

## Case Report

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

**Abstract** :- A rare case of symptomatic Rathke cleft cyst resulting in severe visual compromise in a young 45 yr/f who presented with gradual progressive painless diminution of vision in both eyes. Patient noticed diminution of vision in the Left eye an year ago and later on in the Right eye for which she consulted ophthalmologists. She was diagnosed as optic neuritis and underwent treatment for the same, but vision didn't improve. At presentation, vision in Both eyes was same, 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 menstrual periods since 3-4 months. on further investigation her S. Prolactin levels were markedly raised(59.99ng/ml). Radiology showed well defined hyperintense suprasellar lesion. Patient was operated for right frontotemporal craniotomy with decompression of RCCs by a neurosurgeon. 2 months post surgery visual acuity in RE 4/60 and LE is counting finger close to face.

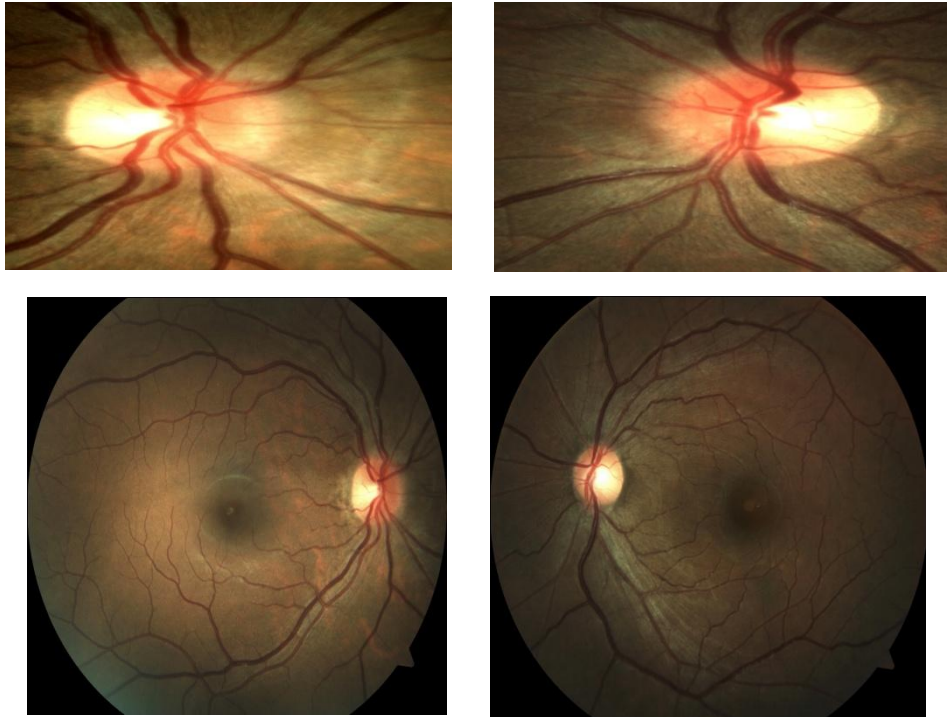
Abbreviations:- Rathke cleft cyst; Ganglion cell complex; Pituitary Adenoma

