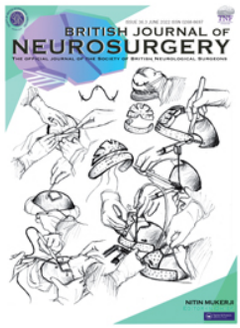

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BRIEF REPORT

First reported case of hydrocephalus in jointly diagnosed bacterial meningitis and a colloid cyst: how Ockham's razor became Hickam's dictum

Gareth May, Simon Lammy*,** , Aditaya Kumar, Ajay Hegde# and Edward Jerome St. George

Department of Neurosurgery, Institute of Neurological Sciences, Glasgow, UK

ABSTRACT

We report the first case in the literature of acute hydrocephalus due to a simultaneous diagnosis of bacterial (not asceptic) meningitis and a colloid cyst. Diagnosing disease is the cornerstone skill of a medical practitioner. Both education and experience allow for sharpening of this skill throughout years of medical practice. Disease is fraught with nuances and inconsistencies which can render an accurate diagnosis a difficult task. Medical practitioners can be guilty of cognitive biases such as Ockham's razor. We present the case of a patient with an initial diagnosis of obstructive hydrocephalus secondary to a colloid cyst. However, pneumococcal meningitis blunted Ockham's razor in favour of Hickam's dictum.

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Neuro oncology; colloid cyst; hydrocephalus; confusion; simplicity

Introduction

A colloid cyst is a benign intracranial lesion which accounts for 0.5 – 3.0% of primary brain neoplasms.[1] Most are asymptomatic and arise in the anterior aspect of the third ventricle. Its proximity to the interventricular foramen of Munro can cause obstructive hydrocephalus which may result in coma and death if left untreated.[2] Although symptomatic colloid cysts often present due to symptoms of raised intracranial pressure (ICP),[3] rare cases of aseptic meningitis have been reported secondary to extravasation of cyst contents.[4] Our case highlights how the quest for diagnostic parsimony resulted in our patient being diagnosed with a symptomatic colloid cyst (Ockham's razor).[5] However, subsequent blood and cerebrospinal fluid (CSF) cultures grew *Streptococcus pneumoniae*. The ensuing diagnostic uncertainty to the actual cause of his hydrocephalus reflects the importance of Hickam's dictum,[6] "*a man can have as many diseases as he damn well pleases*." And in our post-modern society any human being can have as many diseases as they damn well please.

Case report

A 36-year-old Nigerian male presented to his local Accident and Emergency (A&E) department due to a 1-day history of worsening headache, confusion and agitation. He had no past medical history (or recent travel history). He is a non-smoker and does not consume alcohol. His Glasgow Coma Scale (GCS) was 14 due to confusion.

Initial blood tests demonstrated a slightly raised C-reactive protein (CRP) of 11 mg/L and white blood cell (WBC) count of 12.8×10^9/L (being predominantly neutrophilic). The rest of his routine haemoglobin and biochemistry revealed normal indices. A blood film showed hypochromic microcytic erythrocytes and iron studies confirmed an iron deficiency anaemia, i.e. (a) transferrin 1.7 g/L (b) iron 9 µmol/L and (c) transferrin saturation 21%. Despite being initially apyrexial at 36.5 °C he became pyrexial once at 38.5 °C before becoming apyrexial (NB: he never became pyrexial again).

Due to a clinical suspicion of meningitis his peripheral blood was cultured and he was commenced on broad spectrum antibiotics and acyclovir. Routine plain chest radiographs (CXRs) and electrocardiograms (ECGs) were normal.

Given the acute presentation and worsening agitation a computed tomography (CT) was requested (see Figure 1(A)). This unexpectedly revealed a 6 × 5 mm rounded hyper-dense lesion at the intraventricular foramen of Monro resembling a colloid cyst. Acute hydrocephalus was demonstrated by lateral ventriculomegaly, a slightly dilated third ventricle and some obliteration of the basal cisterns. Furthermore, partial opacification of the left sphenoid sinus indicating inflammatory change was noticed (see below).

