Case report

RARE CASE OF MISDIAGNOSED RATHKE POUCH CLEFT LEADING TO SEVERE VISUAL LOSS

Abstract :- We report a rare case of symptomatic RCCs leading to severe visual compromise in a young 45 yr/f who presented with progressive painless loss of vision in BE a year ago, the patient noticed DOV in the LE and later in RE for which she consulted ophthalmologists before, but was diagnosed as optic neuritis and taken treatment for same,but vision didn't improved. . At presentation, the vision in the BE was, counting fingers at 2 meters. Pupillary reactions were normal, however there was bilateral disc pallor and rest fundus was normal. On systemic evaluation patient gave history of irregular mencus since 3-4 months. on further investigation her S. Prolactin levels were markedly raised(59.99ng/ml). MRI showed well defined hyperintense suprasellar lesion. Patient was operated for Right FrontoTemporal craniotomy with decompression of RCCs by a neurosurgeon. 2 months post surgery VA in RE 4/60 and LE is cfcf.

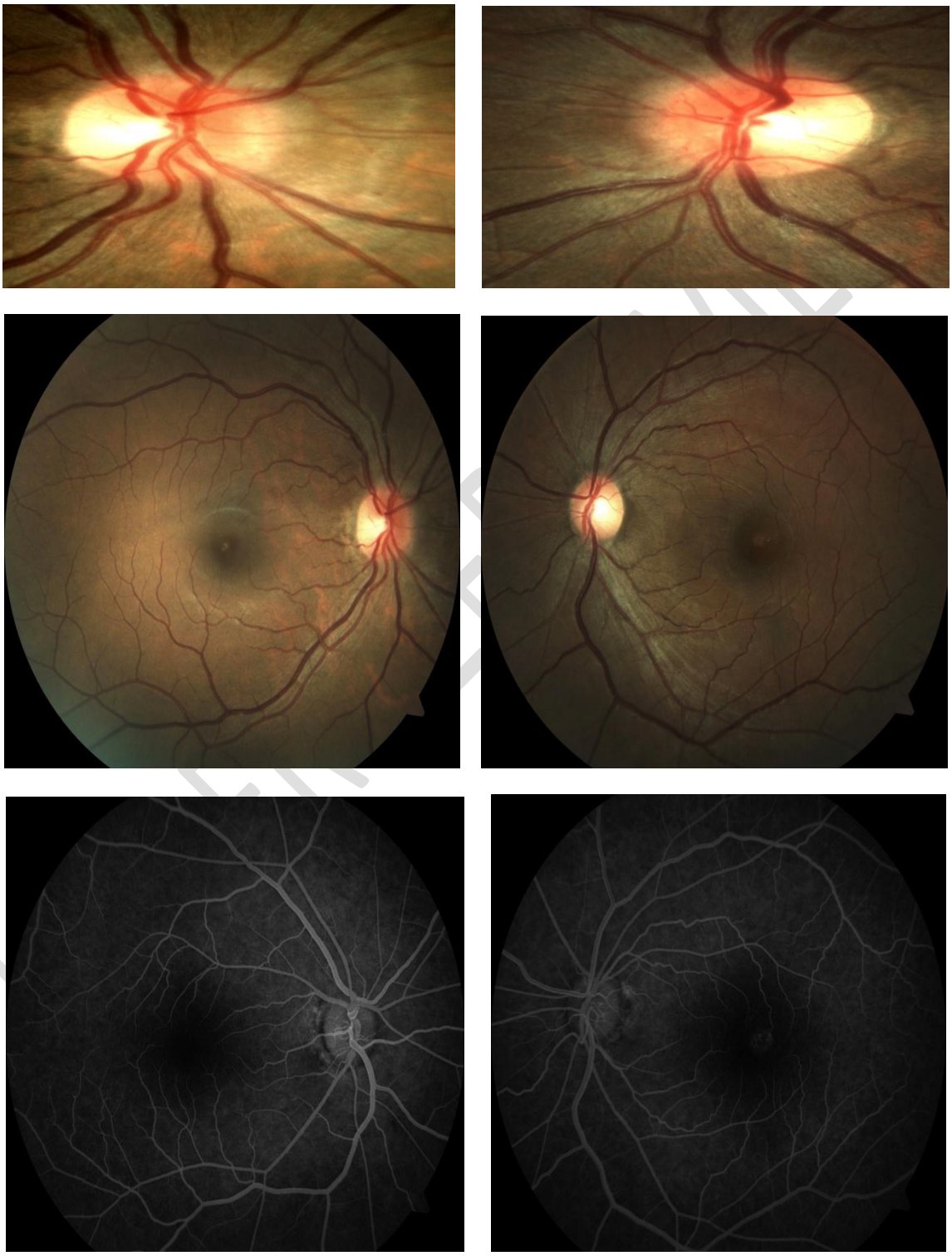
Introduction:- Rathke's cleft cysts are benign epithelial lined intrasellar cysts believed to arise from remnants of Rathke's pouch endoderm(1) or neuroepithelium.(2'3) The cysts are usually small and asymptomatic and have been reported in 2% to 33% of routine autopsies.(5,6) They are generally lined by cuboidal or ciliated columnar epithelium with goblet cells.(5,7) Rathke's cleft cysts that have become large enough to produce symptoms by compression of surrounding structures are rare. However, with modern radiodiagnostic techniques, they are being detected more frequently.(8,9) RCCs are usually asymptomatic, in rare cases secondary to lesional compression visual disturbances, hypopituitarism, headache and dizziness can occur.(1) We present a patient with bilateral gross visual compromise, due to compression by a RCC; and the pituitary hormonal profile was abnormal.

