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CASE REPORT

Headache in a high school student – a reminder of fundamental principles of clinical medicine and common pitfalls of cognition

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ABSTRACT

Primary headache disorders account for the majority of the cases of headache. Nevertheless, the primary objective of a physician, when encountered with a patient with headache is to rule out a secondary cause the headache. This entails a search for specific associated red-flag symptoms or signs that may indicate a serious condition, as well as a heightened suspicion of and evaluation for a don't miss diagnosis.

We present a case of a high-school student whose first manifestation of systemic lupus erythematosus (SLE) was a headache due to cerebral venous and sinus thrombosis, initially misdiagnosed as tension-headache and 'ophthalmoplegic migraine' (now known as 'recurrent painful ophthalmoplegic neuropathy'). The patient made a complete neurological and radiological recovery after systemic anticoagulation and treatment of SLE. An analysis of the clinical errors and cognitive biases leading to delayed referral to hospital is presented. We highlight the fact that adherence to the fundamental principles of clinical medicine and enhancement of cognitive awareness is required to reduce diagnostic errors.

Keywords: Cerebral venous and sinus thrombosis, systemic lupus erythematosus, headache, nephrotic syndrome, principles of clinical medicine, cognitive errors

INTRODUCTION

Cerebral venous and sinus thrombosis (CVST) is an uncommon but potentially life threatening cerebro-vascular disease. Etiologies include prothrombotic disorders, neoplasia, vasculitis, connective tissue diseases, infections and head trauma. Patients with

CVST present with several distinct syndromes including isolated intracranial hypertension, focal neurological deficits, encephalopathy, and cavernous sinus syndrome[1]. Given the plethora of causes and presenting scenarios, CVST may be encountered not only by neurologists but also by general physicians, emergency physicians and general practitioners. Headache is the most common initial symptom of CVST but misdiagnosis and delay in diagnosis are common since there are no pathognomonic features.[2–4]

CASE REPORT

An 18-year-old male, previously healthy Asian student was admitted via the emergency department (ED) with headache of seven days' duration. It started as an intermittent right temporal headache which he ascribed to the stress of his A-level studies and coursework. He had been receiving extra hours of tuition to prepare for his A-level exams and IELTS (International English Language Testing System) tests. The pain was aggravated by walking upstairs and associated sometimes with nausea. Acetaminophen and Codeine based analgesics were prescribed for "tension headaches" by a local general practitioner, but these provided only partial relief. Two days before admission, the headache worsened, this time involving the right temporal, frontal and orbital region, associated with nausea and intermittent blurring of vision but no photophobia. He attended a private hospital where a suspicion of right abducens nerve palsy was raised. A CT scan of the head was done and reported as normal. Non-steroidal anti-inflammatory agents were prescribed for suspected "ophthalmoplegic migraine" and an urgent referral was made to the ED for expert ophthalmological review.

On examination in the ED, he was alert with stable vital signs. Mild puffiness of the face and pitting edema of the feet were noted. There was no fever, rash or neck stiffness. Neuro-ophthalmological examination revealed that his visual acuity was preserved and there was no relative afferent pupillary defect. However, he had left esotropia in primary gaze with the size of convergent squint larger on distance fixation. Ocular motility showed right abduction deficit, indicating right abducens nerve palsy. Optic fundoscopy demonstrated bilateral papilledema. These red-flag signs suggested a secondary cause of his headache.

Initial laboratory studies revealed high ESR (112 mm/1 hour), leukocytosis (16,000/mm^3) and thrombocytopenia (46,000/mm^3). Blood glucose was 5.2 mmol/L and serum creatinine 76 umol/L. Coagulation profile and liver function tests were normal. Lumbar puncture was not carried out in view of thrombocytopenia and papilledema. Brain CT-scan and MRI didn't illustrate any hemorrhage or parenchymal lesion but magnetic resonance venogram (MRV) demonstrated thrombosis of the inferior sagital sinus, right sigmoid sinus, jugular bulb and internal jugular vein (Figure 1A). Further laboratory tests revealed serum albumin of 13 g/L, total cholesterol 7.5 mmol/L and triglycerides 8.64 mmol/L. Urine dipstick analysis showed hematuria and proteinuria but no casts. Proteinuria was quantified at 16 grams/day, identifying nephrotic syndrome not only as the cause of his facial and pedal edema but also as a risk factor of CVST. Renal vein doppler studies were unremarkable. CVST, nephrotic syndrome and thrombocytopenia prompted investigations for systemic lupus erythematosus (SLE) and other pro-thrombotic disorders. Immunological screening revealed positive ANA and anti-dsDNA antibodies and low levels of both C3 and C4. Anti-phospholipid antibodies were negative; protein S activity was mildly reduced on functional assays, while protein C activity and anti-thrombin 3 levels were normal.

He was treated with systemic anticoagulation and steroids. Platelet count increased rapidly; a renal biopsy showed diffuse proliferative lupus nephritis. Mycophenolate mofetil was added and he was discharged three weeks after admission by which time his headache had subsided and diplopia had greatly improved. Six months after discharge, ophthalmological examination was normal and proteinuria had decreased to 0.9 g/day. A follow-up MRV showed almost complete recanalization of the veins and sinuses involved (Figure 1B).

