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CASE REPORT

Clinical, Imaging And Pathological Features Of Infected Atypical Rathke's Cleft Cyst With Secondary Pituitary Abscess Formation

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Abstract

Infected atypical Rathke's cleft cyst (RCC) is a rare cause of secondary pituitary abscess. A thirty year old afebrile female presented with a two year history of diabetes insipidus with loss of vision in the left eye over a period of one month. Brain imaging showed a large heterogenous sellar and suprasellar mass with left parasellar extension. The lesion demonstrated a thick, partially calcified wall and a central cystic component with an intracystic nodule. Craniopharyngioma was considered as a likely possibility due to lack of awareness of the imaging features of atypical RCC. However, the typical pattern of calcification and the MR signal intensity of the intracystic nodules can provide valuable clues to differentiate this uncommon lesion from craniopharyngioma and can guide the further clinical and operative management.

Key words: Rathke's cleft cyst, pituitary, sellar, abscess.

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Introduction

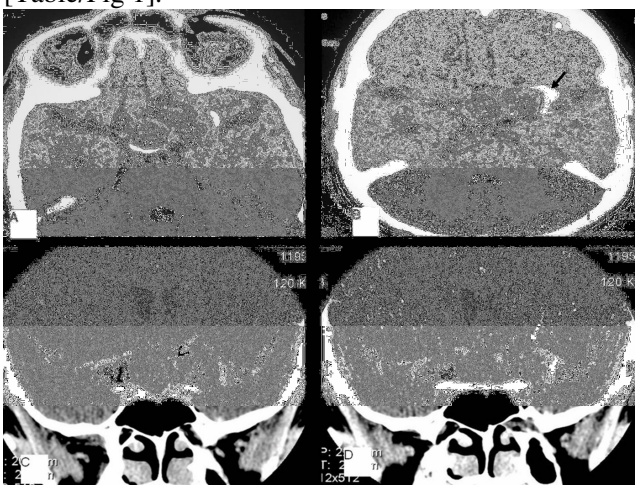
Rathke's cleft cyst (RCC) develops as a macroscopic cyst from the remnant of Rathke's pouch that persists as a cleft between the pars distalis and the pars nervosa of the pituitary gland. [1] Mostly, it is asymptomatic and its typical imaging morphology shows a nonenhancing, thin walled cyst in the pituitary region. Symptomatic cysts are rare and may have atypical imaging features including parasellar extension, cyst wall thickening with enhancement, peripheral calcification and intracystic nodules, when differentiation from craniopharyngioma becomes important due to a different operative approach. [1-4] Abscess formation in RCC as a cause of sellar and suprasellar abscess is only rarely described.[5-7] We describe here, a case of a young female who had atypical imaging features in an RCC along with abscess formation.

Case Report

A thirty year old female presented with a two year history of polyuria and polydipsia and was treated as a case of urinary tract infection. Recently, she developed complete loss of visual acuity in the left eye over a period of one month. The patient was afebrile and had no symptoms of raised intracranial tension. Her total leukocyte count was however mildly raised. Fundal examination revealed partial left optic atrophy.

CT revealed a bifocal lesion, with the sellar and the suprasellar midline component and the left parasellar component. Central attenuation within the midline component was more than that in CSF, while it appeared close to the grey matter superiorly in the left parasellar component. Dense, curvilinear calcification was seen along the margin of the parasellar component. On post contrast CT; it was found that there was thick, partial wall enhancement in the central component and minimal wall

enhancement in the left paracentral component [Table/Fig 1].

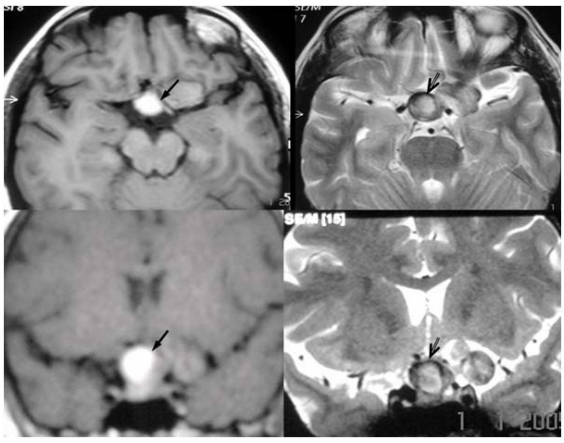


[Table/Fig 1]: (A-D): Pre and post contrast axial CT (A, B) and post contrast coronal CT (C, D) shows the sellar/suprasellar component to be cystic with central attenuation more than CSF with thickened wall and enhancement (open arrow) while left parasellar component appears partially solid (white arrow) and cystic with dense peripheral calcification (black arrow).

