

**Silent Intruder: A Rare Case of Rathke Cleft Cyst in a Young Patient**

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**Citation this Article:** Dr. Shyamnith M, Dr. Priya Margaret, Dr. Shanthi Ramesh, Dr. Sundari.S, “Silent Intruder: A Rare Case of Rathke Cleft Cyst in a Young Patient”, IJMSIR - July - 2024, Vol – 9, Issue - 4, P. No. 56 – 58.

**Type of Publication:** Case Report

**Conflicts of Interest:** Nil

**Abstract**

We report a one year old boy child of Rathke’s cleft cyst in the sellar region with bilateral moderate to severe hearing loss, global developmental delay, AGE with some dehydration. MRI Brain scans of patient who presented with the above complaints revealed a cystic mass in the sellar region. In this case, we advised the patient for BERA and Cochlear implantation surgery.

**Keywords:** Rathke cleft cyst, Global developmental delay, hearing loss.

**Introduction:**

Rathke’s cleft cyst is a nonneoplastic epithelial-lined cyst which develops around the sella turcica. It is derived from the remnant of the Rathke’s cleft in the intermediate lobe of the pituitary gland.[1] This is lined by single layer of cuboidal/ columnar epithelium or rarely with pseudostratified squamous epithelium[2]. These are usually small and asymptomatic, rarely become large enough to cause symptoms. Till now, only 155 cases of symptomatic Rathke’s cleft cyst have been reported, of which 11 cases are suprasellar Rathke’s cleft cyst with

normal sella. We present a rare case of Rathke’s cleft cyst.

**Case Report**

A one year-old boy was referred to our department due to complaints of fever, loose stools and vomiting for 3 days, not responding to when being called and global developmental delay. On examination the oral cavity was dry and Anterior fontanelle was slightly sunken. Investigations with magnetic resonance imaging (MRI) showed a cystic lesion in the sella region measuring 10\*6\*5mm. The lesion was reported as Rathke cleft cyst. The child was found to have bilateral moderate to severe hearing loss on OAE and was advised BERA and Cochlear implantation surgery. The child has a past history of hospital for seizure disorder and was advised to do Tandem Mass Spectrometry. The child was asked to be brought for review with the results of BERA and Tandem Mass Spectrometry.

