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A RARE CASE OF SPONTANEOUS SUB RETINAL HAEMORRHAGE ASSOCIATED WITH IDIOPATHIC INTRACRANIAL HYPERTENSION

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Abstract

***Introduction**: Idiopathic Intracranial Hypertension (IIH)is defined as increase in intracranial pressure due to unspecified causes leading to headache, papilledema, and transient vision loss. Usually the patients present with mild visual loss but about 25% may develop permanent visual loss due to irreversible optic disc damage. One rare cause of severe visual loss is subretinal hemorrhage (SRH) due to underlying Subretinal Neovascular membrane.*

***Case Report**: We report a case of a 36-year-old man diagnosed with IIH and papilloedema with sudden onset profound visual loss in his right eye. On examination, there was a large peripapillary SRH superior to the disc and involving the macula.We stress the importance of recognizing this uncommon complication of papilledema and discuss the possible causes for developing SRH, its most common outcome and the diagnostic as well as treatment modalities available.*

***Discussion**: We stress the importance of recognizing this uncommon complication of papilledema and discuss the possible causes for developing SRH, its most common outcome and the diagnostic as well as current treatment modalities available*

***Conclusion**: This report highlights a rare case of spontaneous sub retinal haemorrhage in a patient with IIH. Measurement of optic nerve sheath diameter can be a vital tool for the diagnosis of raised intracranial pressure in this setting.*

Keywords: Papilledema, Idiopathic intracranial hypertension, Sub retinal haemorrhage, spontaneous sub-retinal haemorrhage.

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INTRODUCTION

Subretinall hemorrhage (SRH) refers to the blood between the neurosensory retina and the retinal pigment epithelium (RPE). SRH can arise directly from the choroidal neovascular membranes (CNV), which grows through breaks in Bruch's membrane. [1] Occurrence of bilateral SRH in patients with pseudo tumour cerebrii is a rare occurrence and can present with sudden onset visual loss.

We bring to light the case of bilateral subretinal haemorrhage in a patient with Idopathic intracranial hypertension presenting with papilledema. [2]

CASE REPORT

A 36 year old male presented to us with sudden onset of painless decreased vision in the right eye three days ago. He also complained that his decrease in vision predominantly affected his central visual field rather than the peripheral field. He had recently been diagnosed with type two diabetes mellitus. There was no history of hypertension or any other pre-existing ophthalmic condition. There was no history of headache or trauma. There were no neurologic symptoms.

The visual acuity in the right eye was counting fingers at two meters and in the left eye, 6/6 by Snellens chart testing . Ishihara's test for colour vision was performed and patient had colour vision defects in right eye- was able to read 2/38 plates .Contrast sensitivity was affected in the right eye (5%). On finger confrontation test, decrease in central visual field of the right eye was noted. Extra ocular movements showed no restriction and were painless. The pupil of the right eye exhibited RAPD Grade 3. Fundus examination of the right eye showed a raised hyperaemic disc with blurring of the disc margins on all sides. A large peri-papillary sub retinal haemorrhage was seen extending 3 disc diameter superior to the disc, and 2 disc diameter temporal to disc involving the macula, 1 disc diameter inferior and nasal to the disc. Retinal striae were seen extending supero-temporally from the disc.(Figure1) VEP recorded prolonged latency of p-100 wave in the right eye. The anterior segment examination in left eye was normal. Fundus examination of the left eye was relatively normal except disc hyperemia and blurring of disc margins.(Figure 2) B scan showed thickening of the optic nerve head, widening of the optic nerve head shadow and an anechoic vitreous cavity in the right eye. In the left eye widening of the optic nerve head shadow was noted .Optic nerve sheath diameter measured 3mm behind the globe was measured to be 8mm and 7mm in the right and left eye.(Figure 3) OCT scan was done which showed large sub retinal haemorrhage involving macula and extending superiorly above the macula in the right eye.The retinal thickness was 397 um at fovea, 418um at the highest point and 307um on an average.(Figure 4)