Due to neurologically deteriorating he was intubated and ventilated for emergency transfer to our neurosurgical theatres for insertion of bilateral external ventricular drains (EVDs). His GCS pre-intubation was nine: localizing, eyes open to pain and incomprehensible sounds. CSF was under high pressure. Both EVDs were set at 10 cm H_20 and the patient was transferred to our neurosurgical intensive care unit (ICU).

CSF analysis revealed a slightly elevated protein 0.54 g/L and glucose 5.1 mmol/L and an elevated white cell count (WCC) 9/mm^3 and red cell count (RCC) 53/mm^3. It unexpectedly demonstrated a gram-positive cocci. He was immediately commenced on intravenous (IV) ceftriaxone and vancomycin. The next day he was extubated and stepped down to our high dependency unit (HDU).

CONTACT Simon Lammy lammy@doctors.org.uk Institute of Neurological Sciences, Department of Neurosurgery, 1345 Govan Road, Glasgow, G51 4TF UK
*University of Glasgow, Glasgow, G12 8QQ, United Kingdom.
**Wolfson Wohl Cancer Research Centre, Institute of Cancer Sciences, Garscube Estate, Glasgow, G61 1QH, United Kingdom.
#Kasturba Medical College Manipal, Manipal, Karnataka, India.
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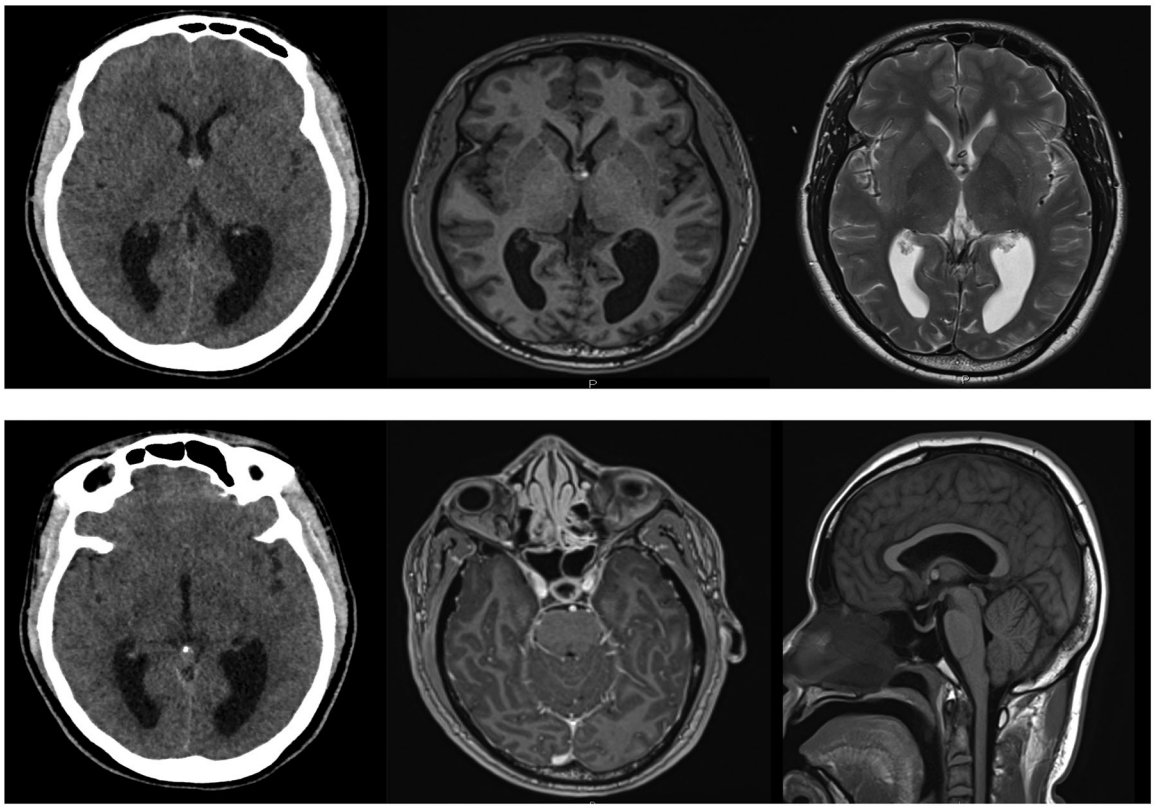

