Diagnostic imaging of Retinal Astrocytomas: a case-series

K. McLean[1,2], R. Hussain[1,3], H. Heimann[1,3]

[1]Liverpool Ocular Oncology Centre, St Paul's Eye Unit, Liverpool University Hospitals Trust, UK, [2]Department of Eye and Vision Science, Institute of Life Course and Medical Science, University of Liverpool, UK, [3]Liverpool Ocular Oncology Research Group, Department of Molecular and Clinical Cancer Medicine, University of Liverpool, UK

Background

- Retinal astrocytomas (RA) are rare tumours of retinal glial cells.[1]
- Usually asymptomatic but rare cases can exhibit aggressive behaviour.[1,2]
- Clinical presentations can be similar to amelanotic melanomas and retinoblastomas but management is very different.
- It is therefore highly important to correctly identify them.

Aim

- To provide an aid which will help practitioners distinguish RAs from other causes of non-pigmented lesion of the fundus, which can sometimes be challenging.

Objectives

- We provide our experience of using multi-modal diagnostic imaging techniques to identify RA.
- Analyse the characteristics of RA on each modality.
- Compare findings with the current literature.

Methods

- Retrospective analysis of notes and imaging of 17 patients diagnosed with RA at a single centre between January 2012 and June 2021.
- Demographics, examination findings and imaging including colour fundus photography, optical coherence tomography (OCT), infra-red (IR) and ultrasound (US) were analysed.
- These were compared to differential diagnoses, including retinoblastomas, amelanotic choroidal melanomas, choroidal metastases and idiopathic scleromas.

Results

Demographics

- Eighteen eyes of 17 patients were included.
- Mostly unilateral solitary lesions, except one bilateral case.
- Ten female (59%) and seven male (41%).
- Mean age at presentation was 43 years old (range 12 - 66).
- Majority were asymptomatic (71%, 12/17).
- Three patients had associated Tuberous Sclerosis.

Ophthalmoscopy and fundus photography

- All lesions were creamy, white.
- **Two distinct appearances:** One type (39%, 7/18) were poorly-defined translucent elevations of the retina *(Fig. 1a)* and the other (61%, 11/18) were solitary, solid, opaque masses with well-defined borders *(Fig. 1b).*
- One patient had associated haemorrhages and exudates tracking towards the macula *(Fig. 1c)*.
- Pathognomonic mulberry-like spherules were present in 35% (6/18).

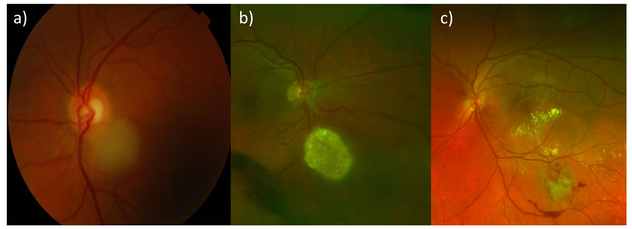


Figure 1: Colour photographs of retinal astrocytomas a) Poorly-defined translucent variant, **b)** Variant which is a solid, opaque mass with well-defined borders, **c)** Aggressive RA with associated haemorrhages and exudates tracking toward the macula.

Infrared imaging

- Six (38%; 6/16) had hyper-reflective calcifications *(Fig. 2b)* and nine (56%; 9/16) were hypo-reflective *(Fig. 2a).*

Optical coherence topography

- Dome-shaped retinal thickening with disruption of normal layers *(Fig.2).*
- Eight (53%, 8/15) had a solid appearance *(Fig. 2a)*. Nine (60%, 9/15) had intra-retinal cystic spaces giving a moth-eaten appearance *(Fig. 2b).*

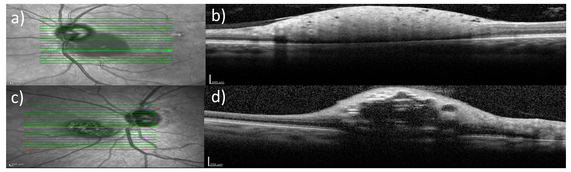


Figure 2: Infra-red (IR) and Optic Coherence Imaging (OCT) of retinal astrocytomas a) IR well-defined hypofluorescence, **b)** OCT solid dome-shaped thickening of the inner retinal layers, **c)** IR hyper-reflective calcifications, **d)** OCT dome-shaped thickening of the inner retinal layers, multiple intra-retinal cystic spaces and posterior shadowing.

Fundal autofluorescence

- Five (38%; 5/13) had hyperfluorescent cystic-like opacities *(Fig. 3b)* and four (31%; 4/13) were hypoflourescent appearance *(Fig. 3c).*

B-scan ultrasonography

- Maximum basal diameter ranged from 3.30 to 6.01mm (mean 4.36mm) and maximum thickness ranged from 0.68 to 2.09mm (mean 1.32mm).
- High internal reflectivity was noted in eleven (92%; 11/12) cases *(Fig. 3c).*
- Significantly larger basal diameter and thickness were measured noted on US compared to OCT (p = 0.001 and 0.004).

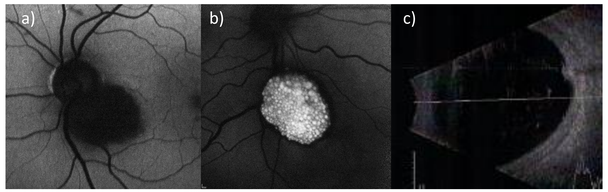


Figure 3: Fundal autofluorescence (FAF) and ultrasound B-scan (US) of retinal astrocytomas a) FAF demonstrating hypofluorescence RA, **b)** FAF demonstrating hyperfluorescent cystic-like opacities and calcifications giving a mulberry-like appearance, **c)** US b-scan of RA demonstrating high-internal reflectivity.

Discussion

- RA patients are older than retinoblastoma ($\bar{X} = 23.5$mths) but younger than choroidal melanoma ($\bar{X} = 60$yrs).
- RA have two distinct appearances:
  > Poorly-defined translucent retinal elevations (Fig. 1a)
  > Solid, opaque mass with well-defined borders (Fig. 1b).
- Clustered, calcified spherules are pathognomonic and present in ~ 35%. OCT, FAF and IR help identify these.
- RA calcifications are 'glistering yellow' compared to retinoblastoma calcifications which are more 'chalky-dull'.[1]
- OCT demonstrates RA as dome-shaped retinal thickening with disruption of inner retinal layers and cavities in ~60% give a moth-eaten appearance.
- Retinoblastomas on OCT are highly reflective disorganisation of the full-retinal thickness, cavities are rare. Choroidal melanomas are evidently in the choroid on OCT.
- RA are mostly hyperechoic on US, whereas retinoblastomas have heterogeneous shadowing and melanomas are acoustically hallow.

Conclusions

- RAs display characteristic clinical, demographic and imaging features which can aid differentiating them from other non-pigmented fundal lesions.
- Use multiple imaging modalities to diagnosed RA.

References

1. Shields et al. *Saudi J Ophthalmol.* 2009; 23: 197–201.
2. Shields et al. *Arch Ophthalmol.* 2005; 123: 856–863.

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K.McLean@liverpool.ac.uk