Imaging papilloedema vs. pseudo-papilloedema

BY VICTORIA COSGROVE

Quite often, in the working week as an ophthalmic photographer, you will be given that patient with 'swollen discs' to image. These swollen discs could be a number of things, but mainly fall into one of two categories: papilloedema or pseudo-papilloedema. So, what is papilloedema? How is it different from pseudo-papilloedema? Papilloedema is defined as swollen optic discs caused by raised intracranial pressure (ICP) diseases such as idiopathic intracranial hypertension (IIH). Psuedo-papilloedema, in comparison, is raised and 'swollen' optic discs caused by structural or other reasons such as optic disc drusen.

Imaging idiopathic intracranial hypertension

The monitoring of IIH is done in numerous ways but close attention is paid to the optic disc and any swelling [1]. Therefore, imaging this correctly and clearly is important to the neuro-ophthalmologists. Optical coherence tomography (OCT) is becoming more and more important for monitoring IIH, recently the paper 'Using Optical Coherence Tomography as a surrogate of measurements of intracranial pressure in Idiopathic Intracranial Hypertension' by Susan Mollan and her colleagues in Birmingham has shown that not only is OCT used as initial analysis of the optic nerve head but it can also be used to monitor ICP in IIH patients, which reduces the need for invasive procedures such as lumbar punctures [2].

From a photographer's point of view there are some different optic disc scans that are available to us. Using imaging equipment, such as the Heidelberg Spectralis, we can perform a retinal nerve fibre layer (RNFL) circular scan and a cross-sectional optic nerve head (ONH) scan. The RNFL shows the thickness of the nerve fibre layer and this is helpful in mild cases of papilloedema. However, when the optic disc is moderately swollen or more then this is not helpful, as the OCT technology cannot segment the layers correctly due to the degree of swelling. You can see in Figure 1 that the nerve fibre layer cannot be seen very clearly as the swelling is too great for the machine to pick up the retinal layers, therefore the thickness measurements are not accurate, though they do indicate the swelling in the disc.

The optic nerve head scan is the second scan which shows us the cross-sectional view of the nerve head. By calculating the distance between the Bruch's membrane (BM) and inner limiting membrane (ILM) a thickness map is produced, enabling the observation of any swelling and potential causes such as disc drusen. If the disc is more than moderately swollen, then unlike the RNFL scan, we still get a fair view into the optic disc plus an indication of the thickness measurement, though this can still be inaccurate due to inaccurate segmentation, as seen in Figure 2.

OCT technology has advanced enough to give us the ability to manually adjust these positions if we are able to move them to the correct positions. The Heidelberg Spectralis has an additional form of scanning possible, called the Anatomical Positioning System (APS). Again, this is only possible on mildly swollen discs. This is because the APS maps within the eye where the BM and the fovea

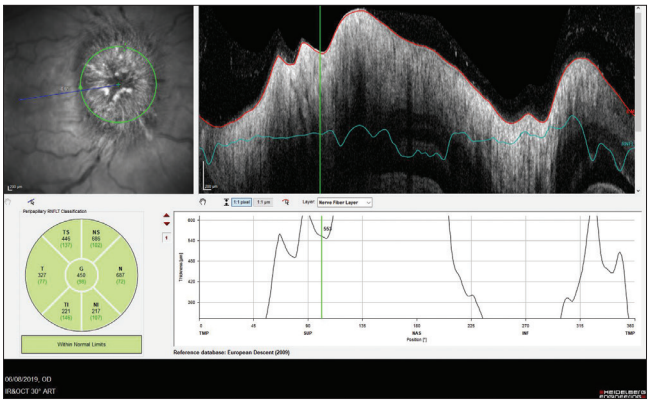


Figure 1: RNFL scan showing inaccurate segmentation on an optic disc with papilloedema.

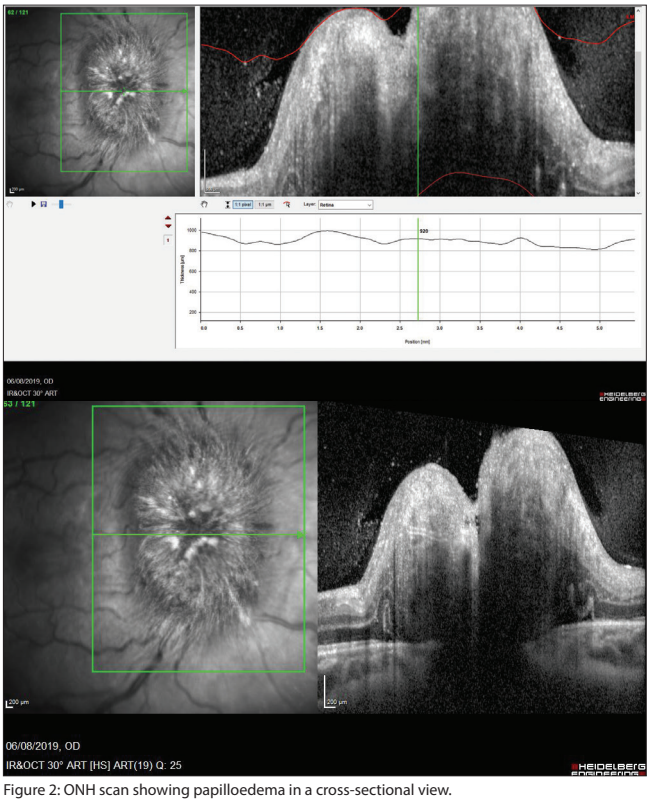
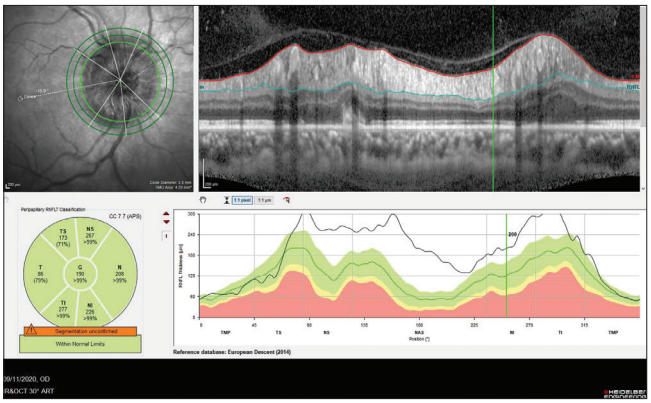


