Intrinsic Third Ventricular Craniopharyngioma: A case report

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Case Report

ABSTRACT

Craniopharyngioma accounts for 2.5-4 percent of all intracranial tumors. The tumor is more observed in the chiasmatic region in adults and the intraventricular subtype is rare. We report an intraventricular craniopharyngioma in a 22-year-old woman presented with chronic headache. Magnetic Resonance Imaging showed hyperintense large mass on T_1-weighted images and hypointense mass on T_2-weighted images in third ventricle with pressure effect on both lateral ventricles and foramen of Monro. The diagnosis of craniopharyngioma was confirmed through histopathological examination of the resected tumor after surgery. After a follow-up period of nine months, neither tumor recurrence nor regrowth occurred. The early diagnosis of this relatively frequent tumor would help to prevent related sequelae.

Keywords: Craniopharyngioma, Headache, Histopathology.

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INTRODUCTION

Craniopharyngiomas are benign partly cystic epithelial tumors, which originate from the squamous epithelial remnants of Rathke's pouch in the subpial space.[1-4] The location of the tumor is determined by embryological events of the suprasellar region.[5] This kind of tumor accounts for 2.5 to 4 percent of all intracranial tumors.[6] Twenty percent of these tumors are located in the sellar (chiasmatic) region in adult,[1] while 5% of them are purely intrasellar.[6] Suprasellar Craniopharyngiomas in 30% of cases extend to the anterior fossa, in 23% to the middle, and in 20% to the posterior fossa and/or retroclival region.[6] Rare ectopic locations include: third ventricle, nasopharynx, pineal gland, sphenoid sinus, and clivus.[6] The intra-ventricular craniopharyngiomas usually present at an older age.[7]

CASE REPORT

A 22-year-old right handed woman was admitted to our institution who presented with chronic headache lasting for 3 years. She had generalized non pulsatile headache accompanied by nausea and vomiting. She had also one episode of generalized tonic clonic seizure.

On the day of admission, she was alert and oriented and neurological examinations were completely normal, except for fundoscopy which revealed mild bilateral papilledema. Non- enhanced computed tomography (NECT) of the head revealed a round and homogeneous hyperdense mass (49 × 54 × 51 mm), below the lateral ventricles, in the third ventricle, which was accompanied by mild dilatation of the bitemporal horns of lateral ventricles due to pressure effect on the foramen of Monro. No calcification was identified in the lesion and posterior fossa and fourth ventricle were unremarkable (Figures 1-3).

Magnetic resonance imaging without injection of contrast media (non enhanced MRI) showed a hyperintense large mass on T_1-weighted and a hypointense lesion on T_2-weighted and fluid attenuated inversion recovery (FLAIR) images in the third ventricle accompanied by pressure effect on both lateral

ventricles and foramen of Monro with interstitial edema due to transependymal leakage of cerebrospinal fluid (CSF) as hyperintensely cloudy like area around the ventricles on FLAIR images (Figures 4-7).

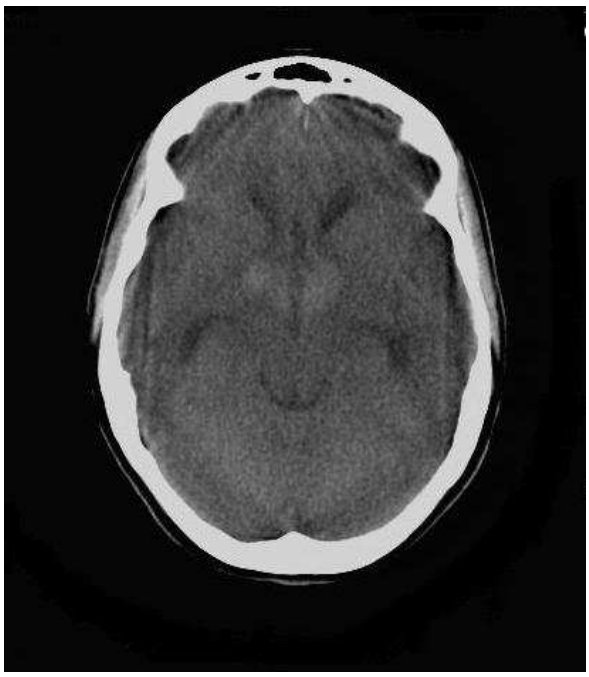

Figure 1. Axial non enhanced CT (NECT) scan of brain (KV: 120, MAS: 60) of a 22-year-old woman with intrinsic third ventricular craniopharyngioma

At the level of temporal horns revealed mid line, homogenous, hyperdense mass below the bifrontal horns with mild obstructive hydrocephaly due to pressure effect on foramen of Monroe.