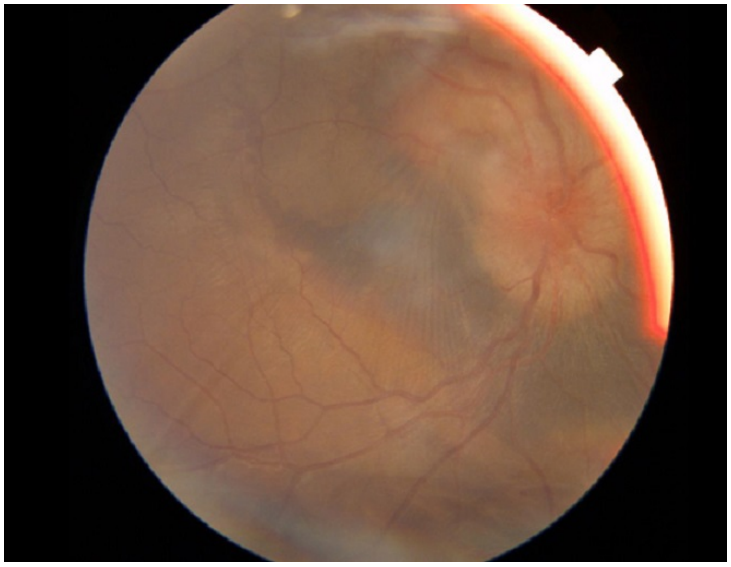

Figure 1: Colour fundus photo of the right eye showing A large peripapillary sub retinal haemorrhage.

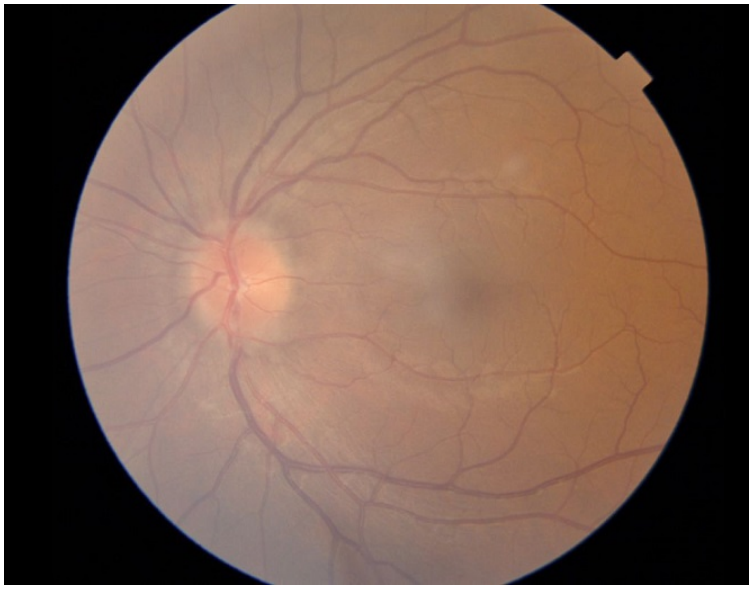

Figure 2: Colour fundus photo of the left eye showing disc hyperaemia and blurring of disc margins

