Rathke's cleft cyst

Rathke cleft cysts (also known as pars intermedia cysts) are non-neoplastic, sellar or suprasellar epithelium-lined cysts arising from the embryologic remnants of Rathke's pouch in the pituitary gland.

Epidemiology

They are common lesions and usually incidentally identified.

They are found in 13-33% of the general population and in 11-22% of autopsies. Although the underlying substrate is congenital, they appear to enlarge during life, as they are rare in childhood.

There is a recognised female preponderance with a female to male ratio of approximately 2:1.

Clinical features

When symptomatic, they manifest themselves by compressing adjacent structures, causing pressure effects such as headache, visual disturbance, or pituitary hormone deficits.

The vast majority of Rathke cleft cysts are asymptomatic, and are incidentally found when the pituitary is imaged for other reasons.

A meta-analysis demonstrates that the resection of RCC in patients presenting with headache is associated with headache resolution $^{1)}$.

Pathology

Rathke's pouch forms during the 4th week of embryologic development as a rostral outpouching from the roof of the primitive oral cavity. The anterior wall of the pouch gives rise to the anterior lobe of the pituitary (pars distalis). The posterior wall of the pouch does not proliferate and remains as the intermediate lobe of the pituitary (pars intermedia). The lumen of the pouch narrows to form a cleft (Rathke's cleft) that normally regresses. Persistence of this cleft with expansion is believed to be the origin of a Rathke's cleft cyst.

In Rathke cleft cysts (RCCs), inflammation by the cyst contents infrequently spreads to the

surrounding structures. Calcification, which is regarded as a consequence of chronic inflammation of the cyst wall, can rarely be found in RCCs, and moreover, ossification is extremely rare

The wall of the cyst is typically lined by a single columnar cell layer of epithelium, often containing goblet cells, and is often ciliated. An intraluminal nodule which macroscopically appears white and is often adherent to the cyst wall (although it may be free floating) is composed of solid tissue that represented desquamated cellular debris.

Diagnosis

The increasingly prevalent use of brain imaging modalities such as MRI has resulted in an increase in the incidental discovery of pituitary lesions.

The majority of radiologically diagnosed RCCs remain unchanged or decrease in size over time. These results suggest that, in the absence of pressure symptoms, it is reasonable to manage a patient with RCC conservatively ².

Radiographic features

On imaging a Rathke's cleft cyst is seen as a well defined non enhancing midline cyst within the sella arising between the anterior and intermediate lobes of the pituitary. 40% are purely intrasellar and 60% have suprasellar extension. Purely suprasellar location, although reported, is rare.

СТ

Non contrast: it is typically non-calcified and of homogenous low attenuation. Uncommonly it may be of mixed iso- and low-attenuation, or contain small curvilinear calcifications in the wall (seen in 10-15% of cases).

Post contrast: typically non enhancing although the cyst wall may enhance in some cases 8.

MRI

The signal characteristics vary according to the cyst composition which may be mucoid or serous.

T1 50% are hyperintense (high protein content) 50% are hypointense T2 70% are hyperintense 30% are iso or hypointense T1 C+ (Gd) no contrast enhancement of the cyst is seen, however a thin enhancing rim of surrounding compressed pituitary tissue may be apparent 9-10 In 70-80% of cases a small non-enhancing intracystic nodule can be identified which is virtually pathognomic of a Rathke's cleft cyst. When seen it is hyper intense to surrounding fluid on T1 and hypointense on T2. Depending on the signal of the surrounding fluid it may be inapparent on one or other sequence.

Occasionally, a fluid-fluid level may be seen (particularly if there has been a haemorrhage).

Differential diagnosis

Rathke's cleft cyst differential diagnosis.

see Rathke cleft cyst treatment.

Recurrence

see Rathke's cleft cyst recurrence

Craniopharyngioma

Its relation to Rathke's cleft cyst (RCC) is controversial, and both lesions have been hypothesized to lie on a continuum of cystic ectodermal lesions of the sellar region.

Alomari et al. report on a 7-year-old boy who presented with decreased visual acuity, presumably of at least 2 years' duration, and was found to have a 5.2-cm sellar lesion with rim enhancement. Histological examination of the resected lesion showed a mixture of areas with simple RCC morphology with focal squamous metaplasia and areas with typical craniopharyngioma morphology. Immunohistochemical staining with CK20 and Ki 67 differentially highlighted the 2 morphological components. Testing for beta-catenin and BRAF mutations was negative in the craniopharyngioma component, precluding definitive molecular classification. Follow-up imaging showed minimal residual enhancement and the patient will be closely followed up with serial MRI. Given the clinical and histological findings in the case, a progressive transformation of the RCC to craniopharyngioma seems to be the most plausible explanation for the co-occurrence of the 2 lesion types in this patient ³⁾.

