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Case Report

Huge colloid cyst: case report and review of unusual forms

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Summary

The authors report a case of a huge colloid cyst of the septum pellucidum with acute worsening. A 42-year-old man was admitted for unexplained lethargy. A brain CT scan disclosed a huge intra-ventricular lesion.

He experienced a sudden deterioration with coma, and was managed with bilateral ventricular external shunting. Total removal was performed a few days later using a right frontal trans-ventricular approach.

The discussion focuses on colloid cyst potential to mimic other intracranial lesions, rendering accurate diagnosis difficult and leading to possibly inappropriate management in similar cases.

Keywords: Huge colloid cyst; acute worsening; septum pellucidum; unusual forms; review.

Introduction

For the neurosurgeon, colloid cysts (CC) usually imply a single tumour, most often situated in the antero-superior part of the third ventricle [27]. Their specific radiological appearance facilitates diagnosis [33], and they can be successfully treated by several surgical approaches [14, 16, 22, 33, 34]. Accurate diagnosis is not, however, a rule. CC may have atypical locations, inappropriate manifestations and unsuitable disease associations, rendering diagnosis difficult before surgery or histological examination. A huge colloid cyst (HCC) of more than 3 cm is rare [5, 7, 22, 25, 28–30, 34, 35], and represents one of these unusual presentations.

The purpose is to report a case of a HCC with acute clinical deterioration, and to highlight the importance of reviewing these odd forms.

Case report

On November 29 2001, a 42-year-old male patient was admitted for unexplained lethargy. Neurological examination found a drowsy patient with a Glasgow Coma Scale (GCS) score of 11, without other physical anomalies. The patient had been suffering from headaches for several months, and according to his wife, had recently had lapses in concentration and had been forgetful for the past twelve months.

A brain computed tomography scan (CT scan) demonstrated a large, partially calcified, ventricular tumour associated with ventricular dilatation.

The patient's clinical state suddenly worsened with a GCS score of 3, bilateral unreactive mydriasis, absent brain stem reflexes, tachycardia at 140 pulses/mn, and blood pressure of 240/120 mm Hg.

He was intubated and managed with 400 ml of mannitol at 20%, and 300 mg of pentobarbital. External catheters were inserted into both lateral ventricles while the patient was in his ward bed. The cerebrospinal fluid (CSF) was crystal-clear on both sides, and not under any increased pressure. Following this external shunting, the GCS improved to 6. CSF analysis was normal.

His clinical course was favourable, except for the development of aseptic meningitis. He was extubated on December 1st, and while the neurological examination was normal, the patient showed signs of short term memory loss but with no long term memory disturbance. Cerebral magnetic resonance imaging (MRI) revealed a large ventricular tumour, (Fig. 1A–D). The differential diagnosis included craniopharyngioma and central neurocytoma.

Surgery was performed on December 4, 2001. The surgical approach was via the right frontal trans-ventricular route, under magnification. The right inter-ventricular foramen was dilated, and the fornix could be distinguished at the level of the foramen, but no further. The septum pellucidum had collapsed around the tumour. Its upper posterior part was completely calcified. The tumour was not connected to the lateral wall of the left ventricle. The third ventricular portion was easily aspirated. It was limited by a fine translucent membrane, which did not adhere to the third ventricle wall. A total resection was performed.

Postoperative course was marked by left hemi paresis, and significant memory disturbance. A cerebral MRI confirmed complete excision of the cyst (Fig. 2A, B).

At one year follow up, clinical examination showed that the patient had completely recovered from his hemi paresis, and was fully autonomous