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

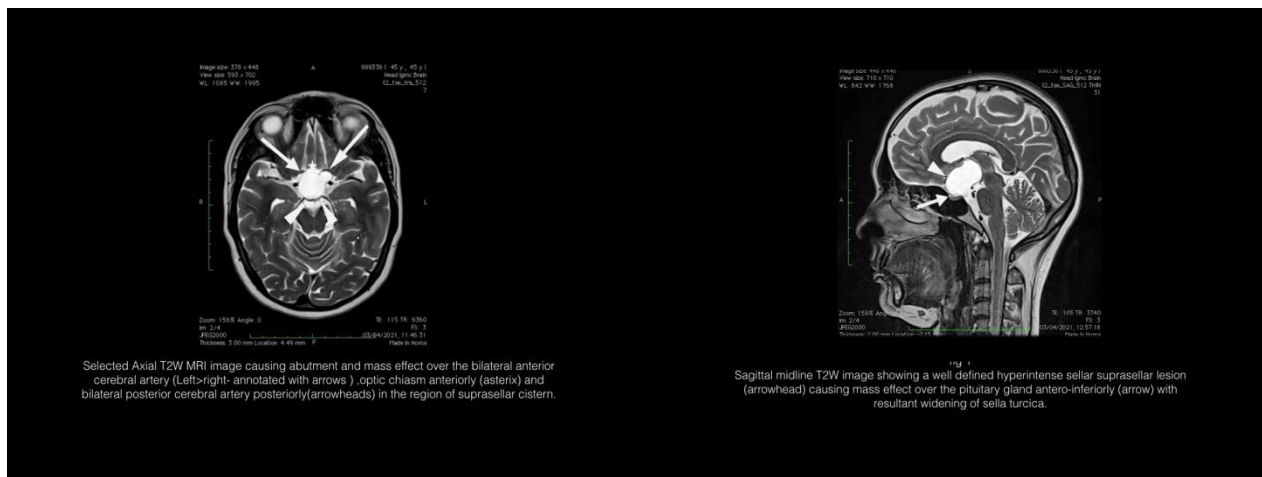
## Case Report

A female aged 45 years presented with progressive painless diminution of vision in both eyes since 1 year, which was first observed in the left eye followed by right eye. She was initially prescribed glasses as part of the treatment. Later on she was treated for multiple sclerosis for which she was put on i/v methylprednisolone. No improvement in symptoms was noted, instead there was a loss of vision in both eyes which had progressed in severity. She had no contributory family history or any other co-morbid condition. At presentation, the vision in both eyes was counting fingers at 2 meters. Pupillary reactions were normal and equivocal bilaterally, however there was bilateral disc pallor, the rest of the fundus being normal.(Figure 1) There was no history of tobacco, alcohol or any other drug intake. Higher mental functions were normal with normal sensory and motor functions. No cranial nerve deficit was demonstrated. Magnetic resonance imaging of brain revealed a cystic mass with possible features of a RCC.(Figure 2) Ganglion cell complex showed gross thinning which was more in Right eye than Left eye.(Figure 4) Fundus fluorescein angiography was absolutely normal.(Figure 3) Visual fields could not be done as on presentation visual acuity was CF 2 meters. On systemic evaluation patient gave history of irregular menstrual periods since 3-4 months. on further investigation her S.Prolactin levels were

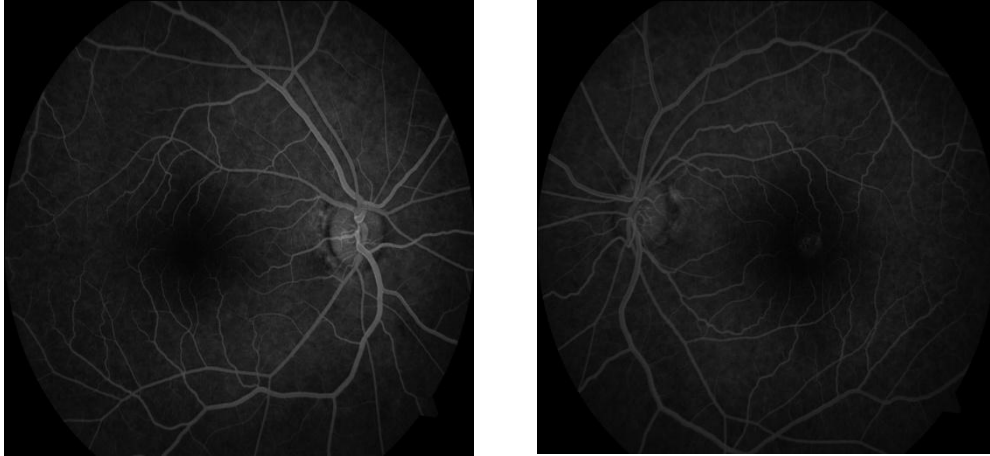
markedly raised(59.99ng/ml). Patient was operated for Right FrontoTemporal craniotomy with decompression of Rathke cleft cyst by a neurosurgeon.