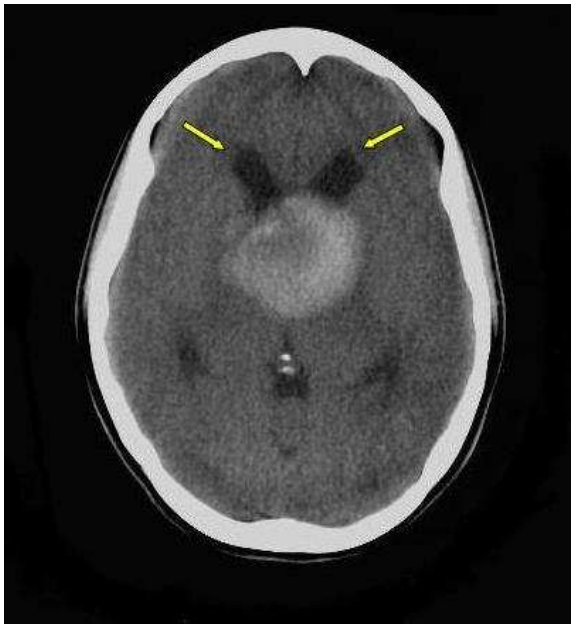

Figure 2. Axial Non enhanced CT (NECT) scan of brain (KV: 120, MAS: 60) of a 22-year-old woman male with intrinsic third ventricular craniopharyngioma
At the level of thalamus revealed midline relatively large homogenous, hyperdense mass in third ventricle with subtle periventricular hypodensity (arrow) due to interstitial edema. Dilatation of frontal horns is due to obstructive hydrocephaly.

Figure 3. Axial non enhanced CT (NECT) scan of brain (KV: 120, MAS: 60) of a 22-year-old woman with intrinsic third ventricular craniopharyngiom
At the level of occipital horns revealed largest dimension of hyperdense mass in third ventricle associated with ventriculomegaly.

Figure 4. Mid sagital T2 weighted spin–echo (SE) magnetic resonance imaging (MRI) of a 22-year-old woman with intrinsic third ventricular craniopharyngioma.
The MRI of brain (Philips intra 1.5 T, TE:100 msec, TR: 3646.8 msec) revealed large round, heterogeneous more prominent hypointense mass placed in third ventricle with obvious bowing of corpus callosum and non communicating hydrocephalus. No pressure effect on optic chiasm was seen.

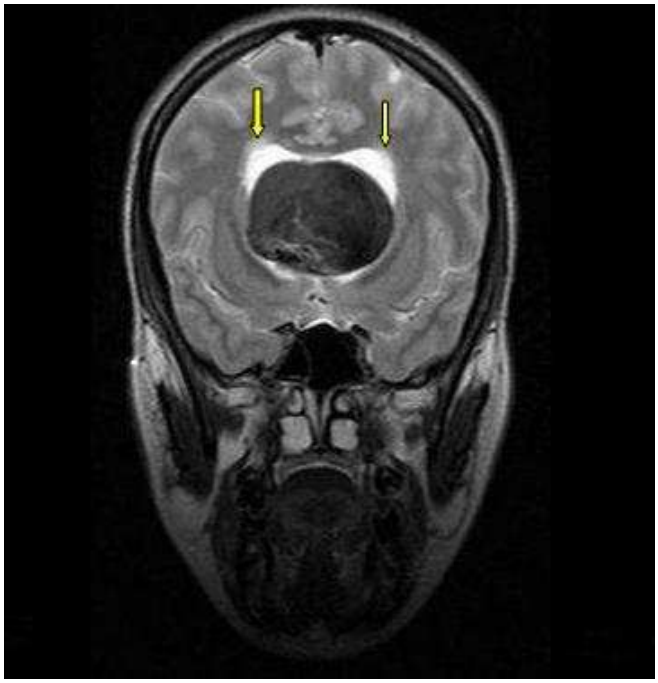

Figure 5. Coronal T2 weighted spin–echo (SE) magnetic resonance imaging (MRI) of a 22-year-old woman with intrinsic third ventricular craniopharyngioma.
The MRI of brain (Philips intra 1.5 T, TE:100 msec, TR: 3641.6 msec) at the level of temporal lobes revealed large midline hypointense mass in third ventricle with dilatation of lateral ventricles and subtle hyperintensity around lateral ventricle due to hydrocephalus (arrow).