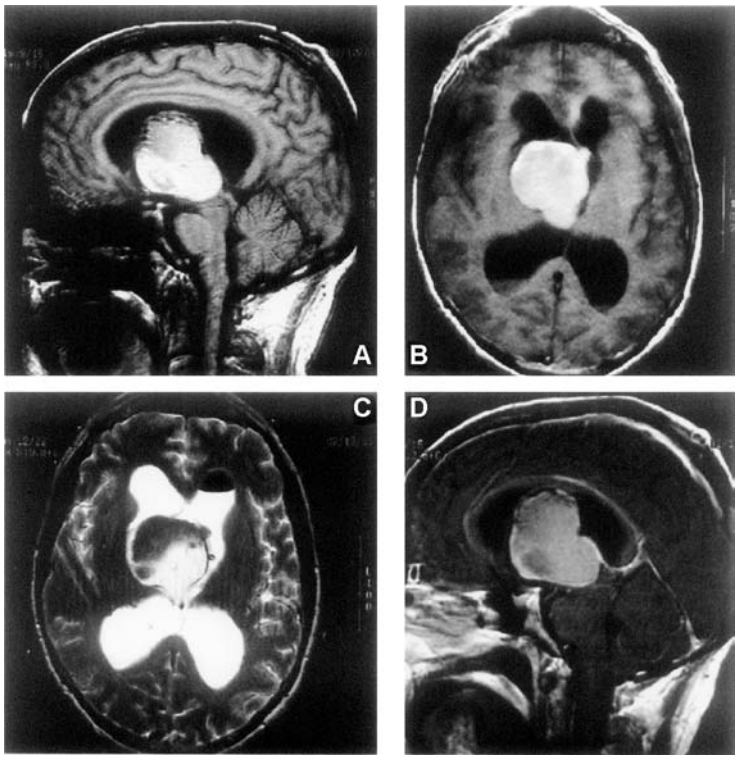

Fig. 1(A). Sagittal T1 weighted MRI shows an intra-ventricular well demarcated mass with high signal intensity. Calcifications are seen at the top of the cyst. (B) Axial T1 weighted MRI shows the intra septal development of the cyst. Note the cyst material at the occipital horns following ventricular puncture. (C) Axial T2 weighted MRI. The cyst appears with two signal intensities, a hypo- and hyper-signal. (D) Sagittal T1 weighted MRI with gadolinium, the cyst does not enhance

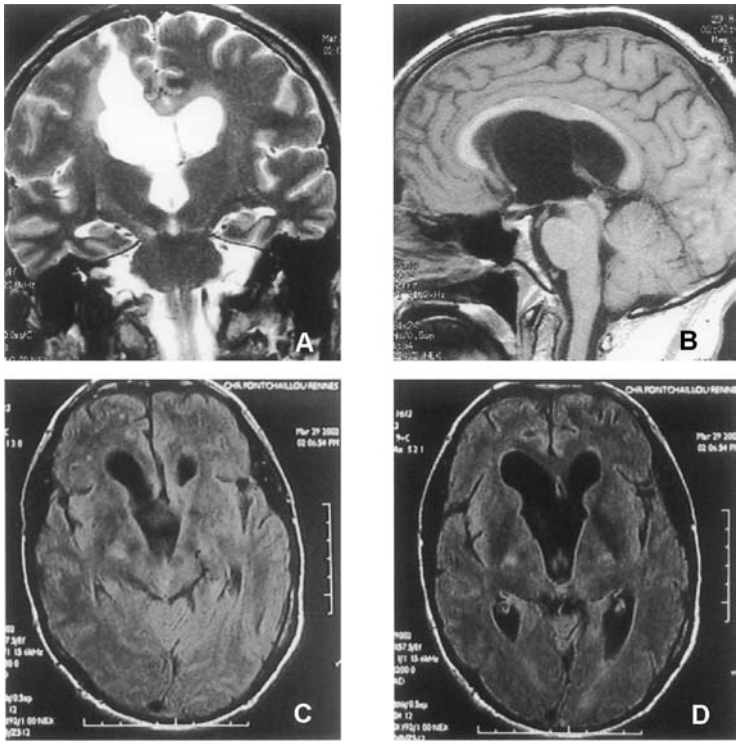

Fig. 2(A, B). Post operative MRI. Coronal T2 weighted MRI (A), sagittal T1 weighted MRI (B), and axial T1 flair (C, D) show a total removal of the cyst, with resection of the middle part of the septum pellucidum and that of the fornix at the foramen of Monro

in his daily activities. However, neuropsychological examination revealed memory disturbance and comprehension disturbance. The patient also had difficulty integrating new experiences and was unable to analyse social facts, interpret the daily news and remember the events of the previous year. The Signoret efficiency score was 33/84, while the average score for the same age and socio-cultural level was 64.3.

