University Hospitals Dorset NHS Foundation Trust

MULTIMODAL IMAGING OF AN INCIDENTALLY DISCOVERED SPORADIC RETINAL ASTROCYTIC HAMARTOMA (RAH)/ASTROCYTOMA

Dr Georgios Tsokolas MSc FEBO FRCOphth[1], Dr. Alina Stinghe MD, Mr Charalambos Tossounis MD FRCOphth[1]
[1]Ophthalmology Department, Royal Bournemouth Hospital, University Hospitals Dorset NHS Foundation Trust, Bournemouth, Dorset, United Kingdom.

PURPOSE

To present a case of sporadic retinal astrocytoma in an otherwise visually asymptomatic patient

MATERIALS-METHODS

The patient underwent full ophthalmic examination under the slit lamp, including dilated fundoscopy. Visual acuities were recorded with the Snellen acuity chart. Multimodal Imaging including Enhanced Depth Imaging (EDI) Optical Coherence Tomography (OCT), OCT-Angiography (OCT-A), Fundus Auto-fluorescence (FAF), Colour Photographs Images were obtained. Zeiss Forum Viewer Software (Zeiss FORUM®) was used.

RESULTS

A 56-year-old female patient was referred to our department due to a lesion that was incidentally discovered by her optician during a routine eye examination. She had no features compatible with tuberous sclerosis and no ophthalmic history of note. From her past medical history, she was diagnosed with Crohn's disease and she was taking Mesalazine and the condition was under control.

Clinical examination of the left eye was unremarkable. Examination of the right eye revealed no signs of inflammation. A creamy yellow-grey lesion was detected along the inferior-temporal vascular arcade of the right fundus (Figure 1A in slide 2). OCT macula showed a healthy foveal contour (Figure 2A in slide 2). OCT over the lesion showed a hyper-reflective dome-shaped thickening at level of the nerve fiber layer (Figure 2B in slide 2). FAF showed hypo-autofluorescenece (Figure 1B in slide 2) and OCT-A demonstrated a dense vascular network within the lesion in the deep retinal plexus (Figure 3 in slide 3). Systemic work-up was unremarkable.

Over a 3-year-period, the lesion has remained stable and the patient is still visually asymptomatic with visual acuity of 6/6 on the Snellen chart.

Contact details: georgetsokolas@yahoo.com
Financial disclosures: All authors declare that they have no conflicts of interest and that no funding was obtained for this work

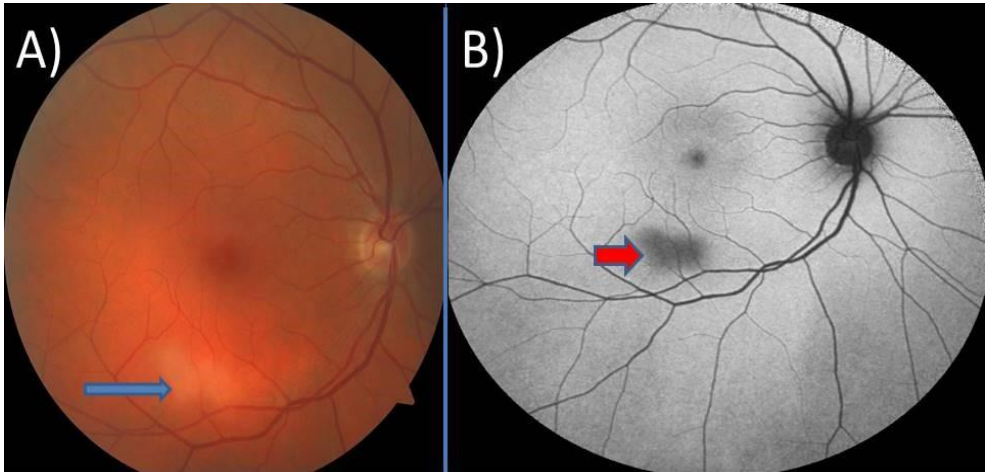

Figure 1. From left to right : A) Colour fundus photograph of the patient's right fundus. Note the creamy-yellow lesion along the inferior-temporal vascular arcade (blue arrow). B) FAF of the patient's right fundus showing hypo-autofluorescence of the lesion (red arrow)

