Title:

Chronic subdural hematoma associated with arachnoid cyst in autosomal dominant polycystic kidney disease (ADPKD). Case report.

Authors:

Dr Gilberto Ka Kit Leung *. M.B.B.S. (equivalent to M.D.)

Dr Yiu Wah Fan *. M.B.B.S. (equivalent to M.D.)

Institution:

Division of Neurosurgery
Department of Surgery
The University of Hong Kong Medical Center
Queen Mary Hospital
Hong Kong SAR
China

Corresponding Address : (Before 30 June 2003)

Dr Gilberto K.K. Leung
Department of Neurosurgery
University of Virginia Health System
PO Box 800212
Charlottesville VA 22908
US.

Telephone No: 434-982-3500 (page 3164)
Email: gilbertleung2002@yahoo.com
Fascimile No: 434 –924-5894

Corresponding Address : (After 30 June 2003)

Dr Gilberto K.K. Leung
Division of Neurosurgery
Department of Surgery
The University of Hong Kong Medical Center
Queen Mary Hospital
Hong Kong SAR
China

Telephone No: (852)-2855-3368
Email: gilbertleung2002@yahoo.com
Fascimile No: (852)-2838-4150

Abstract

Objective and Importance

Chronic subdural hematoma (CSDH) is a known complication of intracranial arachnoid cyst. Autosomal dominant polycystic kidney disease (ADPKD) has an increased incidence of intracrainial arachnoid cyst. The association between CSDH and ADPKD has not been recognized. Increased awareness may lead to early diagnosis and treatment of the complications of ADPKD.

Clinical Presentation

A 27-year-old man had a left middle cranial fossa arachnoid cyst managed conservatively. He later presented with non-traumatic CSDH and previously undiagnosed systemic hypertension.

Intervention

The CSDH was evacuated through burr holes but the procedure was complicated by post-operative extradural hemorrhage due to over-drainage. ADPKD was diagnosed on abdominal ultrasonography as the cause of the systemic hypertension.

Conclusion

ADPKD should be considered as a differential diagnosis in a young patient with hemorrhagic complications of intracranial arachnoid cyst, especially when there is unexplained systemic hypertension. Early diagnosis of ADPKD can facilitate genetic counseling and treatment of its systemic complications, including renal insufficiency, systemic hypertension and intracranial saccular aneurysm. Drainage of CSDH associated with arachnoid cyst may carry risks of over-drainage and should be performed with caution.

Running Title: Subdural hematoma in ADPKD

Key words : arachnoid cyst, autosomal dominant polycystic kidney disease , subdural hematoma

Chronic subdural hematoma associated with arachnoid cyst in autosomal dominant polycystic kidney disease. Case report.

Introduction

Autosomal dominant polycystic kidney disease (ADPKD) is a systemic disorder associated with intracranial manifestations such as intraparenchymal and aneurysmal subarachnoid hemorrhage. (16) Imaging studies have demonstrated an increased incidence of intracranial arachnoid cysts in patients with ADPKD. (15) Chronic subdural hematoma (CSDH) as a complication of arachnoid cyst is well documented (3, 11, 13, 17) but its association with arachnoid cyst in ADPKD has not been recognized. (18) We report a case of chronic subdural hematoma complicating an intracranial arachnoid cyst in a patient with ADPKD.

Case Report

A 27-year-old man presented to his family physician with six months' history of intermittent generalized headache. There was no other symptom or significant past medical history. Magnetic resonance imaging (MRI) demonstrated an arachnoid cyst occupying the anterior and middle parts of the left middle cranial fossa, and foreshortening of the left temporal lobe (Galassi Type II). (7) There was no evidence of hemorrhage (Fig 1). The patient was managed conservatively with oral analgesics.

The patient later presented to our unit with four days' history of vomiting and worsening headache. There was no recent trauma. He was conscious and orientated. Neurologic examination was normal. No skull deformity was found in the temporal

regions. His systolic and diastolic blood pressures were persistently above 170 mmHg and 110 mmHg respectively. This was initially attributed to pain and anxiety.

