https://doi.org/10.5114/oku/217400 (Review Paper)
Received: 11.12.2025 Accepted: 07.01.2026

Nonpigmented retinal tumours – differential diagnosis and management

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Summary: Several tumours and pseudotumours can arise from the retinal structures: astrocytic hamartoma of the retina and optic nerve, acquired retinal astrocytoma and glial cell proliferation, retinocytoma, hamartoma of the retina and retinal pigment epithelium , and retinal pigment epithelium tumours and cysts. This article describes examples of nonpigmented retinal tumours and pseudotumours and differential by reviewing the literature and cases on file in the Ophthalmology and Ocular Oncology Clinic, University Hospital, Krakow.

Key words: astrocytoma, retinocytoma capillary haemangioma, retinal pigment epithelium (RPE) tumours.

Various nonpigmented retinal lesions may appear similar. The most common benign nodular lesions will be presented in the article: retinal glioma, astrocytic hamartoma, retinocytoma, capillary haemangioma, sclero-choroidal calcifications, osteoma [1, 2].

Retinal glioma can manifest as astrocytic hamartoma of the retina and optic nerve, acquired retinal astrocytoma, and glial cell proliferation. Astrocytic retinal tumours are usually benign with very low potential for growth. Patients should be examined for tuberous sclerosis complex and neurofibromatosis

Astrocytic hamartoma of the retina and optic nerve occur in about 30% of patients with tuberous sclerosis, and half of them have bilateral tumours. Lesions are identified in patients of all ages, also in newborns. Hamartomatous tumours arise in the retina, developing from retinal glial tissue. They are composed of elongated fibrous astrocytes that have small nuclei. Areas of calcification may be present. There are two common types of astrocytic hamartoma: noncalcified tumour and calcified tumour. Non calcified lesions can be transparent and flat, while larger tumours are grey-yellow in colour and may cause retinal traction (shields). In calcified tumours the degree of calcification may vary. Ophthalmoscopically, astrocytic hamartoma shows glistening yellow spherules of calcification. Hamartomas may show unilaterally or bilaterally and be solitary or multiple, translucent or opaque, calcified or non-calcified (Fig. 1).

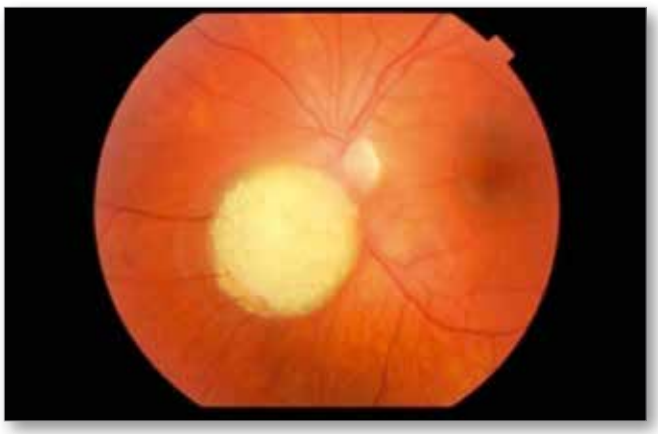

Fig. 1. Colour photograph examination illustrates astrocytic hamartoma.

There are characteristic features in ultrasound (**US**) and optical coherence tomography (**OCT**) imaging.

The A-scan US shows a sharp anterior border and high internal echogenicity, echo deflection in the orbit, and behind the tumour. In B-scan US imaging, astrocytomas show either a high internal echogenicity in the whole or foci of high echogenicity, depending on the level of calcification and shadow.

OCT scans show characteristic structure of the tumour; the internal space appears as if "moth eaten". The relocation of all retinal layers by the tumour resembles and calcifications is well visible (Fig. 2) [2].