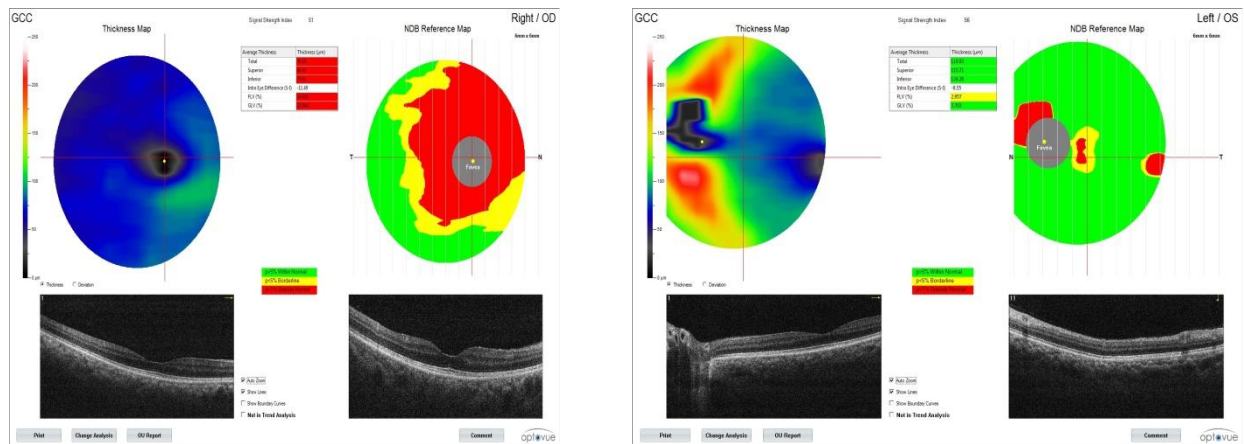
(Figure 1)



(Figure 2) MRI brain indicates features of Rathke's cleft cysts



(Figure 3)



(Figure 4)

## DISCUSSION

“Symptomatic Rcc is a rare intracranial lesion which has till now not received much attention in the ophthalmic literature. On clinical examination signs and symptoms involves a field defect, reduced visual acuity or both. Review of cases of RCCs, which has been previously published in neurosurgical literature, found visual disturbance as the initial symptom in 55.8% of patients” (5). “Pituitary dysfunctions, like hypopituitarism, amenorrhea, galactorrhea, and diabetes insipidus has been reported in 69.4% of cases reviewed by Voelker, Campbell, and Muller”(5). “The other common initial symptom was headache. Pituitary adenoma and craniopharyngioma” (11) “were the two main entities in the differential diagnosis of the patient we had reviewed. Other conditions that can be considered as the differential diagnosis are cysticercosis cyst, ependymal cyst, empty sella syndrome, intrasellar aneurysm, and sphenoidal mucocele. Rathke's cleft cysts usually occur in an older group of patients (mean age, 40 years) compared to craniopharyngiomas (mean age, 24.3 years)”.(5-11) Visual field defects are more commonly seen with Rathke's cleft cysts(4). Suprasellar calcification seen on radiological examination is unusual in Rathke's

cleft cysts(5) which is frequently seen in patients with craniopharyngiomas, 79% in one series.(11) Erosion as well as enlargement of the pituitary fossa can be seen in both conditions. Computed tomography mostly shows a homogeneous low-density lesion with or without contrast enhancement.(8) In T2 weighted magnetic resonance imaging Rathke's cleft cysts often appears as hyperintense, which might be due to the presence of mucopolysaccharide within the cyst and can help in distinguishing them from craniopharyngiomas.(9-12) Generally, Rathke's cleft cysts are intrasellar but can show suprasellar extensions also. They are usually lined by cuboidal or ciliated columnar epithelium containing a gold or white coloured mucinous fluid.It is of utmost importance to distinguish between these conditions, as Rathke's cleft cyst needs a less aggressive treatment; transsphenoidal drainage of the cyst with partial excision of the wall is effective.(13) There is no need of radiotherapy. There is a lower incidence of recurrence and a better visual prognosis in Rathke's cleft cysts.(14) There is also an improvement in Endocrine function and resolution of headache. Early recognition and treatment can result in complete reversal of visual field defects (13)

### **Conclusion**

Rathke's cleft cyst is a benign, sellar, and/or suprasellar lesion which originates from remnants of Rathke's pouch, which mainly disappears in the embryonic period. Most cases of RCC are asymptomatic, while some RCCs can progress to enlarged cysts, which can lead to compression of adjacent structures and thus resulting in clinical symptoms like headache, visual disturbance, and also symptoms of endocrinopathy.(15-17) In symptomatic RCCs, surgical resection provides good symptomatic relief of headaches and visual disturbance, and can even lead to improvement in endocrine dysfunction. After resection the recurrence rates are generally low, on the order of 16–18 %.(18) This case highlights that while evaluating a case with vision loss having a normal posterior segment,a meticulous examination with intracranial imaging is very informative to rule out any intracranial space-occupying lesions.

### **Ethical Approval:**

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

### **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.

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Nil.

### **Conflicts of interest**

There are no conflicts of interest.

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