Case Report

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Spontaneous Intra-Parenchymal Rupture of Craniopharyngioma – A Rare Phenomenon

Radiology Section

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ABSTRACT

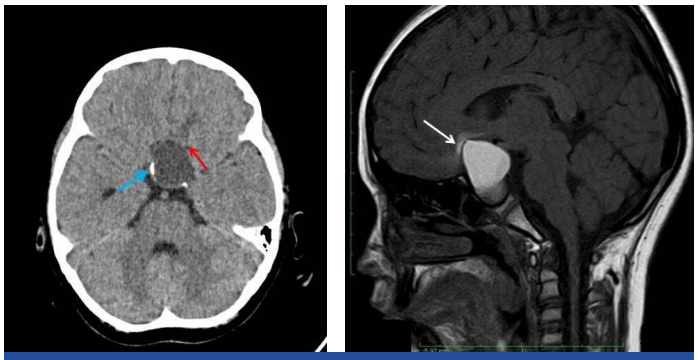
Craniopharyngioma is a relatively benign intracranial tumour that involves sellar and supra-sellar regions. Spontaneous rupture of craniopharyngioma into the sub-arachnoid space or into the ventricles is a rare phenomenon and few cases are reported in literature. We hereby report a case of sellar-suprasellar craniopharyngioma with focal intra-parenchymal rupture into brain causing aggravation of headache in a 12-year-old female child. This complication is a relatively rare phenomenon, which requires attention for early intervention. This was managed by trans-nasal endoscopic trans-sphenoidal surgery. Usually rupture of craniopharyngioma causes chemical meningitis or it can be asymptomatic. Sometimes rupture can cause complete resolution of cyst and symptoms. In the present case, there was only focal rupture and it was into brain parenchyma causing brain oedema with aggravation of headache. Magnetic Resonance Imaging (MRI) in intra-parenchymal rupture of craniopharyngioma has been emphasized in this case report.

Keywords: Chemical meningitis, Cholesterol cleft, Giant cells, Machinery oil of craniopharyngioma

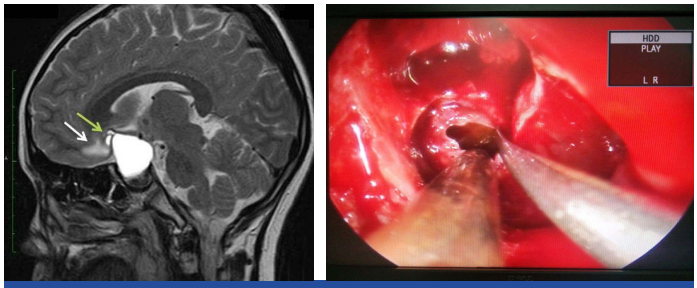
CASE REPORT

A 12-year-old girl presented with complaints of severe headache associated with vomiting for four days. She also gave a history of reduction in vision (Left>Right) and occasional mild headache of six months' duration. She had no meningeal signs, fever nor neurological deficit. On examination, she was of short stature and Tanner Stage II [1]. Her ophthalmologic examination revealed a vision of 6/36 in the left eye and 6/6 in the right. Her lateral skull radiograph showed an enlarged sella turcica. Non-contrast enhanced CT showed a cystic lesion measuring 35 x 26 x 22 mm in supra-sellar region with presence of peripheral curvilinear calcification and oedema in left basifrontal region adjacent to cyst [Table/Fig-1]. Contrast enhanced CT revealed a larger peripherally enhancing cystic component in supra-sellar region with mild enhancement of intra-sellar solid component. T1 weighted MR imaging showed hyperintense cyst in supra-sellar location with isointense solid component in sellar location. There was also a hyperintense signal in the brain parenchyma antero-superior to cyst (left basifrontal region) [Table/Fig-2]. T2 weighted MR imaging revealed a well-defined cystic lesion in sellar-suprasellar location with oedema of left basifrontal lobe and adjacent discontinuity in cyst wall due to intra-parenchymal focal rupture of cyst [Table/Fig-3]. Compression and splaying of optic chiasma and optic tracts were also noted. The history, X-Ray, CT and MR imaging findings were suggestive of sellar-suprasellar craniopharyngioma with focal intra-parenchymal rupture of the cyst antero-superiorly into left basifrontal lobe. This rupture of the craniopharyngioma cyst intra-parenchymally causing excruciating headache is the unique presenting feature of this case.