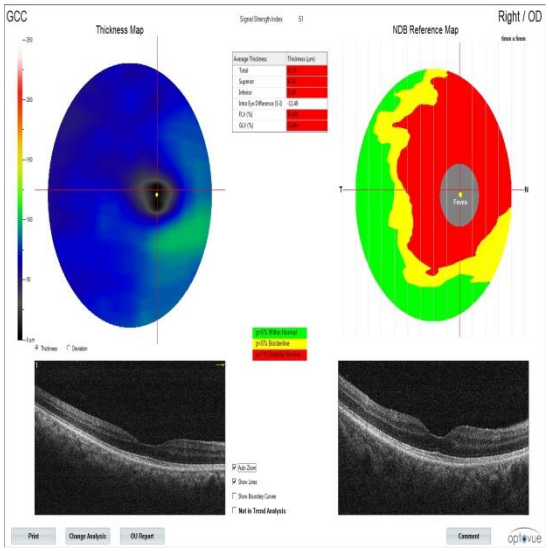
Case Report

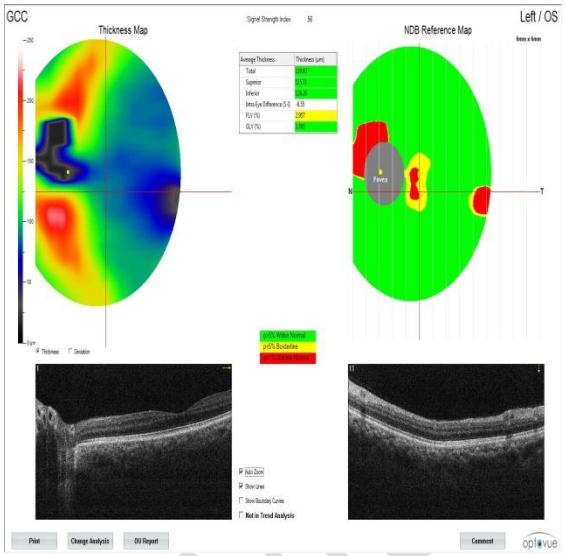
45 years old female presented with progressive painless loss of vision in both eyes. One year ago, the patient noticed diminution of vision in the left eye and later in the right . She had been initially treated with refractive correction, for a few months and then she was treated for multiple sclerosis for which she was started on i/v steroids. There was no improvement in her symptoms, instead loss of vision in both eyes progressed in severity. She did not have any contributory family history or any other co-morbid condition. At presentation, the vision in the both eyes was, counting fingers at 2 meters. Pupillary reactions were normal and equivocal bilaterally, however there was bilateral disc pallor and rest fundus was normal. No history of tobacco use, alcohol intake, drug intake. Higher mental functions were intact with no cranial nerve deficits and no sensory and motor abnormalities. Ganglion cell complex shows gross thinning which was more in Right eye than Left eye. FFA was absolutely normal.Perimetry could not be done as on presentation visual acuity was CF 2 meters. On systemic evaluation patient gave history of irregular mencus since 3-4 months.on further investigation her S.

Prolactin levels were markedly raised (59.99ng/ml). Patient was operated for Right FrontoTemporal craniotomy with decompression of rathke cleft cyst by a neurosurgeon.

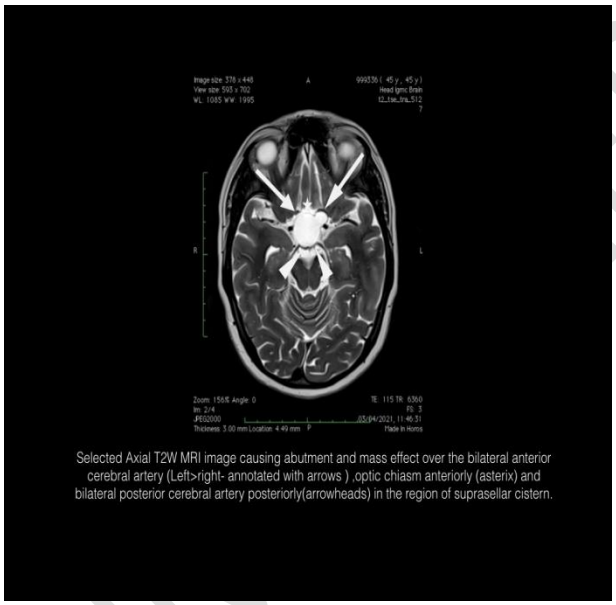
Figure 1 : Ganglion cell complex shows gross thinning







MRI brain (Figure 2) showed a cystic mass with features of a possible Rathke cleft cyst



Selected Axial T2W MRI image causing abutment and mass effect over the bilateral anterior cerebral artery (Left>right- annotated with arrows) ,optic chiasm anteriorly (asterix) and bilateral posterior cerebral artery posteriorly(arrowheads) in the region of suprasellar cistern.