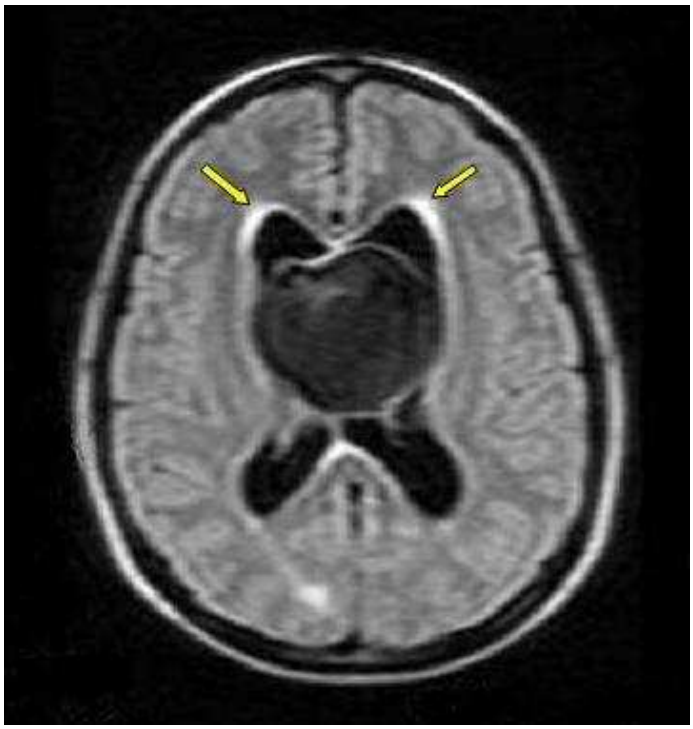

Figure 6. Axial FLAIR image spin-echo (SE) of magnetic resonance imaging (MRI) of a 22-year-old woman with intrinsic third ventricular craniopharyngioma.
The MRI of brain (philips intra 1.5 T, TE:140 msec, TR: 11000.6 msec) at the level of body lateral ventricles showed large midline hypointense mass, with periventricular hyperintensity due to interestial edema (arrow).

Figure 7. Axial unenhanced T1 weighted spin-echo (SE) Magnetic resonance imaging (MRI) of a 22-year-old woman with intrinsic third ventricular craniopharyngioma.
The MRI of brain (Philips intra 1.5 T, TE:15 msec, TR: 486.1 msec) at the level of body lateral ventricles showed large midline, intra ventricular, hyperintense mass.

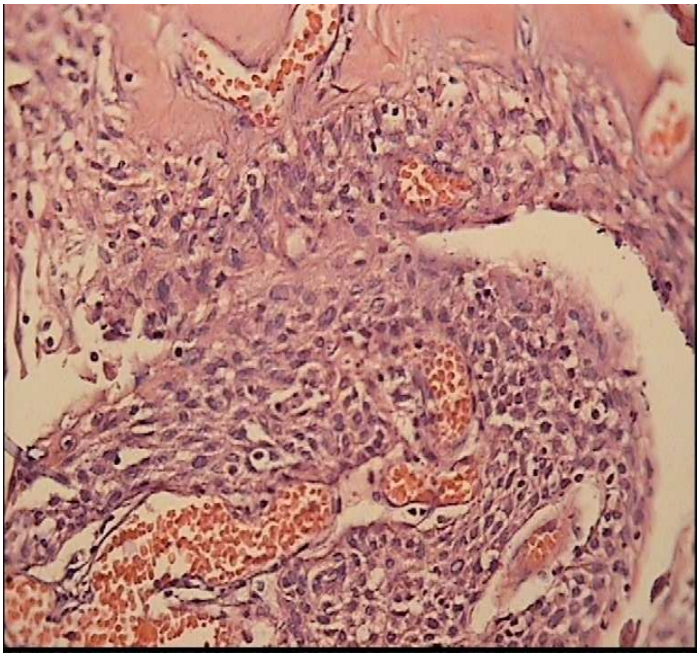

Figure 8. Papillary craniopharyngioma in a 22-year-old woman patient
On H&E staining (Magnification: ×40) it is microscopically composed of solid, well differentiated, pseudopapillary squamous epithelium with separation and desquamation of the epithelium.