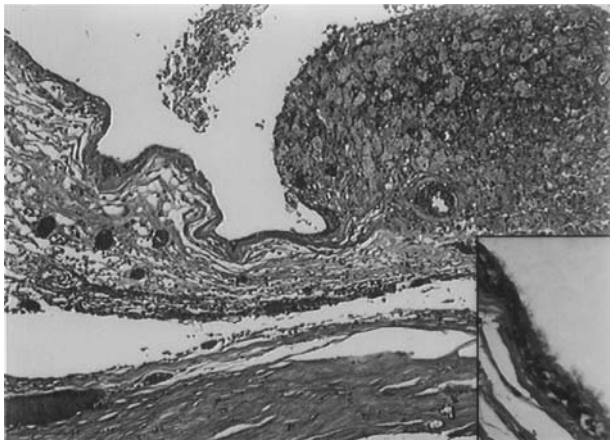

Fig. 3. The cystic cavity, with numerous liphophagic histiocytes, lined by a fibrous stroma, with calcification (HES ×40). Inset: At high magnification (HES ×400), the inner surface of cyst wall is lined with a ciliated epithelium

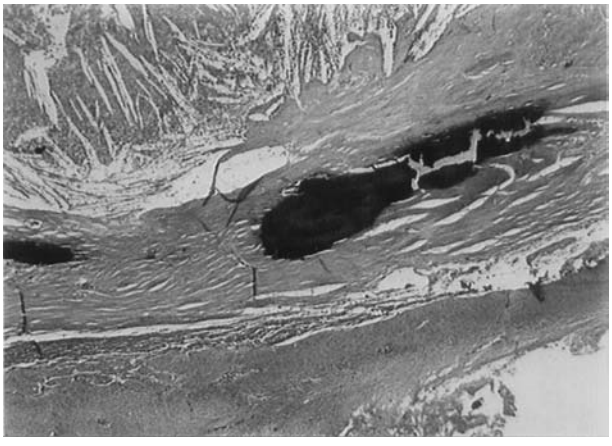

Fig. 4. Fibrous stroma and calcification lining the cyst (HES ×40)

Histopathology

The surgical specimen consisted of a cystic formation measuring $6.5 \times 5 \times 3$ cm, with a white wall which was between 0.2 and 0.5 cm in thickness, and was, in places, covered in yellowish deposits with haemorrhagic areas. Some strips of calcification were also seen.

When examined under a microscope, the wall of the cyst was lined with a cylindrical callous covering made up of ciliated cells with non-secreting cytoplasm, and rounded nuclei which were regular and non-mitotic (Inset Fig. 3). Abundant necrotic haemorrhagic material with lipophagic histiocytes associated with crystals of cholesterol was observed (Fig. 3). The underlying chorion was dense, strongly collaginised and calcified in places. A fine layer of cerebral parenchyma, without significant abnormality, was occasionally observed on the periphery (Fig. 4). The diagnosis was a colloid cyst.

Discussion

CCs are benign lesions which account for 0.55–2% of intracranial neoplasm [2, 8, 14, 16, 20, 22] and represent 55% of the third ventricle's lesions [19]. Relatively accurate diagnosis can be made using a CT scan and MRI, but a diagnostic criterion based solely on topography will be misleading as other lesions can arise in this location, and CC can occur in almost all parts of the neuraxis.

CC can arise in supra tentorial sites from the chiasma [18], sellar area [10], brain convexity [12], lateral ventricle [5, 23, 29, 28], septum pellucidum [11, 16, 28], and the posterior part of the third ventricle [25]. Its occurrence in the posterior fossa has also been reported in the fourth ventricle [25, 27, 28, 30], brainstem [15], cerebellar parenchyma [24], and the subarachnoid space [9]. It may also have a spinal location [26]. These atypical locations are rare, and are, in general, surprising clinical presentations.

CC symptomatology is protean, and most authors have stressed its non-specific nature [2, 8, 20]. However, some symptoms and signs are classic, such as: increased intra-cranial pressure, sudden onset of headaches, normal-pressure hydrocephalus, neuro-psychiatric manifestations, seizure, acute worsening and sudden death [2, 8, 11, 14, 16, 19, 20, 22]. Nonetheless, CC can present unusual signs such as haemorrhage [13, 16, 21, 22], cerebrospinal fluid rhinorrhea [17], hypopituitarism [10], diabetes insipidus [20], aseptic meningitis [2], drop attacks [20], quadriparesis [21], spasmodic torticollis [4] and many others. Fortunately, these inappropriate manifestations are rare, and some cases have even been diagnosed by chance.