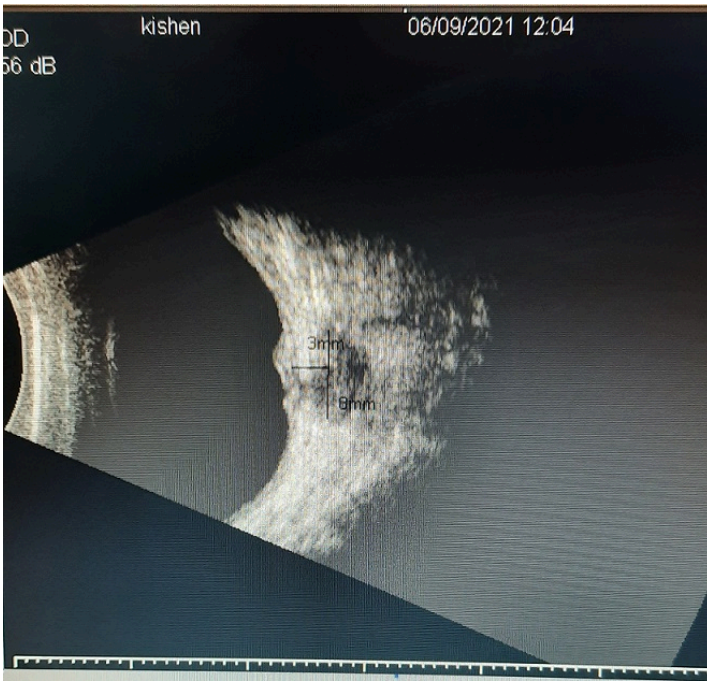

Figure 3A Optic nerve sheath measurement of the right eye using ultrasound B scan.

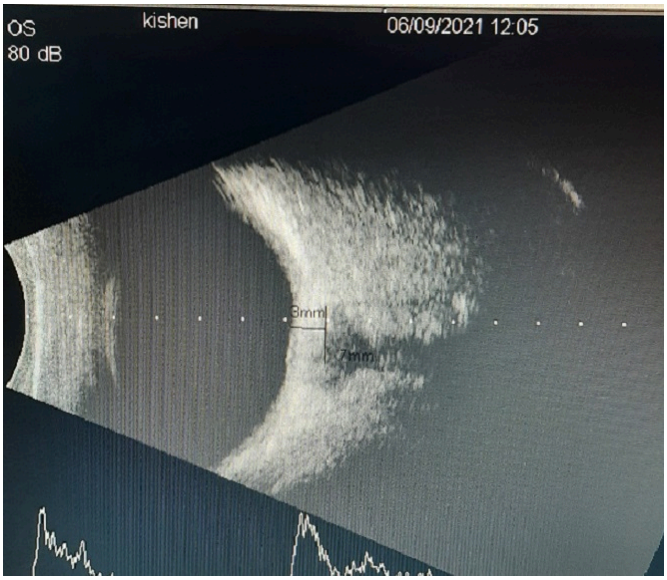

Figure 3B. Optic nerve sheath measurement of the left eye using ultrasound B scan.

The patient was subjected to a complete neurologic examination by a neurologist which was within normal limits. The patient was then admitted to the neurology department for further evaluation. Laboratory investigations were normal including complete blood count, coagulation profile, peripheral smear, serum creatinine and electrolytes Plasma homocystine levels were found to be moderately elevated- 24.6 umol/L. MRI of brain and orbit with MR venogram was reported to be normal. Lumbar puncture demonstrated opening pressure of 30 cm of H2O, the normal CSF opening pressure in males being < 30 cm H2O. [3] CSF evaluation showed a clear colourless fluid with slightly elevated Glucose (79 mg/dL), Protein 57.8 mg/dl and normal chloride level. On microscopy, 100% lymphocytes were noted. CSF was sent for TB CBNAAT as well as cryptococcal antigen which were negative for the same. ANA IF, anti NMO /Anti MOG antibodies was also negative. He was diagnosed with Idopathic intracranial hypertension and treated with IV fluids , Acetazolamide tablets and Paracetamol, and discharged. Patient was subsequently lost to follow up as he moved overseas.

