Glaucoma Masqueraders – Our Clinical Experience – Has OCT Made Diagnosis Easier?

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Abstract

Conditions mimicking glaucoma with field changes or optic nerve head changes similar to glaucoma can puzzle the clinician. We present a series of 17 patients with optic nerve head pit, optic nerve head drusen, ONH coloboma, ocular ischemic syndrome, compressive optic neuropathy, and ocular hypertensives with high cup disc ratio in myopic optic discs which appear glaucomatous but with no field loss. OCT helped us to differentiate some of these. The clinical presentation, visual field analysis, OCT picture and also features clinching the diagnosis in these patients is discussed in detail.

Keywords: Optic pit, compressive optic neuropathy, Ocular ischaemic syndrome, OCT

Introduction

The diagnosis of primary open angle glaucoma (POAG) implies a life long disease with irreversible visual loss for which regular medication and followup is needed for the rest of the life. The visual loss is painless and progressive, and thus may go undetected as it does not involve the central vision initially. With early detection techniques we can halt and prevent visual loss due to glaucoma by appropriate and timely management. At the same time it must be differentiated from other causes which present similarly but with subtle differences in clinical presentation where both the prognosis and management differ.

The ‘glaucoma masqueraders’ mimick glaucoma in one or more ways. They may cause a painless visual loss, that involves the periphery earlier and more severely than central vision involvement, or some may have high intraocular pressure (IOP) e.g. Compressive optic neuropathy (CON), Ocular hypertension (OHT). Others may have optic nerve head (ONH) changes similar to glaucomatous optic neuropathy (e.g. Optic pit and isolated ONH coloboma). Still others may have visual field defects simulating or mimicking glaucoma. Superior arcuate scotoma may be seen in optic pit, anterior ischemic optic neuropathy (AION), compressive optic neuropathy. Glaucoma field loss affects the nasal quadrant initially and may mask an unusual visual field presentation of a pituitary tumor. Differentiating a case of normotensive glaucoma from OHT can be a diagnostic dilemma.
Materials and Methods

This is a nonrandomised, noncomparative, institution based, clinical observational case series study of 17 cases selected from the out-patient department from December 2004 to 2007 June.

Those patients who had at least 1-2 features of POAG, either a high IOP or an enlarged cup, or an arcuate scotoma or had a suspicious small disc with pallor were included.

All patients underwent a complete glaucoma evaluation including applanation tonometry, pachymetry, gonioscopy, automated perimetry with 30-2 on Humphrey, and a dilated stereoscopic optic nerve head evaluation.

OCT was done for all patients using Fast RNFL, Fast optic nerve head scan and Macular protocol on the Stratus OCT Version 4 machine.

Observations

The fundus findings, the HFA visual field, and OCT findings were noted and correlated for all the patients included in the study.

<table>
<thead>
<tr>
<th>Conditions mimicking glaucoma</th>
<th>No. of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Optic pit</td>
<td>6</td>
</tr>
<tr>
<td>Optic Coloboma</td>
<td>1</td>
</tr>
<tr>
<td>Ocular Ischemic Syndrome (OIS)</td>
<td>2</td>
</tr>
<tr>
<td>Compressive Optic Neuropathy (CON)</td>
<td>3</td>
</tr>
<tr>
<td>AION</td>
<td>1</td>
</tr>
<tr>
<td>Orbital apex syndrome</td>
<td>1</td>
</tr>
<tr>
<td>Myopia and OHT</td>
<td>1</td>
</tr>
<tr>
<td>Optic nerve head drusen</td>
<td>1</td>
</tr>
<tr>
<td>Optic neuritis/ Angle closure glaucoma</td>
<td>1</td>
</tr>
</tbody>
</table>

Of the 17 patients (Table 1), 6 had optic nerve head pit-3 unilateral and 3 bilateral. All the 6 patients with optic pit had an arcuate scotoma (Fig. 1b)

Fig. 1. (a) Fundus photo of a case of Optic Pit

Fig. 1. (b) HFA 30-2 of a case of Optic Pit
with normal IOP which did not progress in the 12 months of followup. Central vision was decreased in 3 patients and three had bilateral pits. Pits were seen on the temporal aspect of the disc. The optic nerve head showed characteristic features of an optic pit (Fig. 1a). On OCT the pits could be seen on the temporal aspect of the cup on a horizontal line scan. There was RNFL loss around the disc (Fig. 1c, d, e) and normal optic nerve head parameters were found in all of our patients. One case has cystoid changes and schisis cavity formation in the peripapillary retina. The serous macular detachment on OCT communicated with the optic pit.

Two, had Ocular Ischemic syndrome (OIS) and three had Compressive optic neuropathy. One each had optic nerve head drusen, Optic nerve coloboma, AION, orbital apex syndrome, misdiagnosed optic neuritis.