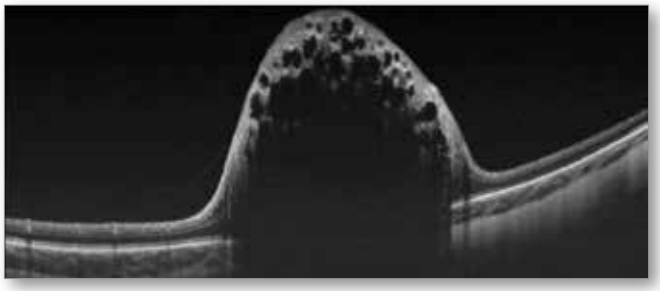

Fig. 2. OCT of astrocytoma – the internal space appear as if "moth eaten".

Tumour shows characteristic features in infrared and autofluorescence examination (Fig. 3).

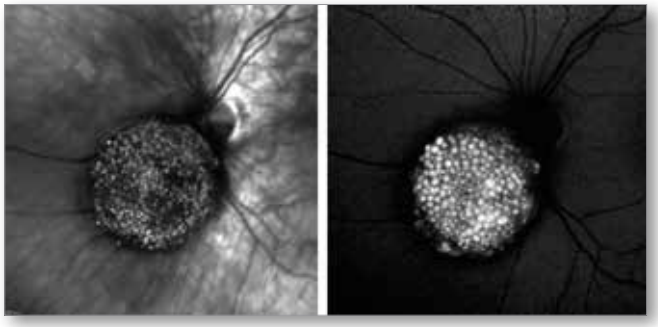

Fig. 3. Astrocytoma in infrared and autofluorescence examination.

A retinal astrocytic hamartoma (RAH) can be an isolated lesion or, more commonly, can associate with genetic syndromes

like tuberous sclerosis or neurofibromatosis. RAH is a benign, congenital tumour of the retina's nerve fibre layer, originating from astrocytes (glial cells).

Management

Treatment is not needed in cases of asymptomatic tumours, but in the case of threat of retinal detachment, a laser barrage should be considered.

Differentiation

In differential diagnosis, the following similar lesions should be taken into account: retinoblastoma, retinocytoma, myelinated nerve fibres, massive retinal gliosis, capillary haemangioma, optic nerve drusen and sclero-choroidal calcifications, choroidal metastases, amelanotic choroidal nevus, intraocular lymphoma, and vasoproliferative tumours.

Differentiation from retinoblastoma is very important, due to similarities in appearance to this malignant tumour.

Astrocytic hamartoma is usually a stable lesion; however, it can show some progressive growth, exudative retinal detachment, and even neovascularisation, glaucoma, and extraocular extension.

In contrast to retinoblastoma, astrocytic hamartoma does not develop prominent retinal feeding and draining blood vessels (shields). Glistening calcifications of astrocytic hamartomas differ from the duller chalky calcifications that characterise retinoblastoma.

Retinocytoma (retinoma)

It is a rare benign intraocular tumour primarily affecting the retina. It is often considered a precursor or a differentiated form of retinoblastoma.

The pathogenesis of retinocytoma is closely linked to mutations in the *RB1* gene, which plays a vital role in regulating the cell cycle. The detection of *RB1* mutations in peripheral blood indicates germline disease, substantially elevating the risk of bilateral retinoblastoma development.

Retinocytoma necessitates vigilant monitoring due to its potential to transform into retinoblastoma (Fig. 4). Management is focused on observation and regular follow-up, and more aggressive treatments are considered if malignant transformation is suspected. There is very low risk of progression to retinoblastoma [2, 3].

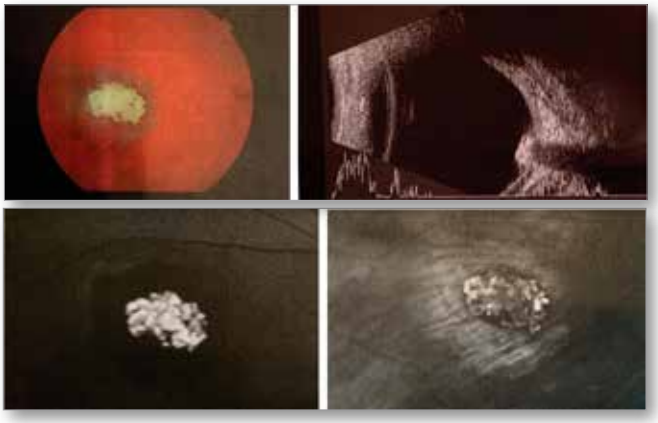

Fig. 4. Retinocytoma: colour picture, ultrasound, autofluorescence and infrared examination.