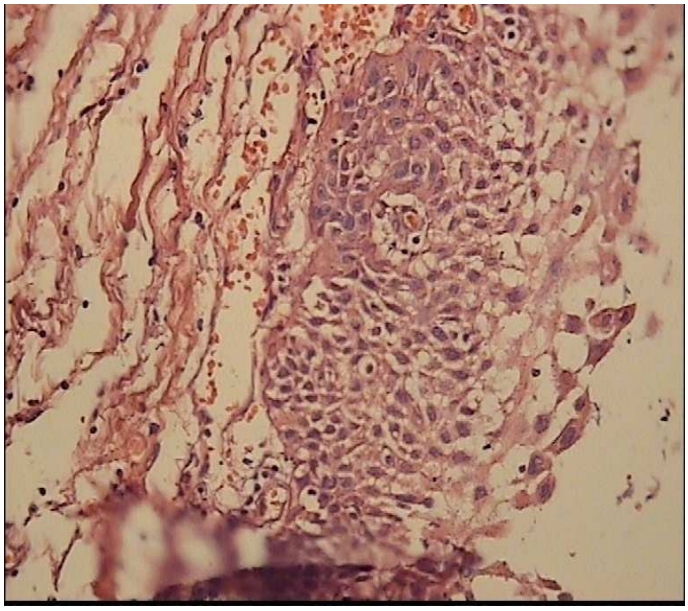

Figure 9. Papillary craniopharyngioma in a 22-year-old woman patient
On H & E staining (Magnification: ×10) it is microscopically composed of solid, well differentiated, pseudopapillary squamous epithelium with separation and desquamation of the epithelium.

Table 1. Information about craniopharyngioma

Etiology	**From remnants of Rathke's pouch epithelium**
Incidence	1.2% to 4.6% of all intracranial tumour
Gender ratio	Men = Women
Age predilection	Bimodal age distribution 5–15 years 40–50 years
Risk factors	Unknown
Treatment	Radical surgery = Gross total resection
Prognosis	Good
Findings on imaging	CT scan finding: Partially Ca++, partially solid, cystic suprasellar mass MRI finding: high signal intensity suprasellar mass on pre contrast T1-W Heterogeneous signal intensity on T2-W T1 C+: Solid portions enhanced heterogeneously

The patient underwent right frontal–precentral–parasagittal craniotomy. The right lateral ventricle was approached through a transcallosal path. The tumor obstructing the foramen of Monro was completely resected. The histological examination revealed a mixed cystic and solid papillary type craniopharyngioma (figure 8,9). After nine months of follow-up, neither tumor recurrence nor regrowth occurred.

DISCUSSION

Craniopharyngiomas are benign partly cystic epithelial tumors, which originate from the squamous epithelial remnants of Rathke's pouch in the subpial space.[1, 4] The location of the tumor is determined by embryological events of the suprasellar region.[5] This kind of tumor accounts for 2.5 to 4 percent of all intracranial tumors.[6] Twenty percent of these tumors are located in the sellar (chiasmatic) region in adult,[1] while 5% of them are purely intrasellar.[6] Suprasellarcraniopharyngiomas in 30% of cases extend to the anterior fossa, in 23% to the middle, and in 20% to the posterior fossa and/or retroclival region.[6] Rare ectopic locations include: third ventricle, nasopharynx, pineal gland, sphenoid sinus, and clivus.[6] The intra-ventricular craniopharyngiomas usually present at an older age.[7] Table-1 summarizes the features of craniopharyngioma.

Behari et al. reported six patients with purely intra-ventricular craniopharyngioma; including 4 patients with cystic lesions and 2 with solid lesions.[8] All of them presented with manifestations of raised intracranial pressure, and papille-

dema. In all patients, the purely intra-ventricular nature of the craniopharyngioma was ascertained on the basis of preoperative MRI. In the above mentioned study, during follow-up period of 8 to 36 months, neither tumor recurrence nor regrowth was detected in any of patients. The symptom of raised intracranial pressure, such as papilledema and visual field defect, were resolved after surgery.[8]

The slow growth of craniopharyngiomas coupled with their location in the third ventricular lumen might delay the encroachment of vital structures and cerebrospinal fluid CSF pathway obstruction for a considerable amount of time. Therefore, they cannot be detected at an early stage, and this might be the reason for the older age at onset of this subtype of craniopharyngiomas. However, when the tumor becomes large enough to obstruct the CSF pathway, the patients present with headache and/or vomiting as the first symptoms.