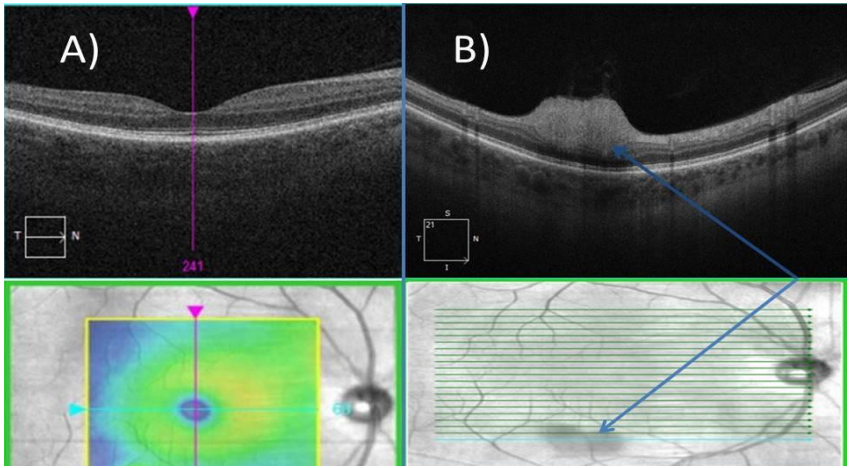

Figure 2. From left to right: A) OCT image of the fovea demonstrating an intact foveal contour. B) High Definition EDI OCT scan over the lesion. Note the hyper-reflective lesion at the level of the nerve fibre layer along the inferior-temporal vascular arcade (2 oblique blue arrows)

COMMENTS-PART 1

Differential diagnosis included retinoblastoma, retinocytoma, amelanotic naevus, amelanotic uveal melanoma, capillary haemangioma, combined hamartoma of the retina and the retinal pigment epithelium, retinal pigment epithelial adenoma, choroidal metastasis and a cotton wool spot. Based on the clinical examination, past medical and ophthalmic history and with the aid of the multimodal imaging, most of the above clinical entities were excluded because they involve the retina and choroid. In our case, the lesion involved exclusively the retinal nerve fibre layer and the outer retina and choroid were normal. Furthermore, retinoblastoma, retinocytoma, combined hamartoma of the retina and the retinal pigment epithelium, retinal pigment epithelial adenoma are usually lesions presenting during early childhood and not during adult life. The patient had also no history of previous cancer treatment and apart from the diagnosis of Crohn's was systemically well with no symptoms suggesting malignancy. Therefore, choroidal metastasis was also excluded.

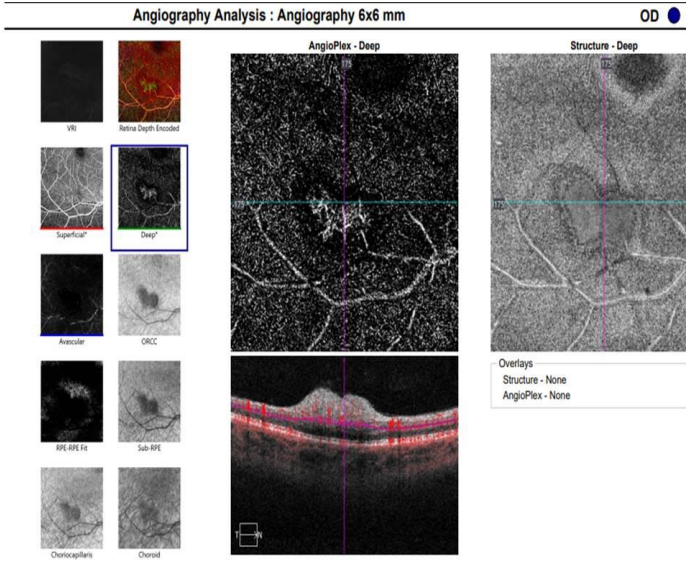

Figure 3. OCT-A of the lesion demonstrating a prominent vascular network within the lesion in the deep retinal plexus.

CONCLUSIONS

Retinal astrocytomas are rare benign glial cell tumours. They may be associated with tuberous sclerosis, or may be sporadic (as in our case). Diagnosis is predominantly clinical and is supported by ancillary tests. Visual prognosis is generally good.

Periodic monitoring is advised to detect those lesions showing aggressive growth or vision-threatening complications.

COMMENTS-PART 2

The patient's blood pressure was normal and HbA1C was negative. Thus, she had no obvious cardiovascular risk factors to explain a presence of a cotton wool spot. Cotton wool spots are localized area of retinal nerve fiber layer necrosis due to retinal ischaemia and capillary drop out. OCT-A in our case did not show any evidence of this. In fact, there was a very rich vascular network within the lesion located at the deep retinal plexus. Therefore, the OCT-A in our case showed the exact opposite features that a cotton wool spot would normally exhibit, and this made the diagnosis of cotton wool spot less likely. In the absence of hypertension and diabetes, with the rest of the blood tests being negative and after having excluded most of the aforementioned lesions, we proposed that this lesion was a presumed sporadic astrocytoma.

REFERENCES

1. **https://eyewiki.org/Retinal Astrocytic Hamartoma**
2. **Shields JA, Shields CL. Intraocular tumours: an atlas and textbook. 3rd Ed Philadelphia Lippincott Williams & Wilkins (LWW) 2015**
3. **Shields CL, Say EAT, Fuller T et al.. Retinal Astrocytic Hamartoma Arises in Nerve Fibre Layer and Shows "Moth-Eaten" Optically Empty Spaces on Optical Coherence Tomography. Ophthalmology. 2016;123(8):1809-1816**
4. **Ulbright TM, Fulling KH, Helveston EM. Astrocytic tumours of the retina. Differentiation of sporadic tumours from phakomatosis-associated tumours. Arch Pathol Lab Med. 1984;108(2):160-163**