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Atypical haemorrhagic colloid cyst: 2 case reports surgical management and review of literature

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ABSTRACT

INTRODUCTION: Colloid cysts are benign cystic lesions located at the anterior part of the third ventricle mostly at the foramen of Monro and contain colloid material. Hemorrhage in a colloid cyst is exceedingly rare. Only 15 clinically diagnosed cases of haemorrhagic cysts were reported in the literature and 5 more cases on autopsy. Here we report two rare cases of a haemorrhagic colloid cyst describing the atypical radiological findings, the undertaken surgical procedures and histopathological results.

PRESENTATION OF CASES: We presented 2 cases of haemorrhagic third ventricle colloid cysts. First case is a 27-year-old male patient, presented with dizziness, nausea, vomiting and blurring of vision. He was operated by transcortical endoscopic transventricular excision of a third ventricular cyst and the insertion of external ventricular drain. The second patient is a 21-year-old male, presented with history of worsening headache for 1 month associated with blurring of vision. The patient had a transcortical microscopic, transventricular cyst excision.

DISCUSSION: Many questions regarding the best way to diagnose and manage such lesions remain unanswered. Hence, we summarize the relevant diagnostic images and best surgical techniques.

CONCLUSION: We concluded that, though exceedingly rare, colloid cyst can bleed and cause rapid deterioration in neurological status, thus, presence of atypical features should alert the physicians to consider atypical colloid cyst that would be valuable in surgical decision making whether endoscopic or microscopic.

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1. Introduction

Colloid cysts are the most common benign neoplasms of the anterior third ventricle, often at the level of the Monro foramen and can often manifest as a sudden onset of headache or loss of consciousness. They make up about 0.5–1.0 per cent of primary brain tumors. The clinical appearance is diverse, ranging from cysts detected accidentally to sudden death due to acute hydrocephalus. Such cysts also have a well-defined cyst wall, mucinous or watery intracystic fluid and the surrounding parenchyma has a demarcation. Colloid cysts are typically 0.3–4 cm in size, well-circumcised, homogeneous hyperdense on CT and low T2/high T1 signal intensity on MRI, radiologically [15].

The cyst wall consists of a cuboidal, columnar, or pseudostratified epithelial lining with interspersed goblet cells of the mucosa [4,10,13]. Intracystic haemorrhage may occasionally lead to xanthogranulomatous inflammatory changes within the cyst resulting in focal thickening of the cyst wall and adhesion to surrounding structures [3,4,16]. Here we identify two cases of atypical colloidal cysts findings and review the literature of this pathology. That work has been reported in line with the SCARE criteria [25].

2. Presentation of cases

2.1. Case 1

27-year-old male patient, medically free presented with history of several attacks of dizziness, nausea, vomiting and blurring of vision for 2 weeks. On physical examination there were no focal neurological deficits, he was dizzy but conscious and fundoscopy examination showed papilledema grade 2 on both eyes.

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Fig. 1. NECT of the brain: Midline third ventricular well-circumscribed hyperdense giant colloid cyst contributing to obstructive hydrocephalus. NECT: Non-enhanced computed tomography, Ax: Axial, Cor: coronal.



Fig. 2. MRI of the brain: Giant colloid cyst obstructing the foramen of Monro and causing obstructive hydrocephalus shows predominant non-enhancing low T2 and intermediateT1 signal intensity with dark T2 signal areas in the inferior aspect of the cyst that depicts hypointense T1 signal and blooming artifacts on SWI sequence suggestive of associated blood products.

Ax: Axial, SWI: susceptibility weighted images, Sag: Sagittal, Cor: Coronal, +C: Contrast enhanced.



Fig. 3. Axial CT brain scan without contrast after transcortical endoscopic transventricular approach showed full excision of a third ventricular cyst and the insertion of external ventricular drain (EVD) with mild pneumocephalus in the left frontal horn of the lateral ventricle.

Non-enhanced CT (NECT) of the brain showed a well-circumscribed midline third ventricular lesion mostly representing a giant colloid cyst that is hyperdense, measuring $3.3 \times 3 \times 3.7$ cm in maximum dimensions and obstructing the foramen of Monro contributing to obstructive hydrocephalus (Fig. 1). MRI analysis of the lesion revealed a heterogeneous appearance of non-enhancing low T2 and intermediate T1 signal intensity with associated central T2 dark signal intensity areas in the inferior aspect of the cyst that depicts hypointense T1 signal and blooming artifacts on SWI sequence suggestive of blood products (Fig. 2).

The patient was subjected to transcortical endoscopic transventricular excision of a third ventricular cyst and the insertion of external ventricular drain (EVD) (Fig. 3). A large third ventricular thickened contents cyst which has triggered auto-septum pellucidotomy was found intraoperatively. The cyst material was initially fenestrated and aspired and produced a brownish yellow fluid. The clot was extracted from the cyst wall upon the opening of the cyst and removed piecemeally. After almost complete cyst content evacuation, the thick wall was dissected from the surrounding parenchymal tissue and then the cyst wall was totally excised. Histopathology results showed a fibrous connective tissue wall lined with Pseudostratified columnar epithelial cells with occasional ciliated and goblet cells. Presence of red blood cells, hemosiderin laden macrophages, cholesterol crystals, and chronic inflammatory cells suggestive of hemorrhage in colloid cyst.

