*Case report*

Child's Nervous System CNS

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Child's Nerv Syst (1992) 8:56–58

# Intrinsic III ventricle craniopharyngioma

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Received September 20, 1990/Revised April 10, 1991

**Abstract.** A case of intrinsic III ventricle craniopharyngioma in an 11-year-old girl is presented. Only five cases in children have previously been reported in detail. The intraventricular location of a craniopharyngioma originates from ectopic remnants and causes intracranial hypertension without hormonal or neurological disorders. The tumor was safely removed by the transcallosal approach, and we confirmed that the floor of the III ventricle was intact.

**Key words:** Pediatric craniopharyngioma – III ventricle – Transcallosal approach

Although the most common sites for craniopharyngioma are suprasellar areas, the tumor can also grow in many directions or in unusual sites, such as the nasopharynx [16, 19], cerebellopontine angle [1], posterior fossa and foramen magnum [21, 26], pineal gland [23], or within the chiasm [6]. The tumor frequently invaginates the floor of the III ventricle. Sweet [27] found that the neoplasm had invaded the ventricular cavity in 23 of the 40 patients operated on. Steno [25], analyzing 30 autopsies, observed 8 cases where the tumor had developed intraventricularly, and Konovalov et al. [13] operated on 85 children with tumors in extra- and intraventricular locations.

Conversely, purely intraventricular cases are rare. Mori et al. [18] found the tumor isolated in the III ventricle in 6 cases out of 155. Sweet [27] presented similar intraoperative evidence in 2 cases. Recently Yasargil et al. [28], reviewing 144 patients who had undergone microsurgical resection, mentioned 6 patients in whom the tumor was confined to the ventricle with few adhesions to the hypothalamus.

Thirty-three cases have been described in detail, including the first one presented as a probable case by Dobos [5] and the 5 most recent cases reported in the Japanese literature by Fukushima et al. [9]. Five of these cases were children: one 11-year-old male was operated on by occipital craniotomy after a ventriculoperitoneal shunt [18]. The second case was operated on at 10 years of age by ventriculocisternostomy and by frontal craniotomy 9 years later [11]. In these cases the tumor was subtotally removed. In the remaining 3 cases, 2 boys and 1 girl (aged 11–15 years), the tumor was removed using the lamina terminalis approach.

In the present case the tumor, which was totally confined to the ventricle, was successfully removed by means of the transcallosal approach.

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## Case report

An 11-year-old girl had been suffering from headaches, nausea and vomiting for 1 month. On admission (24 June 1989) she was somnolent and confused. Neurological examination showed bilateral papilledema and marked bradycardia, but no other neurological changes. General physical examination was unremarkable. Skull radiography was normal with no evidence of calcifications or erosion of the sella turcica.

Computed tomography (CT) scan (Fig. 1) revealed a spherical hyperdense non-enhanced suprasellar mass. The tumor was occupying the III ventricle and the lateral ventricles appeared to be dilated secondary to obstruction of both foramina of Monro. A marked periventricular hypodensity was present.

Surgery was performed the next day. A right free bone flap centered on the coronal suture was removed, the frontal lobe was laterally retracted, and the corpus callosum was opened 2 cm in length, posterior to the genu. The foramen of Monro was found to be greatly dilated and the tumor appeared as a soft greyish-yellow, well-delimited mass, about 25 mm in diameter, occupying the III ventricle. The septum pellucidum was thickened and buckled upwards. After microsurgical dissection of the choroid plexus surrounding the posterior pole of the mass, the tumor, which was not adherent to the ventricular walls, was safely removed. The floor of the III ventricle was absolutely intact. The postoperative course was uneventful and no therapy with cortisone or thyroxine necessary.

The histological examination showed a typical craniopharyngioma (Fig. 2a, b). During long-term follow-up (28 months), no neurological, behavioral or endocrinological disorders were observed. Figures 3 and 4 demonstrate postoperative CT scan and magnetic resonance imaging (MRI).