The patient with Optic nerve head coloboma had an isolated coloboma with no anterior coloboma (Fig. 2a). The patient had a superior arcuate scotoma (Fig. 2c) with a suspicious looking disc and a normal intraocular pressure. There was a white excavation in the disc which was decentered inferiorly, with a thin inferior rim and a normal superior rim (Fig. 2a and b). OCT in this case showed inferior retinal fibre layer (RNFL) thinning (Fig. 2d) and signs of a connection

### Table 2: BCVA of patients of Optic Pit

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Right Eye</th>
<th>Left Eye</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient 1</td>
<td>Optic pit</td>
<td>6/6</td>
</tr>
<tr>
<td>Patient 2</td>
<td>Optic pit</td>
<td>6/6</td>
</tr>
<tr>
<td>Patient 3</td>
<td>Optic pit</td>
<td>6/6</td>
</tr>
<tr>
<td>Patient 4</td>
<td>Optic pit</td>
<td>5/60-6/36</td>
</tr>
<tr>
<td>Patient 5</td>
<td>Optic pit</td>
<td>6/6</td>
</tr>
<tr>
<td>Patient 6</td>
<td>Optic pit</td>
<td>6/6</td>
</tr>
<tr>
<td>Patient 7</td>
<td>Optic pit</td>
<td>6/6</td>
</tr>
</tbody>
</table>
Fig. 2. (a) Fundus photograph showing an isolated optic disc coloboma with a white excavation on disc, decentered inferiorly with a thin inferior rim and a normal superior rim

between the perineural space and the inner retinal layers on the temporal optic disc border (Fig. 2e), as well as schisis-like changes extending from the disc to the macula, with cystoid degeneration and two lamellar holes. There was increased retinal nerve fibre layer thickness and reduced macular thickness.

The patient with Optic nerve head drusen had bilateral visible supertemporal drusen that lead to bilateral inferior arcuate scotoma. The drusen unless seen stereoscopically could be mistaken for notching of the cup. On stereoscopic examination it presented as a raised lesion. On OCT both eyes showed a supertemporal thinning on RNFL scan, and all parameters on ONH analysis were raised. (Fig. 3 a, b and c).

The two patients with Ocular Ischaemic syndrome had a high CD ratio, with a deep cup and extensive field loss with normal IOP’s. On OCT patient had RNFL loss, and decreased ONH parameters like VIRA, HIRW and rim area

History and Carotid Doppler helped clinch the diagnosis of Carotid artery stenosis (Fig. 4 a and b).

Three patients with Compressive optic neuropathy (Fig. 5) in thyroid exophthalmopathy and one with

Fig. 2. (b) Red free fundus photograph of optic disc coloboma

Fig. 2. (c) HFA left eye showing a superior arcuate scotoma

Fig. 2. (d) OCT showing Inferior Retinal Nerve Fibre Layer (RNFL) thinning in an isolated Optic nerve head coloboma
Orbital apex syndrome presented with raised IOP, arcuate scotomas with ONH pallor associated with proptosis. The patients were managed on systemic steroids – (Methylprednisolone in severe cases) along with IOP lowering agents that decrease aqueous production.

On OCT in our patients all ONH parameters were decreased with inferior RNFL thinning with corresponding field changes (Fig. 5b & c). One had a double arcuate scotoma inferiorly more than superiorly (Fig. 5d).

The AION patient [Fig. 6a, b & c] had a small pale disc with a small cup, indicating a resolved neuropathy with resorption of the peripapillary oedema.

The field defect was an arcuate scotoma and IOP was normal. In the acute phase the gross elevation of the disc was clearly evident on OCT and RNFL was normal.

Also one had high intraocular pressure, and high cup disc ratio in a myopic optic discs which appeared glaucomatous. This patient had no field loss even on followup and normal OCT parameters. This patient had ocular hypertension with a borderline thick cornea, open angles, no field defects and normal OCT. The patient is on follow-up regularly and medication has not been started for last 1 year (Fig. 7).

Incomplete and partial Binasal hemianopia was seen in one patient. It appeared like glaucomatous optic neuropathy as the CD ratio was high. The IOP was normal, and had been mistakenly treated as bilateral POAG. After pituitary surgery on the adenoma, the field...
defects dramatically disappeared. In this patient neither RNFL loss nor ONH abnormalities were seen on OCT.

