Rare disease

Spontaneous haemorrhage and rupture of third ventricular colloid cyst

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Summary

Acute bleeding within a colloid cyst of the third ventricle represents a rare event causing sudden increase in the cyst volume that may lead to acute hydrocephalus and rapid neurological deterioration. We report a case of spontaneous rupture of haemorrhagic third ventricular colloid cyst and its management. A 77-year-old ex-smoker presented with unsteady gait, incontinence and gradually worsening confusion over a 3-week period. Brain CT scan findings were highly suggestive of a third ventricular colloid cyst with intraventricular rupture. He underwent cyst excision and histopathology, which confirmed the radiological diagnosis with evidence of haemorrhage within the cyst. A ventriculo peritoneal shunt was performed for delayed hydrocephalus. Surgical management of these patients must include emergency ventriculostomy followed by prompt surgical removal of the haemorrhagic cyst.

BACKGROUND

Colloid cysts are benign neoplasms that develop in the anterior part of the roof of the third ventricle.¹ They are rare, accounting for approximately 0.5–1.0% of primary brain tumours, and comprising approximately 15–20% of intraventricular neoplasms² with an estimated incidence of three people per million per year. Acute bleeding within a colloid cyst of the third ventricle represents a rare event causing a sudden increase in the cyst volume that may lead to acute hydrocephalus and rapid neurological deterioration. We report the first documented case of spontaneous rupture of haemorrhagic third ventricular colloid cyst and its management.

CASE PRESENTATION

A 77-year-old ex-smoker presented with unsteady gait, incontinence and gradually worsening confusion following a fall 3 weeks prior to the presentation. He had a history of mild cognitive impairment, atrial fibrillation, type II diabetes mellitus and had a cardiac pacemaker in situ. He was on warfarin and his international normalised ratio (INR) on admission was 1.2. On examination, his Glasgow Coma Scale was 14/15, with mild right-sided weakness; power 4/5. Cranial nerve examination was normal.

CT of brain demonstrated a well-defined, 2×2 cm, hyperdense lesion in the roof of the third ventricle consistent with a colloid cyst with haemorrhage within the cyst (figure 1). It also revealed features in keeping with small vessel ischaemic disease.

He underwent right frontal craniotomy via transcortical approach to the right lateral ventricle with excision of the cyst from the roof of the third ventricle. Histopathology sections revealed a strip of ciliated columnar epithelial cells with adjacent haemorrhage (figure 2A,B). Postoperative CT of brain revealed satisfactory cyst excision (figure 3).

Figure 1 Non-contrast brain demonstrating a hyperdense colloid cyst with evidence of haemorrhage in the lateral ventricles (hyperdensity in the dependent portion of the occipital horns of lateral ventricles).

OUTCOME AND FOLLOW-UP

Ten days postoperation, he became increasingly drowsy, and a repeat brain CT scan revealed hydrocephalus. He underwent a ventriculo peritoneal shunt placement.

Figure 2 (A) Low-power view of the surgical specimen comprising haemorrhage, blood clot and a small strip of epithelial cells. (B) Columnar epithelial cells, cilia focally apparent, consistent with colloid cyst.

(figure 4). He made a satisfactory postoperative recovery and was discharged to a community hospital for continued rehabilitation.

Figure 3 Postoperative CT of brain demonstrating excision of the colloid cyst.

Figure 4 Ventriculo peritoneal shunt placement to treat hydrocephalus.

DISCUSSION

Third ventricular colloid cysts are benign thin-walled cystic lesions located in the roof of the third ventricle close to the foramina of Monro. They originate from ectopic endodermal elements having migrated into the velum interpositum during central nervous system development.¹ These cysts are composed of a mixed array of epithelial and goblet cells. The goblet cells secrete proteinaceous material that accumulates progressively within the cyst.³

Most patients with a colloid cyst diagnosed have symptoms not evidently caused by or related to the colloid cyst.^{4 5} Memory deficits, headaches and cognitive disturbance are the most related symptoms. The appearance of clinical symptoms in patients afflicted by colloid cysts is considered by most authors to be the result of a cerebrospinal fluid dynamics failure to adjust to changes in the volume and/or position of the colloid cyst.⁶ Rapid expansion of a colloid cyst may produce clinical symptoms associated with increased intracranial pressure, while a gradual growth of the lesion does not manifest clinically due to the fact that intracranial pressures remain within normal limits for longer periods in spite of the progressive enlargement of the cerebral ventricles.

The accurate incidence and mechanisms involved in sudden death from colloid cyst remain poorly understood.^{1 7 8} The frequent finding of ventricular enlargement and brain herniation in autopsy specimens displaying a colloid cyst of the third ventricle has led many authors to postulate that hydrocephalus may play a major role in unexpected deterioration. The rapidity at which hydrocephalus develops, more than the degree of ventricular

dilatation, is the critical factor related to sudden death in these patients.^{4 8}

Colloid cysts appear hyperdense on plain CT scan owing to its colloid content. The evidence of intracystic bleed in our case was subsequently confirmed by histopathological analysis.

Acute intracyst bleeding should be considered an alternative mechanism for sudden deterioration in patients with a third ventricle cyst. It is not clear as to which factors can facilitate colloid cyst bleeding; nevertheless, arterial hypertension⁹ and coagulation disturbances¹⁰ may play a role. It is still possible that the fall preceding the presentation may have caused bleed into the cyst. However, this is very rare and the INR was within normal limits in our case at presentation. Cyst haemorrhage and subsequent rupture may be deleterious as evident from our case. To our knowledge, this is the first documented case of haemorrhagic rupture of third ventricular colloid cyst, which was satisfactorily treated by surgical excision.

Learning points

- ▶ Spontaneous, acute bleeding and subsequent rupture of third ventricular colloid cyst is a rare, potentially fatal phenomenon due to the rapid development of hydrocephalus.
- ▶ Surgical management of these patients must include emergency ventriculostomy followed by prompt surgical removal of the haemorrhagic colloid cyst.
- ▶ The possibility of delayed hydrocephalus must be anticipated and if encountered, a ventriculo peritoneal shunt may be necessary.

Competing interest None.

Patient consent Obtained.

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