Capillary haemangioma

Retinal capillary haemangiomas are a benign, vascular tumours that can lead to vision loss from fluid leakage, macular oedema, or retinal detachment. The tumours may occur alone or as part of the genetic condition von Hippel-Lindau (VHL) disease (Fig. 5). Treatments include laser photocoagulation, cryotherapy, photodynamic therapy (PDT), anti-VEGF injections, or surgery, with the specific method depending on the tumour's size and location [4, 5].

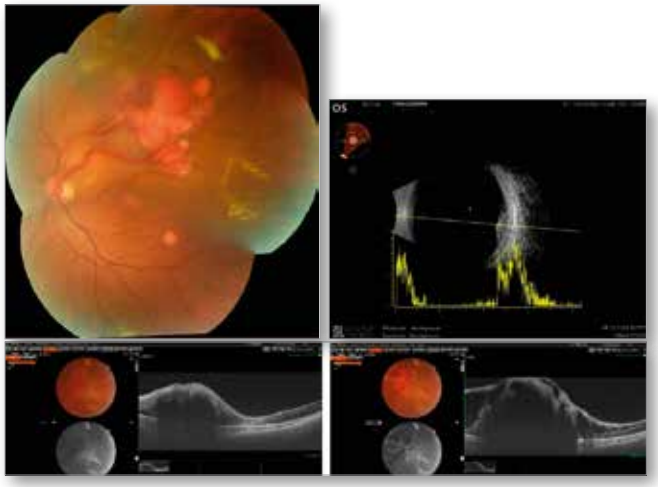

Fig. 5. Capillary haemangioma colour picture, ultrasound, and OCT examination.

Sclerochoroidal calcifications

Lesions usually affect the elderly and middle-aged populations. Most of them have idiopathic features and can co-occur with hypercalcaemia. The lesions are bilateral in 40–80% cases, and they are usually located along the superior or inferior vascular arcade. Sclerochoroidal calcifications are characterised by the presence of isolated yellow and white or cream and orange foci grouped together or large lesions that are elevated (Fig. 6).

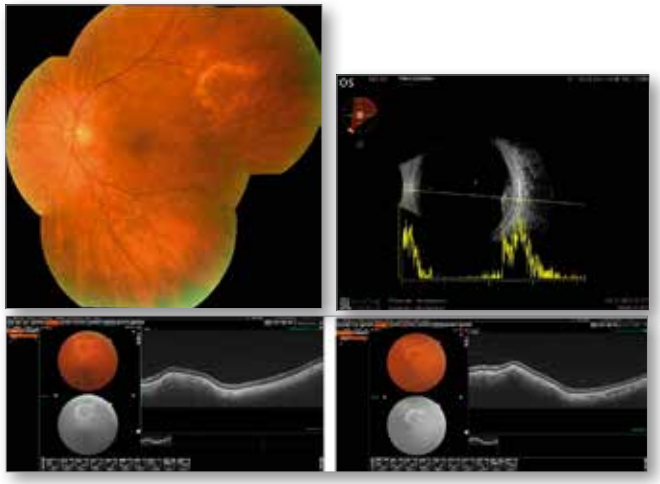

Fig. 6. Sclerochoroidal calcifications colour picture, ultrasound, and OCT examination.

