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ABSTRACT

Introduction: Intracranial epidermoid cysts are congenital inclusion tumours. Cerebellopontine angle and parasellar locations are the common locations. This is a report of an intrasellar epidermoid cyst with haemorrhage, which is rare.

Case report: A 70-year-old female presented with bifrontal headache, vertigo, and nasal discharge. Contrast-enhanced Magnetic resonance imaging [MRI] showed heterogeneously enhancing lesion in sella turcica. Internal hemorrhagic foci were seen. Computed tomography [CT] scan showed a slightly hyperdense tumour of sella. Transnasal transsphenoidal excision was done. Hemorrhagic and colloid material came out. Histopathological examination showed cyst lined by stratified squamous epithelium with keratohyalin granules and keratin flakes, suggestive of an epidermoid cyst.

Discussion: Usually epidermoid cyst is hypodense in CT scan. But hyperdensity can occur due to calcification of keratinized debris, increased protein content, and recurrent haemorrhage. Enhancement with gadolinium in MRI is mild and in cyst wall. Haemorrhage and enhancement are probably due to foreign body granulation tissue developing from leakage.

INTRODUCTION

Intracranial epidermoid cysts are congenital inclusion tumors arising from the remnants of epithelial tissue during the closure of neural tube [1,2]. They constitute 0.8-1.2% of intracranial tumors. Cerebellopontine angle is the most common location. But they can be seen in parasellar, suprasellar, middle fossa, and diploic locations. Intrasellar location is rare. MacCarty et al. reported four cases of sellar epidermoid with suprasellar or parasellar extension [3].

CASE REPORT

A 70-year-old female presented with bifrontal headache, vertigo, and nasal discharge. She had systemic hypertension, diabetes mellitus, and dyslipidemia. Neurological examination was noncontributory. Visual fields were normal. Pupillary reaction was normal. There was mild ataxia on walking.

Contrast enhanced Magnetic resonance imaging [MRI] showed heterogeneously enhancing lesion in sella turcica [Fig.1]. Coronal image showed capsular enhancement and thin sellar floor. Internal

Keywords
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hemorrhagic foci were seen. There was no compression on optic chiasm. Computed tomography [CT] scan showed slightly hyperdense tumor of sella [Fig. 2]. Septation was seen in right half of sphenoid sinus.

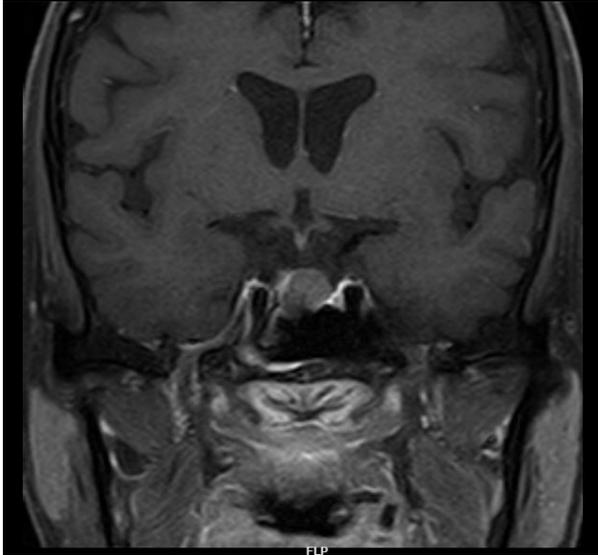


Figure 1. MRI coronal image showing heterogeneous tumor with capsular enhancement and thin sellar floor



Figure 2. CT scan showing hyperdense tumor of sella

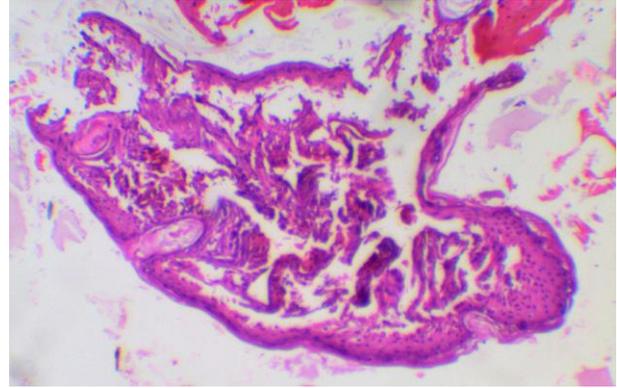


Figure 3. Cyst lined by stratified squamous epithelium and hemorrhage with inflammatory cells

