CFEH Facebook Case #112

A 50 year old Caucasian female presented for an assessment. She reports having problems in low light and driving at night time. She is generally healthy and has no significant family or medical history. Visual acuity is 6/4.8 in each eye. Imaging results are below. What is the likely diagnosis for this patient?
Optomap images show bone spicule intra-retinal pigmentation around the inferior arcades in both eyes while the superior retina appears unaffected. The retinal changes are clearly evident on fundus autofluorescence with large areas of the inferior retina showing hypo-fluorescence. There is a corresponding superior field defect in both eyes.

This presentation is consistent with a diagnosis of sector retinitis pigmentosa.

Sector retinitis pigmentosa (RP) is an atypical presentation of RP that typically involves the inferior retina. The affected area shows the characteristic bone-spicule pigmentation while the other retinal areas have a normal appearance. This condition is considered slowly progressive but given this patient’s excellent acuity and current age, the prognosis in this case is likely to be favourable.

A binocular Esterman visual fields test was conducted to assess the patient’s vision for driving and the RP diagnosis was confirmed with electrophysiological testing.