Figure 1. (a) plain axial CT demonstrating a hyper dense lesion consistent to a colloid cyst and some acute hydrocephalus evidenced by bilateral frontal horn dilation and temporal ventricular enlargement and slight dilation of the third ventricle; (b) axial T1 MRI demonstrating a hyper intense lesion and a post-T1 image showing no significant contrast enhancement of the basal leptomeningies; (c) axial T2 MRI revealing an isointense lesion and ongoing hydrocephalus and a mid-sagittal T1 pre-contrast showing no depressed floor the third ventricle.

His CRP increased sequentially on post-operative day two to a peak of 312 mg/L and normalized on day eight. His peripheral WCC increased to a peak of 25.1×10^9/L on post-operative day two and took 10 days to normalize. Several days later the peripheral blood cultures taken in A&E grew *Streptococcus pneumoniae* in both aerobic and anaerobic bottles. The final CSF culture from his emergency EVD insertion cemented the diagnosis of meningitis by demonstrating *S. pneumoniae* serotype 15B (this serotype is the in 23-valent vaccine). CSF polymerase chain reaction (PCR) was negative for viruses. Further CSF PCR for *Haemophilus influenzae* (hel gene), meningococcus (ctrA gene) and pneumoccous (lytA gene) was negative. All tests for blood borne viruses, i.e. hepatitis B surface antigen (HBV), hepatitis C antibody (HCV) and human immune-deficiency virus (HIV) were all negative. Tests for sickle cell and malaria were both negative. Interestingly, a second CSF specimen on the second post-operative day demonstrated WCC 39/mm^3 (PMNs 83%) and RCC 23/mm^3. The organism bacillus simplex was isolated. No more CSF was analysed.

A magnetic resonance image (MRI) conducted on the second post-operative day revealed the 6 mm colloid cyst to be of heterogeneous signal intensity on both T1 and T2 demonstrating no contrast enhancement (see Figure 1(B,C)). A plain CT post-EVD removal demonstrated minor reduction in ventriculomegaly.

The patient improved neurologically. Both EVDs were removed later that week and he was managed on a non-critical care ward. A repeat MRI prior to discharge on the ninth post-operative day showed unchanged appearances of the colloid cyst and left sphenoid sinus opacification retrospectively scrutinized.

Our patient was discharged on the seventeenth post-operative day recovering full neurological function. He was given the meningococcal vaccine prior to discharge. He required admission to his local hospital in the post-operative period due to developing a chest infection and a deep venous thrombosis (DVT). Subsequent MR imaging revealed a defect within the wall of his left sphenoid sinus.

Discussion

Our case demonstrates diagnostic uncertainty as to the culprit pathological process that caused acute hydrocephalus and neurological deterioration. Was it meningitis? Was it the colloid cyst? Was it both? So let us present evidence for culprit uncertainty. This report will focus on the development of hydrocephalus in pneumococcal meningitis and its role in confounding the finding of a colloid cyst. We found no evidence on literature review of colloid cysts causing both bacterial meningitis and hydrocephalus. Nor have we found evidence of hydrocephalus in the presence of both septic meningitis and a colloid cyst.

