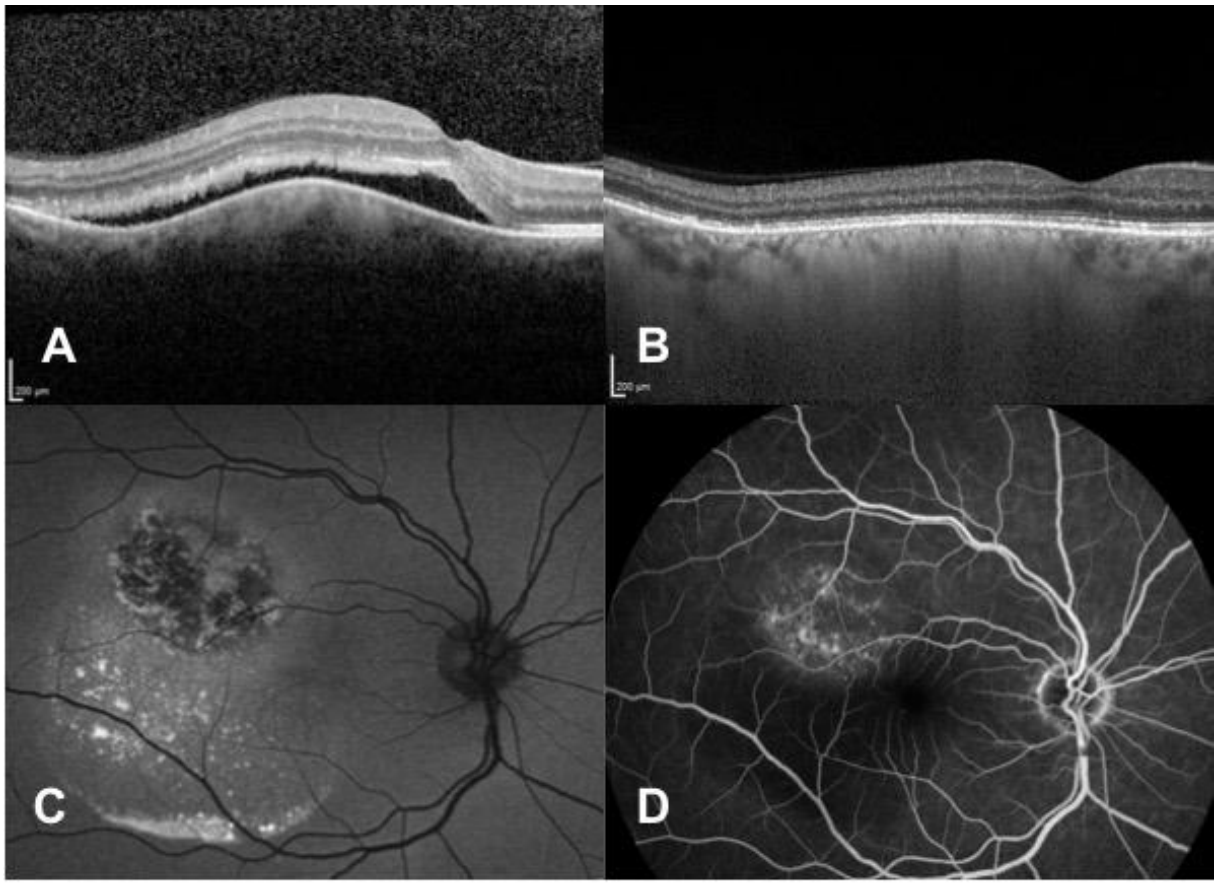


A: right choroidal pigmented mass with a halo, initially diagnosed as a leaking nevus

B: aspect of the lesion after PDT with verteporfin

C: increase in the surface of both the pigmented choroidal lesion and its halo, making the diagnosis of choroidal melanoma most likely

Figure 2



A: macular OCT done on initial presentation showing a choroidal mass with overlying subretinal fluid

B: resolution of the subretinal fluid following treatment with PDT

C: autofluorescence showing the subretinal leakage from the choroidal lesion

D: fluorescein angiogram showing diffuse punctuate hyperfluorescence in the area of the choroidal mass

E and F: increase in the thickness of the choroidal mass compared to the initial OCT scan in figure 2A

G: ocular ultrasound of the choroidal mass on presentation. The tumour was 1.6 mm thick

H: ocular ultrasound showing an increase in tumour thickness to 1.8 mm with acoustic hollowness