Figure 2: ONH scan showing papilloedema in a cross-sectional view.

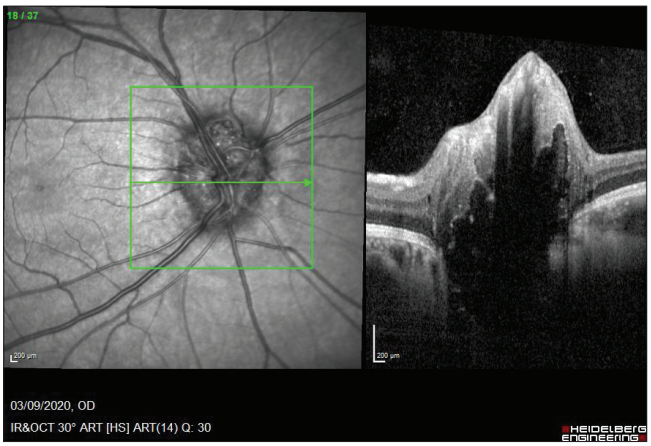
are, providing measurements of RNFL, ganglion cell layer (GCL) and others specific to the eye in question. This then provides more accurate measurements of nerve head thickness and specifically volume to show the degree of swelling in a patient, plus information on how affected the macula GCL is which can thin in some neuro-ophthalmic conditions.

You can see in Figure 3 that the segmentation cannot be confirmed to be accurate, so in this case this particular scan has been manually



(Left) Figure 3: Spectralis APS scan of optic nerve head showing papilloedema.

(Right) Figure 4: Superficial disc drusen.



(Left) Figure 5: ONH scan of optic disc showing superficial disc drusen.

(Right) Figure 6: FAF showing superficial optic disc drusen.

adjusted to the best of the photographer's ability to show the truest thickness possible.

Overall, in imaging IIH the most important thing to remember is that the quality and accuracy of the scan you get depends if the swelling in the disc is mild to moderate or more severe. The more severe cases cannot be imaged with a great deal of accuracy, however, the more moderate swollen discs can be imaged to a high quality and therefore provide accurate information to the neuro-ophthalmologist without performing a non-invasive procedure.

Imaging optic disc drusen

Quite often, we are sent patients from A&E who are generally young, usually teens to early twenties, with suspicious swollen discs. On scanning these patients, we can see there is in fact no true papilloedema. Instead, the patient has optic disc drusen and it is mimicking the appearance of papilloedema.

Optic disc drusen are congenital calcified hyaline abnormalities that occur within the disc. They can be superficial or buried and what type a patient has determines how easy it is to image them. Superficial disc drusen are easy to see on fundus examination and give the optic disc a bumpy and uneven appearance, as seen in Figure 4.

Because it is superficial we can use OCT to image within the disc. The initial scan best used is the optic nerve head scan, but with the setting amended to enhanced depth imaging (EDI). This causes the choroid to be the focus rather than the vitreous interface and therefore provides a clearer view of the drusen, as seen in Figure 5.

In most cases these cause no problems, but it is important to be aware that some patients experience visual field defects because of the drusen, especially as they enter their 20s. In fact, I have only ever come across one case. Another way to image disc drusen is by using autofluorescence (FAF) imaging. Because of the make-up of disc drusen they give off a hypofluorescence, the level of which depends on how superficial or buried they are. Logically the more buried the drusen, the less hypofluorescent they are.

As a general rule, I now find that if I am presented with a young patient with suspicious discs, I will always perform these imaging techniques to ensure that all available information I can provide is there to aid the doctor with diagnosis.

Buried disc drusen is more difficult to image and normally cannot be seen on the OCT scan or the FAF images. In these cases, we normally perform B-scan ultrasonography to show the drusen, as it shows the drusen much more clearly.

Because all these imaging modalities are non-invasive and painless to the patient, it is easy for us to perform them, as long as the patient is dilated. Neuro-ophthalmology is beginning to realise the potential of ophthalmic imaging, in particular OCT to aid them with diagnosis and monitoring of their patients. For our ophthalmology doctors in A&E, we can help aid them in differentiating between papilloedema and pseudo-papilloedema, which is important not only for us but also for the patient's care pathway.

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