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Case report

RATHKE'S CLEFT CYST PRESENTING AS BILATERAL OPTIC ATROPHY

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ABSTRACT

PURPOSE: Reporting a case of Rathke's cleft cyst presenting as bilateral Optic atrophy. Optic atrophy could have been prevented with appropriate surgery if the patient would have consulted ophthalmologist earlier. **Case report:** A 23 year-old male presented with bifrontal headache and diminution of vision over 6 months period. On fundus examination right eye showed partial optic atrophy and left eye total optic atrophy. MRI Brain showed well defined lobulated cystic lesion in the sella with suprasellar extension, peripheral rim calcification causing widening of sella and displacing anterior cerebral artery and optic chiasma superiorly. Histopathology of the biopsied material was suggestive of Rathke's cleft cyst. Endoscopic endonasal transsphenoidal sinus surgery was done and biopsy was sent which suggested Rathke's cyst. Following surgery there was improvement in the field of vision in right eye. **Conclusion**: Early diagnosis and removal of a Rathke's cleft cyst has a good prognosis in visual acuity and visual field. Patient ended with optic atrophy because of delayed consultation for headache.

KEYWORDS: Rathke's cleft cyst, Optic atrophy, Headache.

INTRODUCTION

Rathke's cleft cysts (RCCs) are benign, nonneoplastic sellar and suprasellar lesions believed to arise from remnants of Rathke's pouch. Most of the cyst are usually asymptomatic and found only on autopsy findings ^[1]. Symptomatic cysts are very rare. Only around 150 cases have been documented in the literature. Due to rarity and unusual presentation of the disease we are writing this case report.

CASE REPORT

A 23 year-old male presented with bifrontal headache and diminution of vision over 6 months period. His visual acuity was 6/60 with pin hole improvement 6/36 in right eye and perception of light and projection of rays in all quadrants was absent in left eye. Anterior segment was normal. RAPD present in Left eye. On Fundus examination right eye showed partial optic atrophy and left eye total optic atrophy. Perimetry showed Superior Temporal Quadrant field of view present in right eye and No field of view in left eye. Routine Investigation and Pituitary Profile were normal.



Figure 1. (RE) Partial Optic atrophy. (LE) Total Optic atrophy.

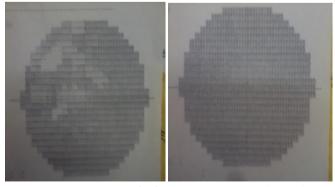


Figure 2. Preoperative (RE) Superio-Temporal field of veiw present, (LE) No field of veiw.

Beladiya et al.,





Figure 3. Post operative. (RE) Temporal Field of veiw - Present, (LE) No field of view.

MRI Brain showed well defined lobulated cystic lesion in the sella with suprasellar extension, peripheral rim calcification causing widening of sella and displacing anterior cerebral artery and optic chiasma superiorly.

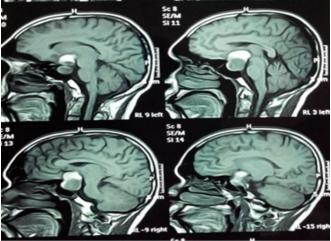


Figure 4. MRI Brain

Patient underwent Endoscopic endonasal transsphenoidal pituitary surgery. Following surgery the patient has noted a subjective improvement in his right eye field of vision. Histopathology of the biopsied material was suggestive of Rathke's cleft cyst.

DISCUSSION

Rathke's cleft cysts (RCCs) are benign, epithelium-lined intrasellar cysts believed to originate from remnants of the Rathke's pouch. RCCs commonly have a round, ovoid or dumbbell shape. These cysts are found during routine autopsies in 13% to 22% of cases [1].

In general, Rathke's cleft cysts are less than 3 mm in diameter and are usually asymptomatic. The most common clinical manifestations of enlarged cysts include headache, hypopituitarism, diabetes insipidus and visual disturbance [2]. Ross et al^[3] reported on data obtained from 43 patients with Rathke's cleft cyst treated by one neurosurgeon. They noted that headache is the most common symptom and that galactorrhea, visual field loss, and hypopituitarisms are the next most common signs. The most common symptom in this study was headache.

In Voelker's report, a retrospective study of 155 patients with symptomatic RCC, the cyst was found in intrasellar and suprasellar locations in 71% of the patients ^[4]. The sella was enlarged in 80%. In our study cystic lesion was found in the sella with suprasellar extension. MRI is the modality of choice in the detection of RCCs ^[5]. Thin-section sagittal and coronal MRI scans should be obtained through the sella.

CONCLUSION

Early diagnosis and removal of a Rathke's cleft cyst has a good prognosis in visual acuity and visual field. Patient ended with optic atrophy because of delayed consultation for headache.

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Beladiya et al.,