Colloid cysts are benign central nervous system (CNS) lesions which account for $0.5 - 3.0\%$ of primary brain tumours.[1] Most are asymptomatic.[3] They are histologically composed of a thin outer capsule of fibrous connective tissue on which a lining of simple cuboidal and pseudostratified columnar epithelial cells is interspersed with mucus-secreting goblet and ciliated cells.[7] Cyst contents consist of an eosinophilic amorphous periodic acid-Schiff (PAS)-positive material.[8] Cyst rupture has been reported to

cause presentations of both acute and chronic aseptic meningitis.[4]

Cyst enlargement, haemorrhage and positional obstruction of the interventricular foramen of Munro can cause ventriculomegaly and raised intracranial pressure (ICP).[2,3,9] Most symptomatic patients will present with signs and symptoms of raised ICP, e.g. non-specific headache, vomiting, and papilloedema.[2] Due to a peduncular attachment to the roof of the third ventricle a colloid cyst can intermittently occlude the interventricular foramen of Munro (in a ball-valve mechanism). This can result in sudden onset headaches and loss of consciousness (or drop attacks).[3] Irreversible obstruction to CSF flow results in obstructive hydrocephalus. Without prompt neurosurgical intervention death can occur.[2] Risk factors for being symptomatic include: age <50 years, presence of headaches, cyst diameter >8 mm, large cyst volume and presence of ventriculomegaly.[2]

Cysts are usually hyperdense on CT.10 Both calcification and contrast enhancement are uncommon. These lesions are usually hyperintense on T1 and hypointense on T2-weighted MR imaging. This reflects the cholesterol and protein content within the cyst.[10] The case presented demonstrated heterogeneous signal intensity.

If it is clinically indicated once hydrocephalus has been treated cyst resection can then be achieved by either microsurgical or endoscopic techniques. A systematic review by Sheikh et al in 1278 patients demonstrated higher rates of gross total resection and lower rates of reoperation and recurrence in patients who underwent microsurgical resection compared to endoscopic resection.[11] But microsurgical techniques are also associated with a higher seizure and readmission rate when compared to endoscopic techniques.[12] Our patient did not warrant cyst resection due to (a) his hydrocephalus not being singularly attributable to his cyst (b) the cyst dimensions not warranting resection and (c) his concomitant meningitis on re-assessment was thought to be the cause of such a rapid ventriculomegaly.

However, the impact of meningitis to confound diagnostic certainty cannot be underestimated. It remains a medical emergency and has an annual incidence of $2-5$ per 100,000 in a developed country.[13] The most common infective organisms in the United Kingdom (UK) are *Neisseria meningitidis*, *Streptococcus pneumoniae* and *Haemophilus influenzae*.[14] Although *S. pneumoniae* is responsible for less than 15% of cases of septic meningitis it has a higher mortality (16–37%) and morbidity (30–52%) rate.[15] Since the pneumococcal vaccine was introduced in 2006 UK hospital admission rates have dropped from 4.45 per 100,000 to 2.03 per 100,000 in 2011.[16]

Once inside the CNS *S. pneumoniae* activates an immune cascade, causing collateral neuronal and meningeal inflammation through cytotoxic storms and oxidative stress.[17] Our case had a defect in an infected sphenoid sinus on retrospective examination of the admission CT in A&E. Certain well-known complications and their prevalence rate within the adult population include (amongst others) cerebral infarction (36%) and hydrocephalus (7%).[18] Our patient though demonstrating hydrocephalus did not develop radiological evidence of infarction on both post-operative MRIs.

Hydrocephalus is an uncommon complication of bacterial meningitis. A 2010 meta-analysis by Edmond et al found a global prevalence rate of 7.1% ($n=18132$ [1998–2008]) for post-bacterial meningitis hydrocephalus.[15] This is similar to the pooled prevalence rate of 6.8% in a 2010 meta-analysis by Jit ($n=3408$; [1991–2009]) for hydrocephalus associated with pneumococcal meningitis in developed countries.[19]

Most commonly, communicating hydrocephalus is seen in the context of pneumococcal meningitis.[20] This is caused by reduced CSF resorption at the arachnoid granulations and is due to (a) increased CSF turbidity from inflammatory cells and proteins; and (b) fibrotic damage of the arachnoid villi due to collateral damage from the inflammatory cascade.[20] Non-communicating (obstructive) hydrocephalus is much less common and is usually caused by an obstructive purulent inflammatory exudate.[21] Most cases of obstructive hydrocephalus in bacterial meningitis occur at the level of the aqueduct or lateral apertures of Luschka.[22]