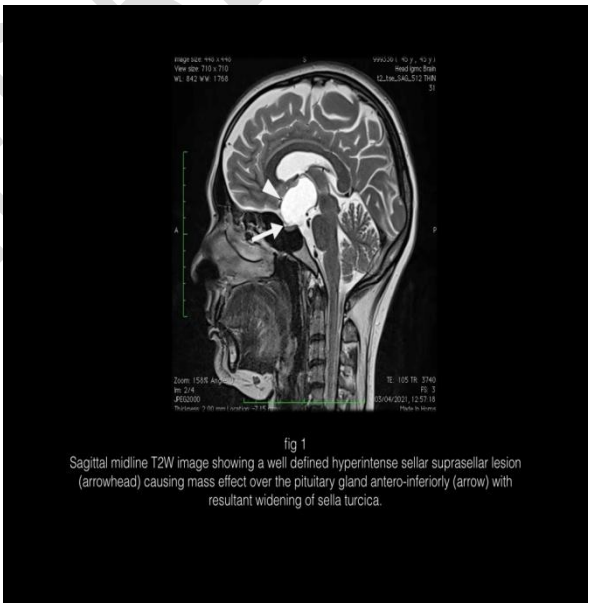


fig 1
Sagittal midline T2W image showing a well defined hyperintense sellar suprasellar lesion (arrowhead) causing mass effect over the pituitary gland antero-inferiorly (arrow) with resultant widening of sella turcica.

DISCUSSION

Symptomatic rathke's cleft cyst is a rare intracranial lesion that has not received much attention in the ophthalmic literature. The condition often produces ocular symptoms and signs. On clinical examination, a field defect, reduced visual acuity, or both. A review of cases of Rathke's cleft cyst, previously published in the neurosurgical literature, found visual disturbance to be an initial symptom in 55.8% of patients.(5) Pituitary dysfunction, including hypopituitarism, amenorrhea-galactorrhea, and diabetes insipidus, was reported in 69.4% of cases reviewed by Voelker, Campbell, and Muller.(5) The only other common initial symptom was headache. Pituitary adenoma and craniopharyngioma(11) were the two

main entities in the differential diagnosis of the patients we reviewed. Other conditions that may be considered in the differential diagnosis include cysticercosis cyst, ependymal cyst, empty sella syndrome, intrasellar aneurysm, and sphenoidal muco-cele. Rathke's cleft cysts occur in an older group of patients (mean age, 40 years) than do craniopharyngiomas (mean age, 24.3 years).(5 11) Visual field defects are seen more commonly with Rathke's cleft cysts.4 . On radiologie examination, suprasellar calcification is unusual in Rathke's cleft cysts.5 In contrast, suprasellar calcification is frequently present in patients with craniopharyngiomas, 79% in one series.(11) Erosion and enlargement of the pituitary fossa may occur in both conditions. Computed tomography usually shows a homogeneous low-density lesion with or without contrast enhancement.(8) In T2 weighted magnetic resonance images Rathke's cleft cysts often appear hyperintense, which may be caused by the presence of mucopolysaccharide within the cyst and can help distinguish them from craniopharyngioma.(9 8,12) In general, Rathke's cleft cysts are intrasellar but may show suprasellar extension. They are lined by cuboidal or ciliated columnar epithelium and contain a gold-colored or white mucinous fluid. It is important to distinguish between these conditions, because a Rathke's cleft cyst requires less aggressive treatment; transsphenoidal drainage of the cyst with partial excision of the wall is effective.(13) Radiotherapy is not required. Rathke's cleft cysts have a much lower incidence of recurrence and a better visual prognosis.(14) Endocrine dysfunction and headaches also improve or resolve in most cases.(13) In conclusion, Rathke's cleft cysts are important because visual disturbance is the most common symptom for which treatment was sought; more than one third of our patients initially came to the Ophthalmology department. Early recognition and treatment may lead to complete reversal of visual field defects.

Conclusion

Rathke's cleft cysts are usually intrasellar or suprasellar or both, and are rarely symptomatic. They have a much lower incidence of recurrence and a better visual prognosis. Endocrine dysfunction and headaches also improve or resolve in most cases. Mri to be done in cases of unexplained vision loss to rule out ICSOL(15,16). Rathke's cleft cysts are important because visual disturbance is the most common symptom for which treatment was sought; more than one third of our patients initially came to the Ophthalmology department. This case highlights that when evaluating vision loss, with a normal posterior segment a meticulous intracranial imaging is very informative to rule out Intracranial space occupying lesions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Reference

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