CASE REPORT

Polypoidal lesions associated to choroidal naevus: spectrum of pachychoroid disease?

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SUMMARY

A 78-year-old woman was referred by an optician for an incidental finding of peripapillary naevus in the absence of visual symptoms. On examination, the right eye showed a choroidal naevus of around 1.5 disc diameter, adjacent to the optic disc. Optical coherence tomography detected the presence of pigment epithelium detachment and subretinal fluid within the context of the naevus. Fundus fluorescein angiography and indocyanine green showed multiple polypoidal lesions (Polypoidal Choroidal Vasculopathy, PCV) in the choroid within the spectrum of a pachychoroid, without any sign of malignancy related to neovascularisation. The patient underwent a follow-up of 6 months, showing stability of the lesions over this timeframe. No treatment has been required since the polypoidal lesions were outside the fovea and visual acuity remained stable. The association between choroidal naevi and polypoidal lesions is rare¹; however, this seems to confirm that these findings are part of the spectrum of pachychoroid disease.

BACKGROUND

Despite the choroidal naevus being a common finding on the retina,¹ the association with PCV is rarely described. Few cases in the literature show that whenever this association has been found, the choroid has a particular aspect of pachychoroid, proving that the naevus can influence the choroidal haemodynamics. A thorough clinical assessment is essential in order to differentiate a choroidal neovascularisation (CNV) from a malignancy.

CASE PRESENTATION

A 78-year-old Caucasian woman was referred by her optician with a history of choroidal naevus in the right eye. She did not notice any distortion

on the Amsler grid and her visual acuity was 0.08 logarithm of the minimum angle of resolution (logMAR) and 0.1 logMAR in the right and left eyes, respectively. Anterior segment examination was unremarkable, while fundoscopy examination of the right eye revealed the presence of a pigmented choroidal naevus of 1.5 disc diameter, adjacent to the superior edge of the optic disc. The naevus appeared flat, with distinct margins and drusen in its context.

INVESTIGATIONS

Optical coherence tomography (OCT) was performed along with fundus autofluorescence (FAF), fundus fluorescein angiography (FFA) and indocyanine green (ICG). OCT (figure 1) revealed pigment epithelium detachment (PED) and a small amount of subretinal fluid, while the choroidal naevus and the surrounding choroid appeared to be thicker with distended vessels, as demonstrated in the enhanced depth imaging (EDI) (figure 2). FAF (figure 3) showed mottled pigment epitheliopathy, while FFA (figure 4) an indistinct area of hyperfluorescence in correspondence with the naevus, which appears to be stable until the late stages. ICG (figure 5) detected multiple polypoidal lesions in the context of the naevus, without any sign of active leakage.

DIFFERENTIAL DIAGNOSIS

In the presence of choroidal naevi, the first approach should aim to rule out any suspicion of malignancy, such as melanoma and choroidal metastasis.² Since the naevus was found at the posterior pole, the OCT was essential to measure the size and thickness, while the FFA and ICG excluded an independent circulation within the naevus, which would have suggested choroidal melanoma. In the presence of neovascularisation, differential diagnosis of choroidal naevus must include melanoma in order to undertake a prompt and adequate treatment.

TREATMENT

The patient was followed up for 6 months and did not require any treatment.

OUTCOME AND FOLLOW-UP

We decided to monitor the polypoidal lesions since they were outside the fovea and not influencing visual acuity. If these lesions will cause active leakage on angiogram, a course of intravitreal

Figure 1 Optical coherence tomography displays the choroidal naevus with polypoidal lesions above it.

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Figure 2 Optical coherence tomography enhanced-depth imaging illustrates the increased thickness of the choroid surrounding the naevus with enlarged vessels.

anti-Vascular Endothelial Growth Factor (VEGF) injections will be considered.

DISCUSSION

The association between the choroidal naevus and PCV has rarely been described so far.^{1,3–7} Asao *et al*³ report a case of an Asian woman with a choroidal naevus at the posterior pole with underlying PCV treated with anti-VEGF agents since the polyps were threatening the fovea and hence visual acuity. Hanhart *et al*⁵ illustrate a similar case of a patient with choroidal naevus and CNV and the importance of OCT angiography to discriminate different types of neovascularisation from malignancy. Wong *et al*⁶ show a good outcome of photodynamic therapy, instead of anti-VEGF agents, as treatment of polypoidal lesions.

The largest case series has been reported by Papastefanou *et al*,⁷ demonstrating an association between the choroidal naevus and different types of CNV in 17 patients. However, in this series only the distinction between classic and occult CNV has been made, without mention of polypoidal lesions and choroidal features.

Another article by De Salvo *et al*⁸ reports the typical findings on OCT of a choroidal naevus complicated by polyps and the importance of excluding melanomas when neovascularisation is present.

Pachychoroidopathy is a recent term that includes pigmentary changes, central serous chorioretinopathy, some cases of type 1 CNV and PCV that share common findings with pachychoroid and dilated vessels.^{9–11} The evidence suggests that in the

Figure 3 Fundus autofluorescence shows mottled pigment epitheliopathy above the disc, corresponding to the area of the naevus and polypoidal lesions.

Figure 4 Fundus fluorangiography demonstrates an indistinct area of hyperfluorescence corresponding to the polypoidal lesions.

Figure 5 Comparison between fundus fluorescein angiography and indocyanine green at the same timeframe clearly shows the hyper-reflectivity and shape of the polypoidal lesions on ICG, without evidence of active leakage or additional neovascularisation from the naevus.

Learning points

- ▶ The association between a choroidal naevus and polypoidal lesions is rare.
- ▶ In the presence of neovascularisation, suspicion of malignancy should be ruled out.
- ▶ Choroidal naevus may influence the haemodynamics of the choroid and trigger neovascularisation.

presence of a choroidal naevus, choroidal haemodynamics are altered, and as a result there is a dilation of the Haller's vessels (pachyvessels), and thinning of the Sattler's vessels and choriocapillaris. Furthermore, abnormal choroidal vasculature seems to lead to photoreceptor dysfunction and apoptosis in the retina. Particularly, long-standing retinal pigment epithelium changes and PED may lead to disruption of the Bruch's membrane and subsequent type 1 neovascular membrane. The new vessels acquire a dilated configuration, resembling polyps.¹¹

This evidence supports the importance of a thorough assessment of choroidal naevi with signs of neovascularisation. It is essential to rule out malignancies and to bear in mind that a space-occupying lesion, such as a naevus, might develop alterations in the choroidal vessels. These features seem to be a risk factor for polypoidal lesions, which need to be detected promptly in order to guarantee appropriate treatment.

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