Blood investigations were normal including blood coagulation and renal function. Computerized tomography (CT) showed a left frontotemporal CSDH with extension into the temporal arachnoid cyst (Fig. 2a & b). No subarachnoid hemorrhage was found.

Surgical drainage was performed through two burr holes under general anesthesia. Intra-operatively, liquefied subdural hematoma was found and there was no acute hemorrhage. The subdural space was irrigated with warm 0.9% sodium chloride solution and a subdural catheter was inserted for post-operative drainage. The arachnoid cyst was not explored.

The patient recovered well with relief of the headache. The subdural drain was opened on the ward and immediately yielded more than 30 ml of blood-stained fluid within 5 minutes when inadvertently placed low at 15 cm below ear level. The patient soon developed severe headache followed by deterioration in conscious level. A repeated CT scan revealed two acute extradural hematomas immediately deep to the burr hole sites. There was no acute subdural hematoma (Fig. 3). The extradural hematomas were evacuated through a craniotomy. The subdural space was explored and was found to be in free communication with the temporal arachnoid cyst, which had ruptured in its outer membrane. No obvious bleeding source was identified in the extradural or intradural compartments. The arachnoid cyst was widely fenestrated with excision of abnormal arachnoid membrane.

The patient recovered well without neurologic deficit. Post-operative CT scan showed satisfactory brain decompression and a residual cystic space in the left middle cranial fossa. The patient continued to have unexplained systemic hypertension, for which further biochemical tests did not reveal any underlying cause. Abdominal ultrasonography later demonstrated bilateral multiple renal cysts. Despite the absence of positive family history, autosomal dominant polycystic kidney disease (ADPKD) was diagnosed. No other intra-abdominal cyst was found. The patient was put on anti-hypertensive medications and remained asymptomatic with normal blood pressure and renal function 18 months after surgery. In view of the patient's history of headache, digital subtraction cerebral angiography was performed, which did not reveal any intracranial aneurysm. We have not performed genetic study on the patient or screening for ADPKD for his other family members.

Discussion

Intracranial arachnoid cyst is an uncommon developmental aberration characterized by splitting and duplication of the arachnoid membrane. (14) The majority of cases are incidental imaging findings while symptomatic lesions may present with headache, seizure or neurologic deficit. (8) Complications of arachnoid cysts resulting in subdural hygroma, intracystic hematoma and subdural hematoma have been reported. (4,10) The exact incidence of hemorrhage is unknown and has been estimated to be approximately 0.04%. (7,12) Typically, the subdural hemorrhage occurs in young male patients with middle fossa arachnoid cysts, as in the present case. (12)

Altered cell growth and abnormal extracellular matrix is believed to play an important role in the pathogenesis of ADPKD and contribute to abnormal cyst formation. (5) The incidence of arachnoid cyst has been shown to be ten times higher in patients with ADPKD (8.1%) than in the general population (0.8%) (15), and familial arachnoid cysts associated with ADPKD have been described. (1) The majority of these cysts were asymptomatic and hemorrhagic complications are extremely rare. (2) To our knowledge, association between CSDH and ADPKD has been discussed previously in only one report. (18) In this report, five young men with ADPKD presented with CSDH. Four harbored intracranial cysts but only one cyst was in close proximity to the subdural hematoma. In the present case, direct communication between the ruptured arachnoid cyst and the subdural hematoma was confirmed during operation. Although no distinct bleeding source could be identified, it is likely that hemorrhage occurred during cyst rupture, causing damage to a nearby blood vessel. (17)