2.2. Case 2

21-year-old male, medically free, presented with history of worsening headache for 1 month associated with blurring of vision, fully conscious and no neurological deficit. NECT of the brain revealed a midline ill-defined heterogeneous hyperdense lesion anterior midline third ventricular lesion at the level of the foramen of monro suggestive of colloid cyst, measuring $2 \times 1.4 \times 1.6$ cm in maximum dimensions and causing obstructive hydrocephalus (Fig. 4). Further evaluation with brain MRI demonstrated heterogeneous high T1 and low T2 signal intensity appearance of the lesion that showed irregular non enhancing wall and central dark T2 and T1 signal intensity changes which showed blooming



Fig. 4. Non contrast axial CT brain image showed midline, ill defined, heterogeneous mass at the foramen of monro causing obstructive hydrocephalus and suggestive of colloid cyst.

artifacts on SWI images that is suggestive of hemosiderin staining related to prior intra-cystic haemorrhage. No abnormal contrast enhancement was detected (Fig. 5).

The patient had a transcortical microscopic, transventricular cyst excision. The wall was markedly thick intraoperatively and there was dark yellow cholesterol content coming out of the cyst until it was opened. Histopathology of the examined material showed cystic lesion lined by columnar epithelium with subepithelial extensive xanthogranulomatous reaction in the form of cholesterol crystals, foamy histiocytes, fibrosis, hemosiderin-laden macrophages and multinucleated giant cells (Fig. 6).

3. Discussion

3.1. Generalities

Colloid cysts are benign thin walled cystic lesions located midline at the anterior part of third ventricle, close to foramen of Monro and contain colloid material [6,13]. The origin of these cysts continues to be uncertain. Diencephalic ependymoma, invagination of neuroepithelium of the ventricle, or the respiratory epithelium of endodermal origin are the other etiological possibilities [4]. They constitute approximately 0.5–1.0% of primary brain tumors and 15–20% of all intraventricular masses [1,5,10]. Most found in the 3rd and 4th decades that peaked at around the age of 40 and distributed evenly among males and females with few recorded family cases. While colloidal cysts are congenital tumors, their presentation in children was found to be extremely rare (only 8 percent in patients younger than 15-year-old) as these lesions are usually become symptomatic in patients aged 20–50 years [1].

3.2. Clinical features

Colloid cyst can cause extreme morbidity and sometimes mortality due to acute obstructive hydrocephalus, and very rarely due to intracystic haemorrhage called 'cyst apoplexy' [2,4,6]. For this

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Fig. 5. MRI of the brain: Irregular walled heterogeneous T1 high and T2 low signal intensity anterior midline third ventricular colloid cyst that contributes to acute ongoing hydrocephalous. Noted central dark T2 and T1 signal intensity changes which showed blooming artifacts on SWI images (not shown) consistent with hemosiderin staining likely related to prior intra-cystic haemorrhage. No abnormal contrast enhancement was detected.

reason, surgical intervention is strongly recommended in order to avoid lethal complications, although a long-term follow-up analysis of 68 subjects with asymptomatic colloidal cysts indicates that only (8%) subjects go on to develop symptoms at 10 years [18].

With the advent of CT/MRI, asymptomatic colloid cysts have increased in numbers [1]. Memory deficiency, headache, and cognitive disorders are the most common symptoms mostly associated with increased intracranial pressure [1,6]. Dementia, gait disturbances, and urinary incontinence are other modes of presentation [1,13]. The most likely explanation for sudden deaths is the noncommunicating hydrocephalus. In addition, hemorrhagic changes in colloid cysts can lead to an acute increase in the cyst dimension resulting in an acute and complete blockage of CSF circulation [6,12].

Haemorrhagic 'cyst apoplexy' colloid cysts are rare and the cause of hemorrhage is not yet understood [1,10]. Just 15 clinically diagnosed cases of hemorrhagic cysts were reported in the literature, and 5 more cases on autopsy were reported [11]. Two cases had hypertension and one case had hemorrhagic tendencies. However, it is unclear whether these factors may have contributed to colloid cyst bleeding-arterial hypertension and coagulation abnormalities and should be carefully considered in future in patients with colloid cysts [11].

Beems et al. described the first hemorrhagic colloid cyst with a disastrous clinical course [15]. A 35-year-old female patient known to have colloid cyst, who had a sudden deterioration due to acute intracystic hemorrhage one day before surgery, was reported. Notwithstanding emergency ventriculostomy and endoscopic subtotal cyst excision, the outcome remained relatively poor. The author concluded that care decisions would weigh up the risk of hemorrhagic changes in a colloid cyst [15]. Only one paediatric case of haemorrhagic colloid cysts, reported by Farooq et al., was that of a 9-year-old girl who had initially presented a sudden deterioration in her consciousness level She had urgent ventriculostomy, craniotomy, and cyst resection. She'd had a great recovery [14].