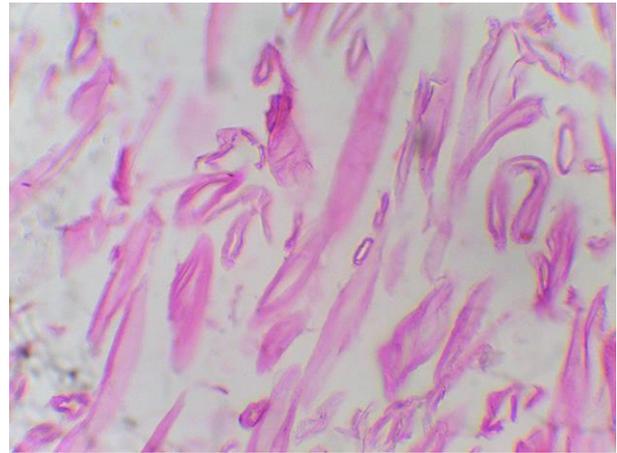


Figure 4. High power view of keratin flakes

Transnasal transsphenoidal excision was done. Left side was chosen because of sphenoid septation on right side. Left sphenoidal ostium was found following middle turbinate. Ostium was enlarged. Dura was opened and piecemeal decompression was done. Hemorrhagic and colloid material came out. Decompression was done till increased urine output was noticed. Fat pad was used for closing the sphenoid.

She recovered clinically. Hourly urine output was in normal range. Hormonal levels such as thyroid function tests, cortisol, and prolactin were within normal limits. Postoperative CT scan showed gross total removal. Initial squash cytology showed fibrocollagenous fragments, cells with round nuclei and eosinophilic cytoplasm, and red blood cells. This was suggestive of inflammatory tissue with hemorrhage. Histopathological examination showed cyst lined by stratified squamous epithelium with

keratohyaline granules and keratin flakes [Fig. 3, and 4]. Immunohistochemistry was positive for p63, suggestive of squamous differentiation. The features are of an epidermoid cyst.

DISCUSSION

The clinical presentations of sellar epidermoid tumors are frontal headache, bitemporal hemianopia, visual loss, diplopia, amenorrhea, galactorrhea, diabetes insipidus, failure of sexual development, and endocrine disturbances [2,3,4,5]. Presentation with features of pituitary apoplexy also is reported⁶.

The usual finding in CT scan is a hypodense lesion, due to lipid and cholesterol content [1,2]. But hyperdensity can occur due to calcification of keratinized debris, increased protein content, and recurrent hemorrhage. Epidermoid tumor appears hypo-, iso- or hyper-intense on T1-weighted MRI⁷. On T2-weighted imaging, it appears hyper-intense. Heterogeneous appearance also is described. Diffusion weighted imaging of MRI shows a restricted pattern as hyperintensity. The cyst appears insinuating into nearby structures. Enhancement with gadolinium is mild and in cyst wall. Differential diagnoses are pituitary adenoma, craniopharyngioma, arachnoid cyst, Rathke's cleft cyst, and dermoid cyst. Pituitary adenomas are usually solid and has homogeneous enhancement⁸. Craniopharyngioma has calcifications in CT and mixed solid and cystic appearances in MRI⁷. Arachnoid cyst is isointense to cerebrospinal fluid (CSF) in all sequences. Dermoid cyst resembles fat and appear hyperintense in T1-weighted image. Because of avascular nature, hemorrhage is rare in epidermoid cyst⁹. Hemorrhage and enhancement are probably due to foreign body granulation tissue developing from leakage.

Surgery is by endonasal transsphenoidal microsurgery or by endoscopic endonasal transsphenoidal approach [2,3,5,6]. Often, the adherence of capsule with neurovascular structures prevented complete removal of the cyst wall. Modification of endoscopic endonasal approach according to the extent of the tumor, can help in gross total removal². Total removal prevents recurrence with malignant change in future.

On histopathological examination, epidermoid cyst is lined by keratinizing stratified squamous epithelium¹⁰. Keratohyaline granules are basophilic

granules in the cytoplasm of granular cells. Keratin flakes also can be seen. The cyst is filled with keratin debris, lipid, and water. Immunohistochemical positivity for p63 is useful in confirming squamous differentiation [11]. Dermoid cyst is lined by simple stratified squamous epithelium¹. Rathke's cleft cyst is lined by simple cuboidal or columnar epithelium with goblet cells. Carcinoma in situ can occur in residual epidermoid cyst as a sequelae of inflammatory response to recurrent rupture and foreign body reaction [1,2].

The sellar tumors which were reported to present with hemorrhage are pituitary adenomas, craniopharyngiomas, epidermoid cyst, undifferentiated sarcoma, tuberculoma, atypical teratoid/rhabdoid tumor, and primary melanocytic tumor [12,13,14,15,16]. So these differential diagnoses also should be considered in case of a hemorrhagic sellar tumor.

CONCLUSION

This is a report of an intrasellar epidermoid cyst with hemorrhage, which is rare. CT scan showed hyperdensity due to hemorrhage. There was enhancement with contrast in MRI. Hemorrhage and enhancement occur due to inflammatory tissue. Transsphenoidal microsurgery and endoscopic endonasal approach are the preferred surgical methods. Total resection is necessary to prevent malignant transformation.

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