Discussion

Optic Pit: The visual field defects in an optic pit is a superior arcuate scotoma similar to glaucoma, but it is nonprogressive, and management does not include an IOP lowering agent. The danger to visual loss is due to neurosensory detachment or retinoschisis. It is only the discerning trained eye of a clinician, that picks up the optic pit on stereoscopic fundus examination rather than diagnose it as optic cupping. OCT has made the diagnosis easier, as on OCT the pit is clearly visible on the temporal aspect of the cup on a horizontal line scan, and so is the associated neurosensory detachment. RNFL and optic nerve head parameters are normal. Optic pit is one condition where OCT helps in differentiating it from glaucoma. According to literature 15% have bilateral disease, while three out of six of our patients had bilateral disease. The cystoid changes and schisis cavity formation in the peripapillary retina can be seen. The serous macular detachment on OCT can be seen communicating with the optic pit.
Fig. 4. (a) Fundus findings in a case of Ocular Ischemic Syndrome (OIS)

Optic Nerve head coloboma when isolated, i.e. with no anterior or retinal coloboma, presents with a superior arcuate scotoma in a suspicious looking disc in a patient with normal intraocular pressure. It is caused by the failure of complete closure of the proximal end of the embryonic fissure. It is characterized by a white excavation in the disc which is decentered inferiorly. The inferior rim is usually thin or absent whereas the superior rim is relatively normal.

OCT showed signs of a connection between the perineural space and the inner retinal layers on the temporal optic disc border, as well as schisis-like changes extending from the disc to the macula, with cystoid degeneration and two lamellar holes in their nasal portion. There is

Fig. 4. (b) OCT findings in OIS
increased retinal nerve fibre layer thickness and reduced macular thickness.

**Optic Nerve Head Drusen (OND)**

Optic disc drusen occurs in about 1% of the population and are found more frequently in Caucasians. 75% are bilateral. Inherited or sporadic, it is a form of calcific degeneration in some of the axons of the optic nerve.
Visual acuity is often not affected, but the visual fields of these patients can be abnormal and deteriorate over time. The drusen may be buried or evident. The disc picture shows an elevated disc that may be confused with papilledema (Fig. 3a). The lesion may be progressive. Buried drusens are less symptomatic. There is no existing treatment for optic nerve head drusen. Proper diagnosis and patient education is the best-available modality of care. Patients need to be aware of potential complications which, while rare, can affect vision. Visual-field testing can aid in monitoring for subtle changes in vision.

OCT has shown a thinning of peripapillary retinal nerve fiber layer. The lowest values were found in the superior and inferior quadrants. Calcification, picked up both on USG B scan and CT scan, is the characteristic feature for diagnosis for ONH drusen.

Visual field defects are uncommon in eyes with buried OND. Eyes with buried OND may have focal RNFL defects but have normal average RNFL thickness.

Coexistence of ONH drusen and POAG in eyes can be most effectively picked up by OCT earliest where the disc may appear elevated on examination but RNFL thinning may be picked up even prior to appearance of visual field defects.
In Ocular Ischemic syndrome OCT did not provide any corroborative evidence, rather, the OCT changes were indistinguishable from glaucoma. The clinical picture is hard to distinguish from normotensive glaucoma. The ONH picture was confusing and history and Carotid Doppler helped clinch the diagnosis of carotid artery stenosis.

Compressive optic neuropathy in thyroid exophthalmopathy or Orbital apex syndrome can present with raised IOP, arcuate scotomas with ONH pallor in a patient of proptosis with or without thyroid disease. The treatment is with systemic steroids, methylprednisolone in severe cases along with IOP lowering agents that decrease aqueous production. Episceral venous pressure is raised in these cases, thus medication that acts on the outflow mechanism may not work.

On OCT in our patient with CON all ONH parameters were decreased with inferior RNFL thinning. He had a double arcuate scotoma inferiorly more than superiorly (Fig. 9). Eyes with CON had significantly larger rim area and smaller cup parameters but similar RNFL thickness compared with controls on Heidelberg Tomograph but no study has been reported using OCT. However, caution has been advised by experts while interpreting the parameters obtained from the eyes with CON. One patient with bilateral field defects & severe proptosis was found to have improvement in visual fields and ONH parameters after radiotherapy.

AION patients have a small pale disc with a small cup especially in a longstanding one where the associated peripapillary edema has subsided.

The field defect can be an arcuate scotoma and IOP is normal. In the acute phase the gross elevation of the disc is clearly evident on OCT and RNFL is normal. Disc topography of eyes with AION was quantitatively

Fig. 6. (c) OCT findings in AION with elevation of nerve fibre layer on ONH scan and raised RNFL thickness

Fig. 7. Fundus picture in Ocular Hypertension
characterized by small and shallow cupping and a relatively large rim area compared to eyes with OAG matched for age and VFD. In eyes with AION, significant correlation with VFD was found only for the RNFL thickness evaluated with SLP but not for the HRT II parameters.

**Conclusion**

OCT can help in confirming the diagnosis in certain cases, and prevent overdiagnosis of POAG in clinical settings similar to glaucoma. Thus, patients of optic nerve head pit, coloboma, drusen, CON, AION, Ocular Ischemic Syndrome and ocular hypertensives can be managed better. OCT has the advantage vis a vis GDX that apart from the glaucoma evaluation, the retina can be simultaneously evaluated. CON and OIS have indistinguishable findings from glaucoma on OCT. The OCT helps in distinguishing optic pit, ONH coloboma, ONH drusen and AION.

**References**


