DISC DRUSEN AND ANTERIOR ISCHAEMIC OPTIC NEUROPATHY
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ABSTRACT
Sudden loss of vision due to the effects of drusen on the optic nerve is rare. We describe a white woman aged 42 who had sudden loss of vision in the right eye, followed by the left in a similar fashion at an interval of twelve months. The results of complete medical and neuro-radiologic examinations were entirely normal. Summarised evidence showed that this was due to drusen causing compression anterior ischaemic optic neuropathy.

INTRODUCTION
Optic disc drusen are anomalous hyaline bodies of the optic nerve head situated in front of the lamina cribrosa. They are laminated, homogenous masses which frequently become calcified. They are bilateral in 75% to 80% of cases, 25% being unialateral even after a long period of follow up. Superficial drusen appear as irregular, glistening yellow globules that may be isolated or clustered. Deep drusen are not directly visible, thus causing pseudo-papilloedema; they evolve slowly often requiring decades to develop. Axoplasmic transport alteration is the anatomic substrate for the formation of drusen of the optic disc. In a histopathological study made on 18 autopsies Giarelli et al\(^1\) reported deformation and displacement of papillary vessels including the retinal artery and vein in most of the severe cases. Visual field defects occur in 85% of cases by compression causing atrophy of the adjacent nerve fibres or on a vascular ischaemic basis. Sudden visual loss is not common.

CASE REPORT
A 42 year old white woman was referred with papilloedema to the neurosurgery department on 17th December, 1990, with a history of painless sudden loss of vision in the right eye of four days’ duration. The visual loss in the right eye was first noticed in the inferior nasal quadrant and over the four day period had involved the complete nasal half. The patient was admitted and investigated for papilloedema. In the past she was diagnosed to have mild hypertension of two years’ duration for which she took tablets Diltizam hydrochloride 20 mgs b.d. She gave a history of occasional blackouts over the previous ten years. She also described occasional episodes of dizziness followed by loss of consciousness for a few minutes, associated with nausea on regaining consciousness. The last such episode was six months previously. In the past she had an appendicectomy and hysterectomy and she remained otherwise healthy and active. She had smoked 20 cigarettes per day for the last 20 years. There was no history of headache, diplopia, fits or injury etc. After normal CT scan she was seen for ophthalmic consultation by us on 21st December, 1990. On examination she had best corrected visual acuity of light perception in the right eye and 6/9 in the left with generalised constricted field on Humphrey perimetry. There was moderate right relative afferent pupillary defect, and fundus examination showed elevated and blurred disc margin right > left with right disc pallor (Figs. 1a and b). Fundus fluorescein angiography performed two weeks later showed auto-fluorescence in the pre-injection phase and hyper-fluorescence of the disc in the late phase. At this stage the CSF biochemistry was unremarkable and the intra-cranial pressure was recorded to be 13 cm of H\(_2\)O. The various investigations like FBC, ESR, LFT, TFT, U.&E., blood sugar, serum B\(_12\), folate, VDRL, TPHA, auto-antibodies and CT scans were unremarkable.

Fig 1 (a) (b) – At presentation, right and left disc margins are blurred and elevated with right disc pallor.
Fig 2 (a) (h) - One year later showing prominent disc drusen right > left with optic atrophy right and pallor left disc.

Fig 2 (c) (d) - Auto-fluorescence in the pre-injection phase of both discs.

Fig 2 (e) (f) - Ultra-sound B. scan showing disc drusen and calcification.

Visual evoked potentials from the left eye were within normal limits, but none was obtainable from the right eye. Somatosensory potential recordings were within normal limits. The patient was further followed up in December, 1991. The patient had sudden loss of vision in the left eye involving first the inferior nasal quadrant and nasal half of the left visual field which later, in days, affected the central vision also. At this stage repeat CT scan with contrast and 24 hour CSF pressure monitoring was unremarkable. The best corrected visual acuity was no perception of light in the right eye and hand movement shadow in the temporal field of the left eye with prominent optic disc drusen and optic atrophy. (Figs 2a to f).

DISCUSSION

Disc drusen is considered as one of the differential diagnoses in papilloedema\(^5\), but one should note that they can cause transient disc oedema or ischaemic neuropathy as they cause mechanical distortion of blood vessels by their size and location. In chronic atrophic papilloedema,\(^5\) the disc appears yellow-white and focal accumulation of drusen-like bodies may become visible which may lead to diagnostic error. Calcification, however, does not occur, unlike optic disc drusen, and the yellow-white drusen-like bodies disappear when either the intra-cranial pressure is relieved\(^6\) or atrophy advances. Drusen have been reported with chiasmal tumours\(^6\), abnormal visual evoked potential\(^6\) and central retinal artery occlusion\(^7\). Disc drusen and pseudo-papilloedema\(^5,\,9\) are a common cause of neurological consultation because of their association with headache,
migraine\textsuperscript{(6-8)}, convulsions, visual field loss\textsuperscript{(9,10)}, and transient visual obscurations\textsuperscript{(6,11,12)}. Sudden loss of central vision due to drusen is not common, but a few cases have been reported. There is evidence that optic disc drusen may induce more or less severe circulatory disturbances in the vascular system of the optic disc\textsuperscript{(13,14,15)}. The drusen themselves are hard unyielding concretions that may compress adjacent vessels\textsuperscript{(16)}. These haemodynamic disturbances have been shown to result in ischaemia of the optic disc\textsuperscript{(16)}. Drusen of the optic disc are, therefore, an important cause of compression anterior ischaemic optic neuropathy.

A review of previously reported cases\textsuperscript{(19,20,21)} and the presentation of our own case indicated that central visual loss due to ischaemic optic neuropathy associated with optic disc drusen seems to follow a characteristic pattern.

In our present case the field loss affected the inferior nasal quadrant first, followed by loss of central vision in days to weeks with concomitant afferent pupillary defect. This is followed by optic atrophy and remaining island of vision in the temporal field. Both the eyes had a similar pattern of visual loss within a 12 month period. Thus in view of this typical presentation and the persistently negative results for any other intra-cranial or systemic pathology, we believe that in our patient drusen of the optic disc were responsible for loss of central vision.

REFERENCES