Choroidal osteoma

It is a benign choristoma, localised mainly in the posterior pole of the eye, characterised by slow growth. The tumour is made of osseus tissue of sponge-like structure. It more often occurs in adults and in young woman. This white, partially transparent tumour is located in the posterior segment of the eye, often near the optic disc. Osteomas evolve and enlarge, and they may decalcify, and cause choroidal neovascularisation (CNV) and bleeding.

Diagnosis is based on its characteristic appearance on ophthalmoscopic examination and typical features visualised in ultrasound examination. Additionally, an OCT scan shows the osteoma's structure in detail. This is possible because the tumour is partially transparent [2].

US scans reveal very high echogenicity of the tumour (Fig. 7)

Fig. 7. Choroidal osteoma: colour picture, ultrasound, and OCT examination.

Management

In cases of asymptomatic tumours, only observation is recommended. The appearance of choroidal neovascularisation (CNV) is an indication for intravitreal anti-VEGF injections, laser treatment, or photodynamic therapy.

Vasoproliferative tumour

It is a rare, usually benign, blood vessel growth in the retina, often appearing as an orange-yellow mass, located in the periphery. The tumour is most often idiopathic but rarely can be secondary to other conditions like retinitis pigmentosa. Vasoproliferative tumours can cause visual symptoms (floaters, blurriness) and have a tendency of leakage (exudates), macular oedema, and retinal detachment (Fig. 8) [2].

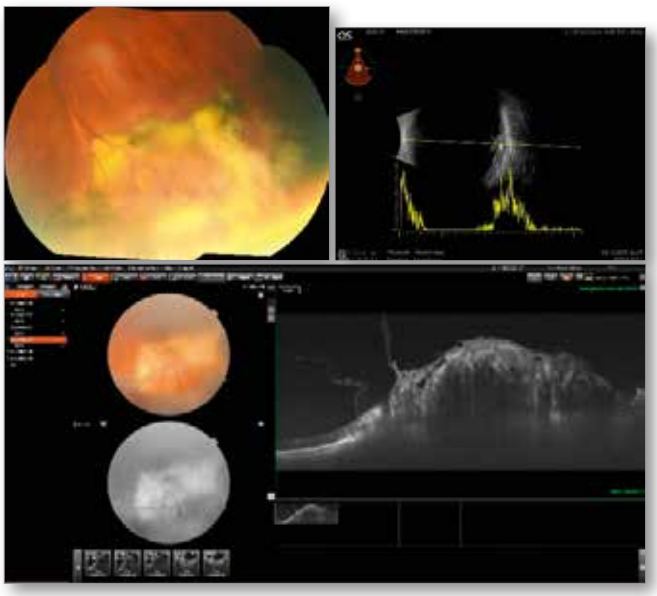

Fig. 8. Vasoproliferative tumour colour picture, ultrasound, and OCT examination.

Management

Leaking tumours should be treated with cryotherapy or radiation therapy (brachytherapy with 106 Ru).

Metastatic tumour

Choroidal metastases, the most common intraocular malignant tumours, spread to the eye's choroid from primary cancers elsewhere, most commonly breast (women) and lung (men) cancer, appearing as yellow/white masses with fluid, causing vision deterioration. There can be many tumours in one eye, and both eyes can be involved (Fig. 9).

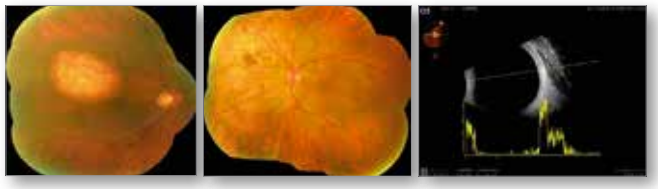

Fig. 9. Choroidal metastases colour picture, ultrasound, and OCT examination.

Management

Systemic treatment can influence intraocular tumour growth, if local treatment is necessary, it should be palliative to preserve sight through transpupillary thermotherapy (TTT) dedicated to smaller lesions or radiation therapy for thicker tumours [6, 7].

Disclosure
Conflict of interests: none declared
Funding: no external funding
Ethics approval: Not applicable.

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