Though familial forms have been described [1, 16, 22], most CCs occur sporadically as a solitary lesion of the third ventricle, though paired [23, 27], or multiple locations have been reported [31]. CC has sporadically been associated with others diseases, such as: xantogranuloma [1, 2] craniopharyngioma, astrocytoma, nevoid basal cell carcinoma, neurofibromatosis [1], leukaemia [13] and multiple sclerosis [18], or with congenital defects, such as: congenital vascular anomalies [32], agenesis of the corpus callosum, encephalo meningocele, atrial diverticula [1] and bone anomalies [21]. Such associations are fortuitous, and result in the cyst, and its diagnosis, being overlooked.

Usually, CC diagnosis is unproblematic; it is a homogeneous hyper-dense and round lesion, without calcification, inside the third ventricle. The cyst may also present as a hypo dense [1, 16, 23, 35], or calcified lesion [16, 25, 26, 33, 35], showing occasional rings [6] or heterogeneous enhancement after administration of contrast medium [5, 16, 24, 35]. In addition, differing densities and signal intensities within HCC have also been described [34].

CCs also have a specific appearance on microscopic examination, but several associated histological features have been recorded: haemorrhage [13, 21, 22], presence of elastic tissue within the cyst wall [21], xanthogranuloma [2], etc.

HCC with acute clinical deterioration [22, 29], and/or sudden death [7, 28] is a rare combination. Worsening may occur spontaneously (as in our case), or follow a stereotactic procedure [5] or lumbar puncture [29]. The risk of acute deterioration in a symptomatic patient with a CC has been estimated at 34% [14]. The mechanism behind acute aggravation and sudden death has been subject to controversy [28], and no valid explanation has yet been found.

Several surgical approaches have been proposed for the treatment of CC [3, 14, 33, 34], but some cannot be applied to HCC, in particular, endoscopic aspiration. Thus, most HCCs have required open surgery for a radical excision as none of their characteristics suggested this possibility (merely that of glioma) [5, 29, 34, 35]. In such a case, there is a debate between the trans-frontal and trans-callosal route of surgical approach, even if the latter is now usually straightforward [3, 14]. Owing to the tumour mass, the fornix was thinned ''in a fashion similar to the elongation of the facial nerve by a large ponto-cerebellar angle tumour'', so it could only be distinguished from the cyst at the foramen, of Monro and we do not believe that the trans-callosal route approach, with total excision, could have preserved it.

Not all reported cases are similar, but in most of cases, memory disorders and handling of the fornix have not been described [28, 29, 34, 35]. A few reports did however show post operative memory disorders [5, 22]. Therefore, the question is whether a total resection should be mandatory?

On the one hand, a total resection protected the patient from recurrence [2, 14, 22] but certainly increased memory disturbance. On the other hand, cyst evacuation with incomplete resection of the cyst membrane is accompanied by a high recurrence rate [8, 22] and the need to reoperate without the guarantee that the same trouble will not reoccur. Consequently, in such cases total resection seems to be the best therapeutic alternative.

Conclusion

Colloid cysts represent the main, third ventricular tumour. Most often, it has a specific radiological appearance, and is surgically curable. In its nonconforming presentations, it is disappointing, and we must remember these possibilities to prevent possibly inappropriate management in similar cases.

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Comments

Drs Hamlat *et al.* have written a report of an unusual case of a huge colloid cyst of the third ventricle. The report is well written. It captures the essence of this specific case and contains a comprehensive literature review of colloid cysts with a slight emphasis on unusual features. The review and the case report may serve as a reminder of unusual features and also of an example of a lesion where the differential diagnoses of this colloid cyst would include craniopharyngioma, central neurocytoma or glioma.

Tiit Mathieson

This is another report describing an atypical, large colloid cyst of the third ventricle and its surgical treatment. While there is nothing spectacular with the present case, the authors add a review of previously described unusual forms of this tumour, which increases the scientific value of this communication.

Helmut Bertalanffy

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