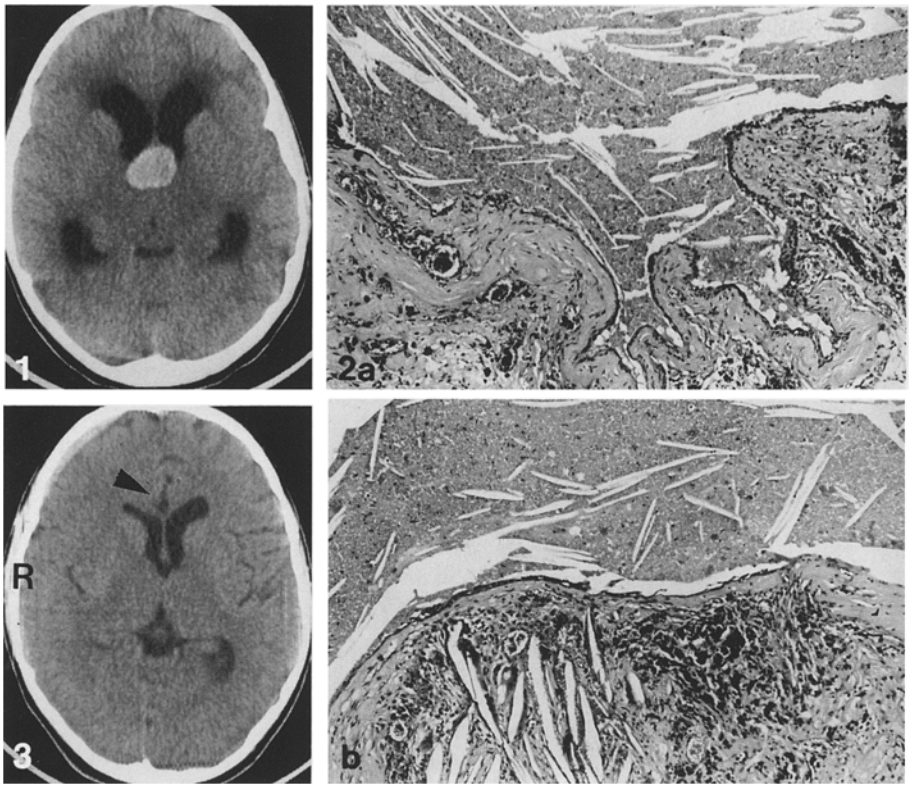


**Fig. 1.** Computed tomography (CT) scan shows a uniformly hyperdense non-enhancing mass occupying the III ventricle with resulting hydrocephalus from blockage of the foramina of Monro

**Fig. 2a, b.** Pathological findings showing the cystic feature of the tumor containing amorphous and necrotic debris areas with elongated cholesterine crystals (at the top of each figure part). The cyst is delimited by flattened pluristratified epithelium surrounded by chronic inflammatory tissue with giant cell reaction to cholesterol clefts

**Fig. 3.** Postoperative CT scan demonstrates the complete removal of the tumor and reduction of the hydrocephalus. The *arrow* indicates the transcallosal approach

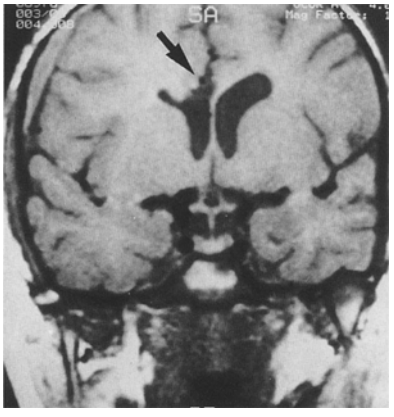

**Fig. 4.** Magnetic resonance imaging 8 months after the operation demonstrates, in addition to the surgical approach (*arrow*), the intact floor of the III ventricle, the patency of the suprasellar cistern and the normal appearance of the pituitary stalk

## Discussion

Craniopharyngiomas intrinsic to the III ventricle are considered to be caused by ectopic remnants of Rathke's pouch [4, 10, 27]. Rush et al. [22] believe that these intraventricular tumors may result from upward growth of squamous cell nests from the floor of the III ventricle near to the tuber cinereum. Konovalov et al. [13] were able to demonstrate microscopic areas of the glial capsule of the tumor in the walls of the anterior part of the III ventricle; these areas represent a possible source of recurrences. Papo et al. [20] emphasized that the origin or craniopharyngioma from Rathke's pouch is still a controversial issue and that there is no clear-cut difference between neuroepithelial cysts with squamous epithelium attached to the floor of the III ventricle and Rathke's cleft cysts.

However, in all the cases described in detail, except in the two reported by Long and Chou [15], an intact hypothalamic floor has been observed. In our case, besides the absence of sellar abnormalities, surgical evidence and MRI clearly demonstrated that the floor of the III ventricle was intact, the suprasellar cisternae patent and the pituitary stalk normal.

An intraventricular tumor causes a syndrome of intracranial hypertension, as occurred in our case, generally without the hormonal and neurological disorders which are usual in suprasellar craniopharyngioma. Hemianopsia and diabetes insipidus were observed in the case reported by King [11] and bitemporal defects were present in the three cases observed by Klein and Rath [12]. King [11] emphasized that in tumors attached to the floor of the III ventricle the lamina terminalis may bulge and the chiasm may be forced downwards towards the tuberculum sellae.

CT does not provide a definite histological diagnosis, even if calcifications or cysts are present [8]. Usually the mass is contrast enhanced [9, 10, 14, 17], but in our case it was hyperdense and non-enhancing, as in the two cases described by Ferrara et al. [7]. Coronal CT and MRI are essential for establishing the intraventricular location of the tumor, and for providing information regarding the surgical approach.

We used the transcallosal approach, which provides easy access to the lateral ventricles and foramina of Monro with minimal retraction upon the brain and very

limited division of the corpus callosum. This procedure allowed us full control and permitted total removal of the tumor. The same approach was used by Long and Chou [15] in four cases, two of them previously operated on using the subfrontal approach, and by Japanease surgeons [9] in another two cases but with only partial removal of the tumor.

On the basis of their experience, many authors [2, 13, 24, 28] have stated that transcallosal and transcallosal interfornicial approaches provide more possibilities for evaluation and radical removal of tumors that impinge upon or enlarge the foramina of Monro, no matter what their nature, with minimal physiological consequences.

Other surgeons have used the subfrontal approach through the lamina terminalis [10–12], the transcortical route [3, 15, 17, 22] or both of these procedures [9]. The lamina terminalis approach offers easy access to the inferior part of the III ventricle and is preferable when the tumor is located in the lower portion of the ventricle and adherent to the floor. The suboccipital approach through the splenium of the corpus callosum has been used in one case [18] in which the tumor was located in the posterior half of the III ventricle, anterior to the pineal body.

Although intraventricular craniopharyngioma remains rare in children, more cases have been identified in the last decade, and our observation further emphasizes the need to consider this tumor in the differential diagnosis.

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