Visual disturbances with endocrinological disorders which are the presenting symptoms in cases of suprasellar craniopharyngioma are very rare in intraventricular subtype.[7] In addition, calcification which is common in suprasellar craniopharyngioma, is rare in intra-ventricular type tumor.[8]

Colloid cyst, germinoma, lymphoma, choroid plexus papilloma and glioma are the main differential diagnoses of third ventricular craniopharyngioma. In computed tomography

Table 2. Differential diagnosis of third ventricular mass

	Colloid cyst	Germinoma	lymphoma	Choroid plexus papilloma	Craniopharyngioma	Glioma
X-Ray	No findings	May show calcification	No findings	Sutural diastasis	May show calcification	May show calcification and sutural diastasis
Ultrasonography	No findings	May show Hyperechoic intra ventricular mass and hydrocephaly	Usually no findings may shows hydrocephaly	Hyperechoic intra ventricular mass	No findings	May show Hyperechoic intra ventricular mass
CT scan	Hyperdense mass	Hyperdense mass	Hyperdense mass	Iso or Hyperdense mass	CT scan finding: partially Ca++, partially solid, cystic mass	Hypo or hyperdense mass
MRI-T1	hyperintense	Iso or hyperintense	Iso or hyperintense	Iso to hypointense	Often high signal intensity	Hypo or isointense
MRI-T2	Majority isointense	Iso or hyperintense	Iso or hyperintense	Iso to hyperintense	Heterogeneous signal intensity	Iso or hyperintense
MRI-DW1	Does not restricted	May show restricted diffusion	Show restricted diffusion	No reported	Variable depending upon the character of cyst	May show restricted diffusion
Pattern of contrast enhancement	No enhancement	Avid homogenous	Avid homogenous	Avid homogenous	Solid portions enhanced heterogeneously	Variable enhancement
PET	No FDG uptake	FDG uptake	FDG uptake	Increased tumor to normal ratio	May increase uptake	May show increased tumor to normal ratio
Scintography	No uptake	Increased concentration of radioactive isotope above the sella	Detectable with 99m Tc-pertechetate	increase uptake	May increase uptake	Increased uptake in high grade glioma
MRS	No finding	Increased coline Decreased NAA	Increased coline Decreased NAA	Increased coline Decreased NAA	Broad lipid spectrum	May show increased coline

NAA: N-acetylaspartate; FDG: Fluorodeoxyglucose; MRS: Magnetic resonance spectroscopy; PET: positron emission tomography.

(CT) scan all aforementioned tumors could appear as a hyperdense mass, while craniopharyngioma usually appears as a partially solid cystic mass.[1]

Preventive related pointes

The tumor is present at birth, but it may not be symptomatic until childhood or adulthood.

The cause is not totally understood, although it is believed to be primarily a congenital illness.

Beta-catenin gene mutations have been identified to be important only in the adamantinomatous subtype.[9,10]

Duringthe fetal period, ultrasonography, later CT scan, and MRI are regarded to be the most effective tools for diagnosis.[11]

Imaging and differential diagnosis

MRI is the best imaging technique for precise anatomical localization of the intra-ventricular craniopharyngioma, however, no specific signal could be observed.[9] Craniopharyngiomas usually appear as heterogeneous masses of variable intensity(often high signal intensity) on T_1-weighted MRI images and hypointense to mildly hyperintense compared to gray matter On T_2Weighted images.[12] Table 2 shows the differential diagnoses of craniopharyngioma and their patterns on different imaging modalities.

CONCLUSION:

Craniopharyngioma accounts for 2.5 to 4 percent of all intracranial tumors, and is more frequently detected in the sellar region and the intra-ventricular subtype is rare. Craniopharyngiomas appear as heterogeneous masses of variable intensity on T_1- and T_2- weighted MRI images. The early diagnosis of this tumor would help to prevent related sequelae.

Conflict of interest statement: All authors declare that they have no conflict of interest.

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