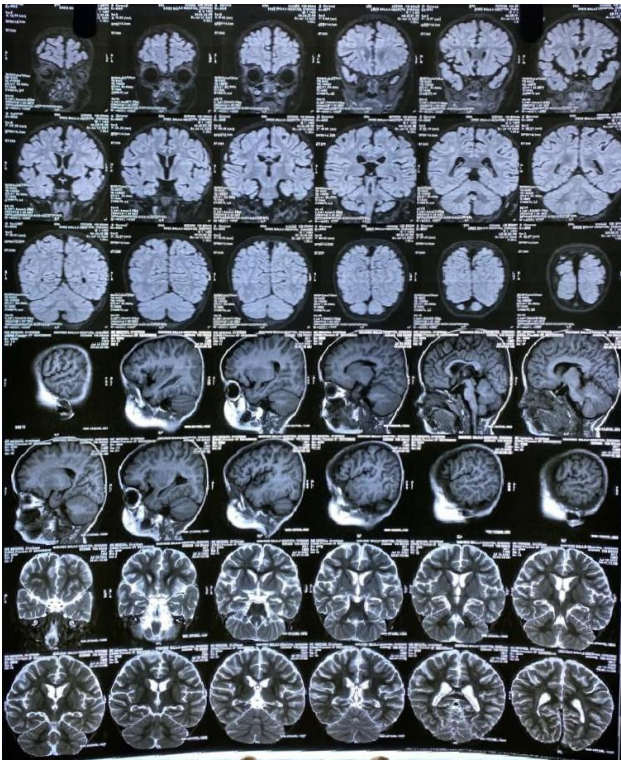


Figure 1

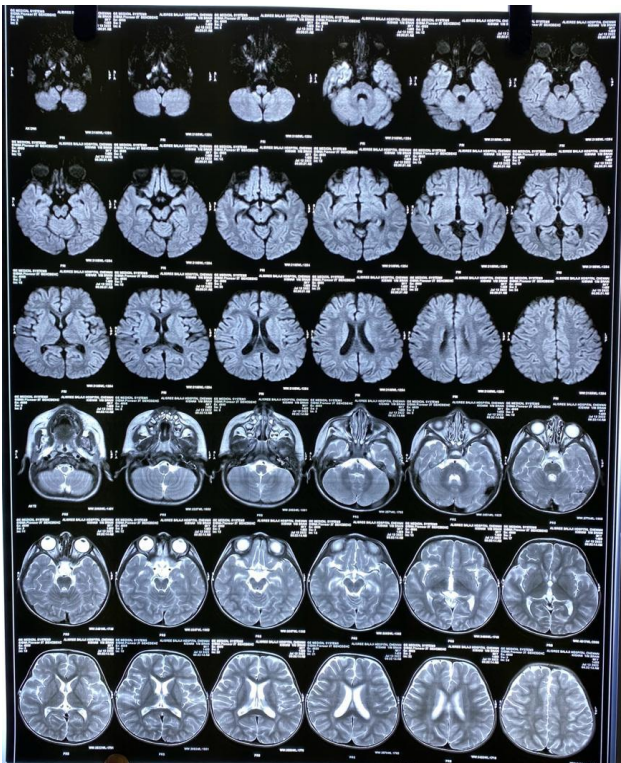


Figure 2

## Discussion

The incidence of the Rathke's cleft cyst has not been clear because most of Rathke's cleft cysts are asymptomatic. The incidence and the number of surgical cases have increased recently due to the advent of MRI. Embryologically, it develops from remnant of Rathke's pouch [3]. Barrows et al., suggested that Rathke's pouch remnant may coexist with anterior pituitary tissue located above the diaphragm. Under these conditions, remnants of Rathke's cleft found above diaphragm sella may give rise to suprasellar and parasellar Rathke's cleft cyst. Isono et al., proposed that neuroepithelium may pinch off to form ependymal lined tubules within the gland and around stalk. The concept would account for mechanism of development of Rathke's cyst outside the sella as in our case. Third concept is of origin from endodermal cell by metaplasia. The histological picture of Rathke's cleft cyst and craniopharyngioma differs greatly, even though they might have common origin [4]. Histologically, Rathke's cleft cysts consist of a single or pseudostratified epithelium with an underlying layer of connective tissue. The epithelium may contain ciliated, goblet, and squamous cells, whereas craniopharyngioma has either adamantinomatous or squamous stratified epithelium invading surrounding brain parenchyma, Nodule formation, keratin formation, calcium deposit, chronic inflammation, and hyaline granule layers. Rathke's cleft cysts are most often small and asymptomatic; occasionally, they may become large enough to cause symptoms by compression of intrasellar and suprasellar structures. The lesion may become symptomatic in children but most reported cases have been in adults. Rathke's cleft cyst are more common in female by 2:1 margin, the most common presenting symptom are pituitary dysfunction, visual field defect, and headache. Hypopituitarism is most common

hormonal abnormality followed by hyperprolactinemia. Aseptic meningitis and pituitary apoplexy are rarely reported.[5] Radiologically, they are located in the midline in and above the anterior portion of the sella turcica with well-defined margin rounded or lobular. Pituitary stalk and gland are usually displaced posterior. Computed tomography (CT) and MRI vary according to morphology of the cyst wall and content of the cyst. These are noncalcified cystic lesion that is important feature that differentiate it from cystic craniopharyngioma. When the cyst lining is of single layer cuboidal or columnar epithelium, it is not clearly seen on imaging studies; however, when stratified squamous epithelium is present or if inflammation occurs in cyst wall enhancement occur on postcontrast studies as in our case. CT scan typically shows an enlarged sella turcica containing a cystic mass, which can be hypodense or isodense with brain.[5] Only symptomatic Rathke's cleft cyst cases require treatment and cyst drainage with marsupialization of cyst is appropriate as recurrence rate of these cyst is very low. Radical excision may result in hypopituitarism and damage to sellar/ suprasellar structures. Since aim is to achieve drainage and marsupialization, these cysts are commonly operated on by transphenoidal route because of low morbidity of this approach.[8] Postoperative adjuvant radiation is also not required as recurrence is very low so just regular follow-up with MRI will suffice.

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