Whether uncontrolled systemic hypertension contributed to the development of CSDH in our patient is uncertain. Abnormal vascular reactivity and systemic hypertension is common in ADPKD, and hypertension is thought to predispose to hemorrhage into renal cysts in ADPKD. (6) Our present patient and all five patients in the report mentioned above suffered from hypertension although any suggestion of a casual link can only remain speculative. (18) Underlying ADPKD does not appear to increase the risk of bleeding in arachnoid cyst. In a series of 247 patients with ADPKD, intracranial arachnoid cysts were found in 20 patients and none developed CSDH. (15)

Although our patient had no positive family history, the appearance of bilateral multiple renal cysts on ultrasonography was characteristic of ADPKD. Only about 60% of patients with ADPKD will report a positive family history and the diagnosis can often be established on imaging findings. (5) Systemic hypertension and CSDH are both uncommon in young patients and further investigations may reveal certain underlying conditions, such as arachnoid cyst and ADPKD in the present case. Because the association is distinctly rare, a high index of suspicion is necessary. Early diagnosis is important for ADPKD. Renal failure is a serious complication of ADPKD and its prognosis is strongly affected by early and satisfactory control of systemic hypertension. (5) ADPKD is a hereditary disease and genetic counseling can be facilitated by early diagnosis in young patients of reproductive age. Improved awareness may also facilitate the treatment of any unruptured saccular aneurysms once the patients develop related symptoms. (16)

Surgical treatment of arachnoid cysts complicated by hemorrhage has been a matter of controversy. (7, 11, 12, 13, 17) In patients with no or only minimal symptoms prior to hemorrhage, only decompression of the space-occupying collection without surgery on the arachnoid cyst has been recommended. (12) In the present case, burr hole drainage achieved satisfactory clinical improvement but was unfortunately complicated by extradural hematoma. This was most likely due to technical fault during operation. However, we had also observed inadvertent over-drainage through the subdural drain which may have contributed to dural detachment and extradural bleeding. Free communication between the basal subarachnoid cistern and arachnoid cyst has been demonstrated in Type I and II middle cranial fossa cysts typically associated with CSDH. (8) Post-operative subdural drainage in these situations

therefore should be performed with caution. Interestingly, extradural hematoma in association with arachnoid cyst also has been described. (9)

Conclusion

Neurosurgeons are acquainted with ADPKD due to the increased incidence of cerebral aneurysm but the possible association between ADPKD, arachnoid cysts and CSDH has not been recognized. Patients with ADPKD have an increased incidence of intracranial arachnoid cysts, which may predispose to the development of CSDH. ADPKD should be therefore considered as a differential diagnosis in a young patient who presents with hemorrhagic complications of an intracranial arachnoid cyst, especially when there is co-existing systemic hypertension. Drainage of CSDH associated with arachnoid cyst may cause over-drainage of cerebrospinal fluid due to communication between the cyst and the cisternal subarachnoid space and should be performed with caution.

Financial Disclosure

There is no financial support received in conjunction with the generation of this submission.

References

1. Alehan FK, Gurakan B, Agildere M: Familial arachnoid cysts in association with autosomal dominant polycystic kidney disease. **Pediatrics** 110: e13, 2002.
2. Allen A, Wiegmann TB, MacDougall ML: Arachnoid cyst in a patient with autosomal-dominant polycystic kidney disease. **Am J Kidney Dis** 8: 128-130, 1986.
3. Aoki N, Sakai T: Intraoperative subdural hematoma in a patient with arachnoid cyst in the middle cranial fossa. **Childs Nerv Syst** 6: 44-46, 1990.
4. Donaldson JW, Edwards-Brown M, Luerssen TG: Arachnoid cyst rupture with concurrent subdural hygroma. **Pediatr Neurosurg** 32: 137-139, 2000.
5. Gabow PA: Autosomal dominant polycystic kidney disease. **N End J Med** 39: 332-342, 1993.
6. Gabow PA, Duley J, Johnson AM: Clinical profiles of gross hematuria in autosomal dominant polycystic kidney disease. **Am J Kidney Dis** 20: 140-143, 1992.
7. Galassi E, Piazza G, Gaist G, Frank F: Arachnoid cysts of the middle cranial fossa: A clinical and radiological study of 25 cases treated surgically. **Surg Neurol** 14: 211-219, 1980.
8. Galassi E, Tognetti F, Gaist G, Fagioli L, Frank F, Frank G: CT scan and metrizamide CT cisternography in arachnoid cysts of the middle cranial fossa: Classification and pathophysiological aspects. **Surg Neurol** 17: 363-369, 1982.
9. Molloy CJ, Jones NR, North JB: Arachnoid cyst presenting as an extradural hematoma. **Br J Neurosurg** 56: 635-637, 1991.