Subsequent MRI revealed mixed signal intensity in the lesion. The midline component was hyperintense superiorly and isointense inferiorly on the T1 weighted sequence and on the T2 weighted sequence, it was hypointense superiorly and iso to hyperintense inferiorly. The left parasellar component was isointense to grey matter on T1 and iso to hyperintense centrally with thick peripheral hypointense signal on T2 [Table/Fig 2].

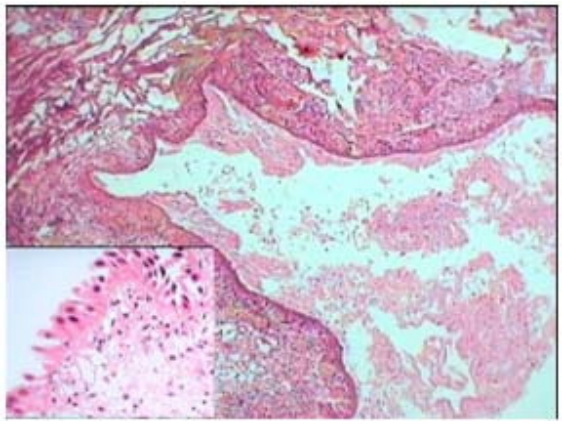
The imaging features of the midline component suggested a thick walled cystic lesion with probably proteinaceous fluid content, while the left parasellar component appeared to be of soft tissue and cystic consistency. These features, along with a calcified wall, favoured a possibility of craniopharyngioma.

Left pterional burr hole craniotomy was done. The wall of the lesion was excised and a yellowish brown pus like material was drained. As the optic nerve on the left side could not be defined separately from the lesion, complete excision of the sac could not be done.



[Table/Fig 2]: (A-D): T1W MR Images (A- axial, C- coronal) shows intracystic nodule superiorly within the central cystic component to be typically hyperintense (black arrows). On T2W images (B- axial, D- coronal) it appears hypointense (open black arrows).

Histopathological examination of the lesion revealed a thickened fibrocollagenous cyst wall lined by pseudostratified columnar epithelium, exhibiting squamoid differentiation at places. There was a dense inflammatory cell infilterate comprising neutrophils, lymphocytes, plasma cells and histiocytes in the deeper tissue, along with focal haemosiderin deposits [Table/Fig 3].

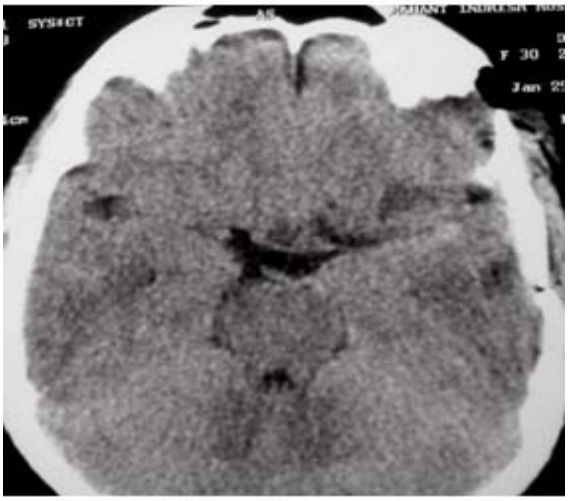


[Table/Fig 3]: Photomicrograph shows thickened fibrocollagenous cyst wall having mixed inflammatory cell infiltrate, congested blood vessels and focal hemosiderin deposits (Hematoxylin and eosin, x 100). Inset shows cyst wall lining of pseudostratified columnar epithelium exhibiting focal squamoid differentiation (Hematoxylin and eosin, x 400).

These findings were suggestive of infected atypical Rathke's cleft cyst with old haemorrhage. The subsequent pus culture was sterile.

The post operative course was uneventful. The patient was put on parenteral ceftriaxone for one

month and was also prescribed desmopressin. Follow up CT after one month showed only a small residual lesion and the patient recovered to finger counting at six feet in the left eye [Table/Fig 4]. The symptoms of diabetes insipidus had also resolved. The patient has still been put on a further long term follow-up to exclude recurrence.