S.A. Al-Saiari et al.

3.3. Radiological features

Studies of computed tomography (CT) found that typical colloid cysts are hyperdense in two-thirds of cases and iso- to hypodense in one-third of cases, with density depending on the content of cholesterols. It does not usually show enhancement on contrast, except for occasional rim enhancement [2,7,8,13]. On MRI, the signal varies depending on the nature of the cyst content; typically, the characteristic presence of being hyperintense on T1W images represents the amount of cholesterol content, and the amount of dense or protein fluid on the T2W images gives the iso- to hypointensity. There may be a peripheral (rim) gadolinium enhancement in some cases. These colloidal cysts typically do not inhibit on the FLAIR studies and show no restriction on diffusion-weighted imaging [2,7,8,13].

Colloid cysts are mostly 0.3–4 cm in size. Cysts larger than 3 cm are called giant colloid cysts(GCC) and are very rare [5,9]. And so, a sudden increase in cyst size due to haemorrhage and subsequent precipitous neurological decline was rarely reported [1,12]. The signal intensity of our first case on T1 and T2 was as defined in the literature, but interestingly, there was inferior heterogeneous signal intensity in T2-weighted pictures showing blooming artifacts on SWI in line with the cyst-confirmed haemorrhage. It also measured over 3 cm consistent with a rare colloid cyst variant of the giant.

Both CT and MRI studies may not be able to differentiate between colloid cysts and their xanthogranulomatous transformation; however, this differential diagnosis should be warned by atypical features [16]. The presence of blood in the cyst may trigger xanthogranulomatous inflammatory response, which is considered to be heterogeneous signal intensity on MRI; in these cases, signal characteristics are variable and depend on the mixture of lipid, fluid, and blood products [4,5,12,16]. The presence of an atypical solid-cystic lesion in the third ventricle near the Monro foramen, as in our 2nd case, can mimic a suprasellar cystic craniopharyngioma although such neoplasms are rarely present in the 3rd ventricle and have calcifications and gadolinium enhancement more often than not.

The presence of atypical features should alert the surgeon to consider alternative diagnoses, such as an atypical colloid cyst or a cystic craniopharyngioma [3]. In addition, the tendency among surgeons is to assess the quality of a colloid cyst's contents and see whether it is watery or mucinous. Also important to look for atypical features such as a heterogeneous focal zone in the capsule as this would offer clues to the possible diagnosis and provide valuable information about the plane of the cyst with the surrounding parenchyma, aiding in the appropriate surgical decision making [3].

3.4. Management

The management of symptomatic colloid cysts has different approaches. The operating procedures typically consist of open or endoscopic microsurgical techniques for excision [17-19]. Open surgical interhemispheric transcallosal technique is the technique of choice in the management of colloid cyst it has the benefit of avoiding cortex incision, but carries several complications including, but not limited to, cortical venous infarction, fornices damage, deep venous system injury, subdural hematoma, disconnection syndrome, ventriculitis, and meningitis [20,21]. Endoscopic approach has been reported by B. Diyora et al. to have advantages over microscopic one including excellent visualization and minimally invasive acccess through the dilated ventricular system. MRI signal intensities of the cyst have a major role in decision making regarding to mode of surgical intervention of the colloid cyst and can direct the neurosurgeon to the way he can deal with. In thin content cyst stereotactic-guided surgeries can be applica-



Fig. 6. Histopathology of case 2 showed cystic lesion lined by columnar epithelium with subepithelial extensive xanthogranulomatous reaction in the form of cholesterol crystals, foamy histiocytes, fibrosis, hemosiderin-laden macrophages

ble [22–24]. On the opposite side, a firm, solid or thickened cyst wall, as well as a cyst containing clot or haemorrhagic part, needle aspiration cannot be accomplished as the cyst mostly adheres to neighbouring vital structures, particularly the fornix. This clot can also not be pulled except by piecemeal removal, which is why open microsurgical technique is the best choice for certain colloid cyst types [1].

4. Conclusions

and multinucleated giant cells.

A haemorrhagic colloid cyst can pose a management challenge, as it cannot be treated as a simple colloid cyst equivalent. A thickened cyst wall and cyst material prevents the possibility of needle aspiration. Manipulation of well-defined solid mass without decompression is often at the risk of damage to the critical structures around it. Also, the strong adherent clot cannot be drained out and decomposition is unavoidable. We believe that while endoscope may have many advantages over microscope, the best treatment choice for a haemorrhagic colloid cyst should be considered an open microsurgical technique.

Declaration of Competing Interest

The authors report no declarations of interest.

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Ethical approval

In our institute, ethical approval is not required for a case report. However, informed consent was received from the patient to participate in the report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

S.A. Al-Saiari et al.

Author contribution

First case operated and managed by the second author, second case operated and managed by the first author. All authors shared in writing, editing, reading and approving the final manuscript.

Registration of research studies

N/A.

Guarantor

The corresponding author Mohamed A Khoudir, is the Guarantor for this work.

Provenance and peer review

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