Books

MRI of the Pituitary Gland By Jean-François Bonneville, Fabrice Bonneville, Françoise Cattin, Sonia Nagi

This clinically oriented book will familiarize the reader with all aspects of the diagnosis of tumors and other disorders of the pituitary gland by means of magnetic resonance imaging (MRI). The coverage includes acromegaly, Cushing's disease, Rathke cleft cysts, prolactinomas, incidentalomas, Clinically nonfunctioning pituitary neuroendocrine tumors, other lesions of the sellar region, hypophysitis, and central diabetes insipidus. Normal radiologic anatomy and the numerous normal variants are described, and guidance is also provided on difficulties, artifacts, and other pitfalls. The book combines concise text and high-quality images with a question and answer format geared toward the needs of the practitioner. MRI is today considered the cornerstone in the diagnosis of diseases of the hypophyseal-hypothalamic region but the relatively small size of the pituitary gland, its deep location, the many normal anatomic variants, and the often tiny size of lesions can hinder precise evaluation of the anatomic structures and particularly the pituitary gland itself. Radiologists and endocrinologists will find MRI of the Pituitary Gland to be full of helpful information on this essential examination, and the book will also be of interest to internists and neurosurgeons.

Case series

see Rathke cleft cyst case series

Case reports

2 cases of RCC in sisters who developed a sudden onset of symptoms in a manner similar to pituitary apoplexy. Interestingly, one of them had a very unusual presentation with seizure. Martinez Santos et al. hypothesized that acute symptoms occur due to aggressive intracystic overproduction of mucopolysaccharides (with or without hemorrhage) and a resulting compressive syndrome or local irritation of surrounding structures by spillage of the cyst contents. RCC can be encountered incidentally in family members or may have a familiar predisposition. Since both sisters presented here developed apoplexy symptoms, they proposed a more frequent follow-up with sequential imaging in patients with a family history of RCC. Transsphenoidal surgery with evacuation of the cyst contents is the treatment modality of choice, with excellent outcomes⁴

A 60-year-old woman presented with headaches, fatigue and weight loss due to panhypopituitarism. Magnetic Resonance imaging revealed a mass lesion in the sellar region, which was comprised of two different parts, with Hypointensity anteriorly and hyperintensity posteriorly on T1-weighted image (WI), and the rim with significant Hypointensity entirely on T2-WI. During the transsphenoidal surgery, the cyst wall was so rigid that it was difficult to cut and remove it. The cyst contained mucinous fluid with both old and new hemorrhages, and yellowish, elastic hard, solid nodule. Postoperative histological diagnosis was RCC with unusual lymphocyte infiltration, massive granulation, and mature bone formation. Six months later, the fluid in the cyst re-accumulated and the patient presented with headaches. Removal of the entire cyst wall and the aspiration of the cyst content were performed to collapse the cyst cavity and, consequently, to prevent further recurrence. Postoperatively, panhypopituitarism was unchanged and the symptoms were treated with hormonal replacement. The cyst has not recurred for 2 years after the second surgery.

Persistent, long-term inflammation induced by the RCC content, mucin-containing fluid, and several phases of hemorrhage, presumably promoted the formation of mature bone on the cyst wall and of the elastically solid nodule within the cyst ⁵.

1)

Altuwaijri N, Cote DJ, Lamba N, Albenayan W, Ren SP, Zaghloul I, Doucette J, Zaidi HA, Mekary RA, Smith TR. Headache Resolution after Rathke Cleft Cyst Resection: A Meta-Analysis. World Neurosurg. 2018 Jan 5. pii: S1878-8750(18)30025-1. doi: 10.1016/j.wneu.2017.12.170. [Epub ahead of print] PubMed PMID: 29309984.

2)

Culver SA, Grober Y, Ornan DA, Patrie JT, Oldfield EH, Jane JA Jr, Thorner MO. A Case for Conservative Management: Characterizing the Natural History of Radiographically Diagnosed Rathke Cleft Cysts. J Clin Endocrinol Metab. 2015 Aug 25:jc20152604. [Epub ahead of print] PubMed PMID: 26305621.

Alomari AK, Kelley BJ, Damisah E, Marks A, Hui P, DiLuna M, Vortmeyer A. Craniopharyngioma arising in a Rathke's cleft cyst: case report. J Neurosurg Pediatr. 2015 Mar;15(3):250-4. doi: 10.3171/2014.11.PEDS14370. Epub 2015 Jan 2. PubMed PMID: 25555112.

4)

Hayashi Y, Oishi M, Fukui I, Sasagawa Y, Aida Y, Nakada M. Rathke cleft cyst with entirely ossified cyst wall and partially solid content in the cyst: a case report and literature review. World Neurosurg. 2016 Oct 18. pii: S1878-8750(16)31040-3. doi: 10.1016/j.wneu.2016.10.052. PubMed PMID: 27769949.

From: https://neurosurgerywiki.com/wiki/ - **Neurosurgery Wiki**

Permanent link: https://neurosurgerywiki.com/wiki/doku.php?id=rathke_s_cleft_cyst



Rathke's cleft cyst

Last update: 2024/06/07 02:51