[Table/Fig 4]: Follow up CT after one month shows only minimal residual lesion.

Discussion

Rathke's cleft cyst (RCC) forms part of a spectrum of epithelial cystic lesions in and around the sellar region, with a common cell of origin which is derived from the remnants of Rathke's pouch. Histologically, the lesion spectrum ranges from RCCs (most simple) to craniopharyngiomas (most complex and aggressive). A majority of the RCCs are asymptomatic. Symptomatic RCCs may present with visual disturbances, endocrine dysfunction, raised intracranial tension and uncommonly, diabetes insipidus. [1],[8-11]

On imaging, typical RCCs present as a central, thin walled cystic lesion with a fluid content which is similar to CSF in the sellar and the suprasellar regions. However, atypical features like variable fluid content (ranging from milky white, yellowish or brownish), thick wall, the presence of calcification and parasellar extension lead to a confounding imaging morphology. The imaging features of the cyst will vary according to the predominance of the content (protein, cholesterol or haemorrhage), with high intensity areas on T1 and low intensity areas on T2 weighted imaging, representing a fluid with high protein concentration or haemorrhage. Sometimes, characteristic intracystic soft tissue nodular appearance can be seen, which is hyperintense on T1- weighted images and low signal on T2 weighted images. Retrospectively, this finding was also seen in this case. Pathologically, these intracystic nodules show mucin clumps and biochemical analysis shows cholesterol and protein as the main constituents. The wall of the cyst can be thin or thick. Haemosiderin deposits due to old haemorrhage can also be rarely seen in the wall, as in this case and can lead to the T2 shortening and the blooming effect. Rim like post contrast enhancement can also be noticed with or without the presence of inflammation in the cyst wall. [2-4],[8-13]

Primary pituitary abscess generally occurs within a normal pituitary gland, while secondary pituitary abscess occurs within sellar tumours like adenoma, craniopharyngioma and RCCs. Pituitary abscess may not present with typical systemic symptoms of infection and can only have visual and endocrine symptoms. [5-7]

In this case, the patient was afebrile with predominantly visual symptoms. However, the peripheral smear revealed leukocytosis. On CT imaging, typically an abscess has a thick wall with central attenuation more than in CSF, while on MR, the lesion is homogenously hyperintense on T2 and iso to hyperintense on T1- weighted images(depending on protein content).On post contrast imaging, peripheral enhancement of the lesion may be seen.

However, this case had the atypical imaging morphology, probably due to intracystic fluid containing pus and haemorrhage, along with a thick fibrocollagenous wall containing hemosiderin deposits. In addition, the left parasellar extension of the lesion with coarse left peripheral calcification led to a probable imaging diagnosis of craniopharyngioma. Retrospectively, however, lack of speckled or floccular calcification should have favoured a possibility of atypical RCC. [3] Necrotic pituitary neoplasm and epidermoid cysts are other imaging possibilities, although calcification is uncommon in these lesions. [1-3]

The lesion was approached by the left transpterional approach due to the presumptive preoperative diagnosis of craniopharyngioma, instead of a transsphenoidal approach, which is generally recommended for abscess. [6],[7]

The colour of the pus can be variable and thick yellowish brown material was drained in this case, probably due to haemorrhage and superadded infection. Acinetobacter iwoffi, Staphylococcus epidermidis, Staphylococcus aureus and Streptococcus pyogenes can be isolated from the pus culture, but mostly, the culture is sterile. Histologically, the lesion is characterized by a thickened fibrocollagenous cyst wall which is lined by pseuodostratified columnar epithelium, exhibiting squamoid differentiation at places due to squamous metaplasia. [5-7]

Parenteral antibiotics like vancomycin, ceftriaxone and metronidazole for at least four weeks, are recommended following surgical drainage. [6],[7] Long term follow up is however required to detect recurrence in cases with the histopathological features of squamous metaplasia in the cyst wall. [9],[14],[15]

Conclusion/ Key message

Atypical RCC can be considered in the differential diagnosis of a heterogenous mass in the sellar and the suprasellar regions if the typical patterns of peripheral calcification and intracystic nodules are seen. The knowledge of this lesion may help in planning the correct operative approach. However, histopathology may be required to diagnose superadded infection and haemorrhage.

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