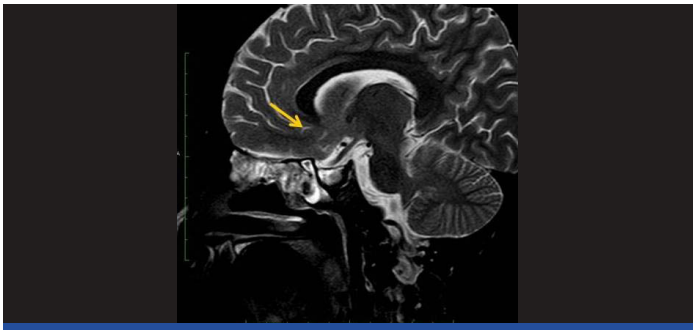
She was hospitalized and started on steroids initially. A full hormonal workup was done (prolactin - 2.54 ng/ml, cortisol - 2.50 µg/dL, growth hormone - 4.4 µg/ml, free T3 - 4.94 pg/ml, free T4 - 6.05 ng/ml) and hormonal replacement therapy was initiated. Patient underwent trans-nasal endoscopic trans-sphenoidal excision of both solid and cystic components [Table/Fig-4]. Postoperatively, the patient recovered well. On seventh postoperative day her headache subsided completely and vision of left eye improved to 6/24. Postoperative MR imaging showed resolution of brain oedema [Table/Fig-5]. The histological examination confirmed the adamantinomatous pattern of craniopharyngioma [Table/Fig-6a,b]. She was later referred to the endocrinologist for further hormonal management.



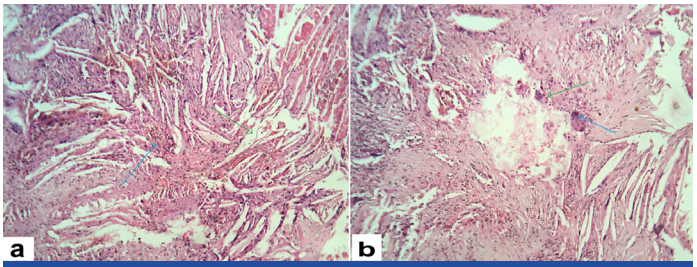
[Table/Fig-1]: Axial NECT showing cystic lesion measuring 3.5 x 2.6 x 2.2 cm in suprasellar region with presence of curvilinear calcification (blue arrow) with mild oedema of left basifrontal lobe (red arrow). **[Table/Fig-2]:** T1WI MR sagittal section showing hyperintense cyst in suprasellar location with isointense solid component in sellar location. Also, note hyperintense signal in brain parenchyma antero-superiorly indicating irritation from leaking cystic fluid due to intra-parenchymal focal rupture (white arrow).



[Table/Fig-3]: T2WI MR sagittal section showing well defined cystic lesion in sellar-suprasellar location with disruption of cyst wall antero-superiorly associated with mild oedema of left basifrontal lobe (white arrow) due to focal rupture (green arrow) antero-superiorly. **[Table/Fig-4]:** Transnasal endoscopic trans-sphenoidal approach of the lesion showing cyst filled with thick oily yellow coloured fluid of machinery oil appearance.



[Table/Fig-5]: Postoperative (trans-nasal endoscopic trans-sphenoidal excision of the lesion) T2W MR sagittal section showing resolution of brain oedema (yellow arrow).



[Table/Fig-6]: Histopathological examination (H&E Staining, x100 magnification) showing: a) Cholesterol clefts (green arrow) and fragments of reactive squamous appearing epithelium (blue arrow); b) Cholesterol clefts associated with multinucleate giant cells (green arrow) and non-caseating granulomatous inflammation (blue arrow).