10. Page AC, Mohan D, Paxton RM: Arachnoid cysts of the middle fossa predispose to subdural hematoma: Fact or fiction. **Acta Neurochir Suppl (Wien)** 42: 210-215, 1988.
11. Page A, Paxton RM, Mohan D: A reappraisal of the relationship between arachnoid cysts of the middle fossa and chronic subdural haematoma. **J Neurol Neurosurg Psychiatry** 50: 1001-1007, 1987.
12. Parsch CS, Krauss J, Hofmann E, Meixensberger J, Roosen K: Arachnoid cysts associated with subdural hematomas and hygromas: Analysis of 16 cases, long-term follow-up, and review of the literature. **Neurosurgery** 40: 483-490, 1997.
13. Prabhu VC, Bailes JE: Chronic subdural hematoma complicating arachnoid cyst secondary to soccer-related head injury: case report. **Neurosurgery** 50: 195-197, 2002.
14. Rengachary SS, Watanabe I, Brackett CE: Pathogenesis of intracranial arachnoid cyst. **Surg Neurol** 9: 139-144, 1978.
15. Schievink WI, Huston J 3rd, Torres VE, Marsh WR: Intracranial cysts in autosomal dominant polycystic kidney disease. **J Neurosurg** 83: 1004-1007, 1995.
16. Schievink WI, Torres VE, Piepgras DG, Wiebers DO: Saccular intracranial aneurysms in autosomal-dominant polycystic kidney disease. **J Am Soc Nephrol** 3: 88-95, 1992.
17. Sener RN: Arachnoid cysts associated with post-traumatic and spontaneous rupture into the subdural space. **Comput Med Imaging Graph** 21: 341-344, 1997.

18. Wijdicks EF, Torres VE, Schievink WI: Chronic subdural hematoma in autosomal dominant polycystic kidney disease. **Am J Kidney Dis** 35: 40-43, 2000.

Authorship

First Author: Dr G K K Leung is the writer of the article and the main operating neurosurgeon in this case

Second Author: Dr Y W Fan is the attending neurosurgeon in this case.

Acknowledgement

There is no specific acknowledgement of grant information for this article. There is no personal or institutional financial interest in any drugs, materials or devices described in the article.

Legends for figures

Figure 1. T2-weighted MRI studies showing a left middle cranial fossa arachnoid cyst.

Figure 2a. CT scan showing a left chronic subdural hematoma

Figure 2b. Extension of the subdural hematoma into the arachnid cyst

Figure 3. CT scan showing two extradural hematomas after drainage of the subdural hematoma.

Article Summary

This article illustrates the unusual association between chronic subdural hematoma (CSDH), intracranial arachnoid cyst and autosomal dominant polycystic kidney disease (ADPKD) in a 27-year-old man. Chronic subdural hemorrhage is a documented complication of intracranial arachnoid cyst, the incidence of which is increased in patients with ADPKD. Improved awareness of this association may lead to earlier diagnosis of ADPKD and treatment of its systemic complications, including renal insufficiency, systemic hypertension, and previously unsuspected intracranial saccular aneurysm. Surgery for CSDH in association with intracranial arachnoid cyst may carry risks of over-drainage, as illustrated in the present case by the development of extradural hemorrhage.