The presence of hydrocephalus in meningitis confers an increase in mortality rate (60% vs 17%) compared to non-hydrocephalic meningitis.[23] The cause of this increase in mortality is multifactorial i.e. delay in presentation, co-morbidities, severity of the underlying infection. This makes our case unusual as both CSF WCC counts were lower than expected for pneumococcal meningitis and reports of normal CSF in adult meningitis patients is rare but does exist.[24] Furthermore, bacterial meningitis in the context of acute obstructive hydrocephalus can evade microbiological detection on ventricular CSF analysis resulting in devastating consequences.[25]

Risk factors for the development of hydrocephalus include; (a) disturbed consciousness at the time of admission and (b) higher mean age.[21] Despite successful vaccination programmes, pneumococcal meningitis continues to evolve and contribute a significant health burden throughout the world.[15] Clinical astuteness must be employed to recognise this disease early and the sequalae which may co-present with it.

It was reasonable to assume given the radiological findings of hydrocephalus and the presence of a colloid cyst (despite its size) that both were related (unifying diagnosis). However, once microbiological analysis of his peripheral blood and CSF confirmed a pneumococcal meningitis then the exact cause for his deterioration was difficult to decipher. Did our patient suffer from hydrocephalus as a result of his colloid cyst alone? Did he have an acute decompensation of a chronic hydrocephalus from concurrent bacterial meningitis in the context of a colloid cyst? This might have been clarified if dedicated CSF MRI sequences were done, e.g. CISS, during his admission and follow up.

Emboldened by initial results and obedience to parsimony, the surgeon may have wielded Ockham's sharpened razor to surgically resect this patient's colloid cyst. However, an understanding of diagnostic heuristics and an awareness of Hickam's dictum has stayed his hand, much to the benefit of the patient.

Diagnostic uncertainty is a reality of practising medicine. Cognitive biases have developed to deal with these uncertainties, and reflect the education and experiences to which the practitioner has been exposed. Awareness of these biases and their limitations is crucial to prevent inaccurate diagnoses and patient harm.[26]

This is true for the philosophy of Friar William of Ockham, an English Franciscan monk who in 1323 wrote, "*it is futile to do with more what can be done with fewer*". In medicine, this represents the urge to correlate multiple signs and symptoms into one unifying diagnosis.[5] In this case, hydrocephalus secondary to a colloid cyst. However, the counterargument from 20th century American physician Dr. John Hickam allows practitioners to embrace uncertainty and enhance the awareness of their own biases. This dictum ensured unnecessary surgery was avoided, and the correct disease treated.[6]

After removal of his EVDs and completion of antimicrobial therapy our patient fully recovered neurologically. This suggested the colloid cyst might have been an incidental finding and that

he had pneumococcal meningitis causing obstructive hydrocephalus. In short, he had as many diseases as he damn well pleased and thus strict outpatient follow-up is required.

Conclusion

Although these biases help practitioners to make sense of the chaotic world of medicine, they can be wrong and mistakes can be made. It is pertinent that doctors worldwide recognise that patients may not present in neat unifying diagnoses (Ockham's razor) but rather as a sum of their parts each of which may present a symptom or sign independent of one another (Hickam's dictum).

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Patient consent

We obtained written consent from the patient. This is available on request

Disclosure statement

No potential conflict of interest was reported by the authors.

Author contributions

GM (Conceptualization; Methodology; Investigation; Resources; Writing – Original Draft; Visualization). SL (Conceptualization; Methodology; Formal Analysis; Resources; Writing – Original Draft; Visualization; Supervision). SL scrutinized the whole manuscript and double checked it for accuracy and fluency. AK (Writing – Review & Editing). AH (Writing – Review & Editing). EJSt.G (Conceptualization; Supervision).

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ORCID

Gareth May http://orcid.org/0000-0002-4790-6446
Simon Lammy http://orcid.org/0000-0003-3199-0038
Ajay Hegde http://orcid.org/0000-0001-6970-2260

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