Study	No. of cases, Age/ Sex	Presenting Symptoms	Site	Management
Russel RW and Pennybacker JB [3]	1 (67, F)	Visual disturbance	Subarachnoid space	Patient expired
Kaemmerer E et al., [4]	1 (46/M)	Headache	Subarachnoid space	Conservative
Patrick BS et al., [5]	1 (21/M)	Headache	Subarachnoid space	Surgical
Scully RE et al., [6]	1 (60/M)	Meningism	Subarachnoid space	Conservative
Ravindran M et al., [7]	1 (12/M)	Intermittent headache associated with vomiting	Intra ventricular rupture	Patient expired
Tokiwa K et al., [8]	1 (43/M)	Visual field defect, transient disturbed consiousness	Subarachnoid space	Surgical
Okamoto H et al., [9]	1 (17/M)	Headache	Subarachnoid space	Surgical
Maier HC [10]	1	Intermittent headache for many years	Rupture into sphenoid sinus	Radiation therapy and conservative
Satoh H et al., [11]	3	Meningism	Subarachnoid space	Surgical
Satoh H et al., [11]	1	Visual disturbance	Subarachnoid space	Surgical
Satoh H et al., [11]	1	Headache	Subarachnoid space	Conservative
Shinohara O et al., [12]	1 (8/F)	Meningism, visual disturbance	Subarachnoid space	Conservative
Shida N et al., [13]	1 (36/F)	Aphasia and drowsiness	Subarachnoid space	Surgical
Kulkarni V et al., [14]	1 (38/F)	Personality change, behavioural disturbance, urinary incontinence, gait ataxia	(Intra-ventricular rupture)-	Surgical followed by radiotherapy
Takahashi T et al., [15]	2 (70/F,69/F)	Headache, visual disturbances	Subarachnoid space	Conservative
Hadden D and Allen I [16]	1 (34/M)	Headache	Subarachnoid space	Patient expired
John-Kalarickal M et al., [17]	1 (61/F)	Headache, blurred vision	Subarachnoid space	Conservative
Yasumoto Y and Ito M [18]	2 (4/F,47/M)	Asymptomatic	Subarachnoid space	Conservative
Kumar A et al., [19]	1 (13 years old child)	Asymptomatic	Subarachnoid space	Conservative
Rajput D et al., [20]	2 (17/F, 16/M)	Headache, vomiting, visual disturbances, meningism	Subarachnoid space	Surgical
Patnaik A et al., [21]	1 (13/F)	Headache, loss of vision	Subarachnoid space	Surgical
Present case	1 12/F	Headache, vomiting, visual disturbances	Intra-parenchymal	Surgical

[Table/Fig-7]: Cases of rupture of cystic craniopharyngioma.

DISCUSSION

Craniopharyngiomas constitute 3%-5% of all primary brain tumours of intracranial origin. Being the most common non-glial brain tumour in paediatric age group they also constitute 50% of all supra-sellar masses in children [2]. Few cases of spontaneous rupture of craniopharyngiomas have been reported in literature. From our review of literature, using keywords “craniopharyngioma” “sellar supra-sellar” “spontaneous rupture” “chemical meningitis” in pubmed and google scholar, 26 cases have been reported on spontaneous rupture of cystic craniopharyngiomas [3-21] [Table/Fig-7].

It may be asymptomatic, associated with chemical meningitis or in some cases reduction in cyst size leading to improvement of neurological symptoms. The ruptured cyst fluid has been reported to cause vasospasm causing infarction. So far no case has been reported in literature on spontaneous rupture of craniopharyngioma into brain parenchyma as seen in our case. Among the reported cases rupture of craniopharyngioma is common among adult males [11]. In our case the spontaneous rupture occurred in a female child. Patient had a history of sudden increase in severity of headache for four days prior to hospitalization which may be due to the spontaneous focal intra-parenchymal rupture of the cyst. Spontaneous rupture of craniopharyngioma into the ventricle or subarachnoid space may cause neurological deterioration. Lateral skull radiography may reveal amorphous sellar and suprasellar calcifications, sellar enlargement, dorsum sella and clinoid erosion. CT and MR imaging have complementary roles in the diagnosis of craniopharyngiomas i.e., CT is superior to MR imaging in the detection of calcification while MR imaging is superior to CT for determining tumour extent and provides valuable information about the relationships of the tumour to surrounding structures. In the present case, intraparenchymal rupture caused severe headache but no neurological worsening, this may be due to focal nature of rupture. Methods of primary treatment include radical surgery, which is gross total resection (complications are hypothalamic injury, endocrine symptoms, vasavasorium injury and pseudoaneurysm) and limited surgery which is subtotal resection along with radiation therapy. Patient had no meningeal signs. Prompt surgery and steroids relieved the headache. The parenchymal oedema resolved in the post-operative MRI indicating the clearance of the irritant. Treatment for residual or recurrent tumour includes surgery, radiation therapy or cyst aspiration, cyst instillation with intra-cavitary radioisotopes, bleomycin or other sclerosing agents. This is the first documented

intraparenchymal spontaneous rupture of craniopharyngioma and prompt surgery is the logical step to reduce the parenchymal irritation and oedema, worsening of which may cause neurological deterioration.

CONCLUSION

We report a case of spontaneous rupture of craniopharyngioma cyst which was focal and into brain parenchyma, without chemical meningitis, which is a very rare phenomenon. This is the first documented intra-parenchymal spontaneous rupture of craniopharyngioma and prompt surgery was the logical step to reduce the parenchymal irritation and oedema, worsening of which may cause neurological deterioration.

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