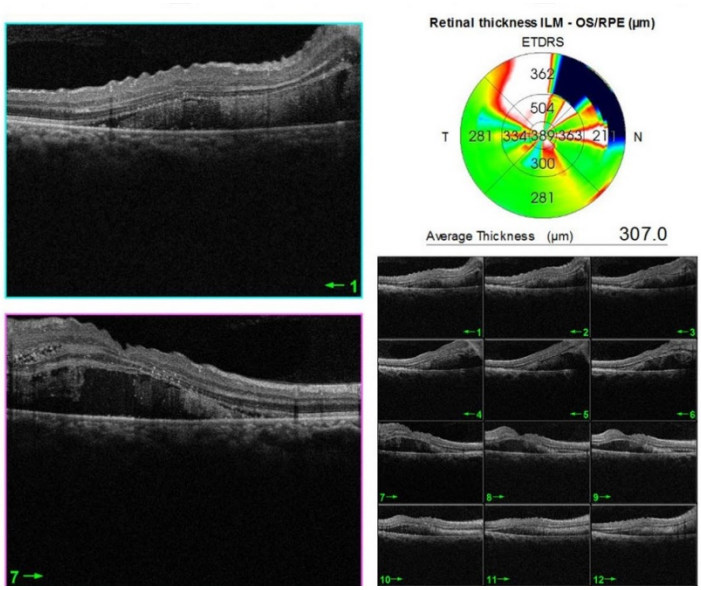

Figure 4: OCT imaging of the right eye

DISCUSSION

Occurrence of spontaneous sub retinal haemorrhage in Pseudo tumour cerebri is rare and fewer than 15 cases overall have been reported describing this condition.[4-12]

The pathogenesis was descrcribed initially by Morese et al, who stated that formation of a sub retinal neovascular membrane ,also known as choroidal neovascular membrane was often antecedent to a sub retinal haemorrhage. The swelling of the axons of the optic nerve head seen in papilloedema casues a mechanical discontinuity in the normal apposition of the chorio-retinal layers and a break in Bruchs membrane . This anatomic dehiscence coupled with hypoxia as a result of the axonal swelling paves way for new vessel formation beneath the retinal pigment epithelium and the neurosensory retina leading to formation of a neovascular membrane.(7)

The inner blood retinal barrier comprises of non fenestrated retinal endothelial cells in contrast to capillary beds of choroidal neovascular (CNMV)membranes which are fenestrated. These fenestrae coupled with the action of Vascular endothelial growth factor (VEGF) facilitates the opening of fenestrations leading to hyperpermeability and the extravasation of macromolecules from the blood vessels. (13) There are other risk factors also which make the CNV prone to haemorrhage. Hypertension is one factor where increased pressure within the lumen can increase the leakage. Microtrauma and Anticoagulant use are other risk factors.(14) Majority of the CNVM are idiopathic.(15) In this patient , sudden visual loss was most likely due to secondary to retinal detachment as a result of haemorrhage from a peripapillary neovascular membrane formation which progressed to involve the macula.

Various treatment modalities have been used for CNVM including argon laser photocoagulation(4,7) standard 3-port vitrectomy(10) and PPV with retinotomy(16). Photocoagulation can only be performed safely when the CNV does not involve the foveola. It also carries the risk of permanent damage to the nerve fibre layer when adjacent to the disk and in the papillomacular bundle. (4,7) Due to the lack of safety and proven efficacy of these methods, newer techniques can be adopted. Displacement of sub retinal haemorrhage involving the macula with a 2step pars plana vitrectomy using tissue plasminogen activator (tPA) and perfluorocarbon liquid (PFCL) tamponade is shown to be a safe and efficient treatment option. (17) Optic nerve sheath fenestration has also been tried, following which the papilledema improved but there was no improvement in visual function(7)

Troost et.al, also suggested that these membranes may not even require treatment due to the fact that they regress spontaneously with the resolution of papilloedema and carry good visual prognosis.(5,11) The value of surgically removing sub retinal hemorrhages to improve visual outcome remains unestablished.(7)

Shirodkar et. Al evaluated the efficacy of optic nerve sheath diameter (ONSD) by ultrasound as a noninvasive method for detecting raised intracranial pressure (ICP). ONSD was measured at 3mm behind the globe.The measurements above 4.6 mm and 4.8 mm in females and males were considered to have increased ICP.In our patient,at 3mm behind the globe, ONSD was measured to be 8mm and 7mm in OD and OS suggestive of raised ICP. Hence this non invasive test can be helpful in the early detection of raised ICP.(18)

CONCLUSION

This report highlights a rare case of spontaneous sub retinal haemorrhage in the setting of IIH. Measurement of optic nerve sheath diameter can be an invaluable cost effective method for the diagnosis of raised intracranial pressure. It is yet unknown whether surgical intervention in sub retinal haemorrhages can improve visual outcomes.

Further research on the efficacy of surgical intervention in non AMD related SRH is warranted.

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