DISCUSSION

This young student presented with one of the most common symptoms - headache, secondary to one of the uncommon and sinister but potentially reversible pathologies, CVST. Headache is a common symptom and doctors at busy workplaces are pressed for time. In addition, the relative rarity of CVST makes it less likely to be included in the list of differential diagnoses of a patient with headache. However, the fact remains

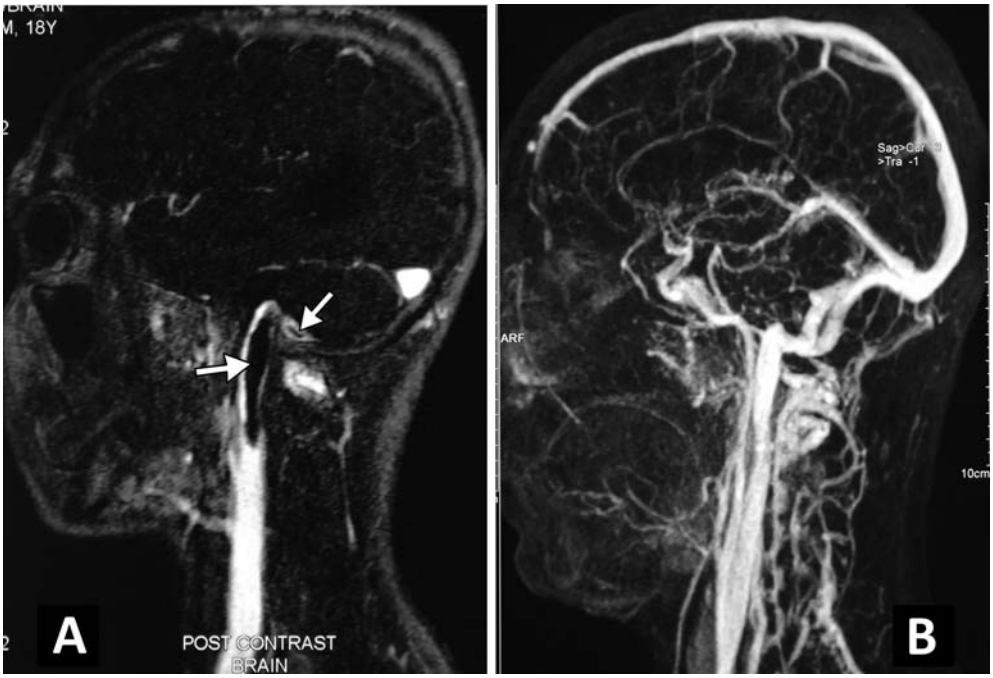

Figure 1. Cerebral venous and sinus thrombosis; A) MRV (source image) showing thrombus in right sigmoid sinus, jugular bulb and internal jugular vein. B) Follow-up MRV 6 months later demonstrating recanalization of the veins involved.

that the primary task of the physician is to identify red-flag signs that may indicate an underlying cause of secondary headache. History and physical examination should facilitate the generation of a differential diagnosis and preliminary classification of the headache type based on the criteria established by The International Headache Society.[2] By the time the patient presented to the ED, he had developed a full spectrum of signs and symptoms of raised intracranial pressure and it was obvious that he was harboring a serious pathology. However, observance of fundamental principles of clinical medicine and knowledge of cognitive biases would have led to an early referral and diagnosis of CVST, SLE and lupus nephritis.

First, a careful history taking is vital as some core aspects of history can facilitate differentiation between primary and secondary headaches. Though tension-type headaches are one of the commonest primary headache disorders, any new onset headache that is daily and persists should never be taken lightly. Tension-type headaches are non-disabling, bilateral, pressing/tightening in quality but generally *not* associated with nausea or vomiting.[2] Similarly, aggravation of pain by walking stairs or any other routine physical activity is *not* a feature of tension headaches. The presence of these symptoms in our patient should have been considered as red-flags.

Recurrent painful ophthalmoplegic neuropathy (the new medical term for ophthalmoplegic migraine) is thought to be an inflammatory demyelinating illness[5–7] that is associated with paresis of one or more of the cranial nerves developing during or within 24 hours of a severe attack of migraine. Headache usually is ipsilateral to the cranial nerve palsy. It is a very rare condition and the diagnosis requires exclusion of other parasellar, orbital fissure and posterior fossa pathologies.

Secondly, a thorough physical examination is crucial to identify causes of secondary headaches.
A neurological examination should include signs of meningeal irritation, pupillary size and light responses, extraocular movements and visual fields. In addition, optic fundi are checked for spontaneous venous pulsations and papilledema. Hasty clinical examinations done in busy clinics rarely include fundoscopy - a mandatory examination at first presentation with headache. Neurological examination is normal in patients with tension headaches. In recurrent painful ophthalmoplegic neuropathy, the pupillomotor fibers of the oculomotor nerve are most commonly involved resulting in mydriasis, ptosis and limitation of medial, upward, and downward gaze.[8] Involvement of abducens or trochlear nerve is uncommon. The clinical features in our patient were caused by impaired

absorption of CSF by CVST leading to raised intracranial pressure with consequent papilledema and stretching of the abducens nerve due to its anatomic location within the Dorello canal. Some patients do not mention a symptom if it is mild or if they think it is irrelevant. Facial puffiness and pedal edema were hence not picked up during the initial examinations. Detection of these physical signs earlier on might have prompted a search for a systemic disorder.

Finally, the physicians must be oriented to common pitfalls of cognition that can distort clinical reasoning leading to misdiagnosis.[9,10] Physicians, in their daily practices often rely on heuristics (experienced-based decisional shortcuts) to help them rapidly synthesize complex clinical information, generate diagnoses and devise treatment strategies.[11] However, because heuristics are intuitive rather than analytic processes, their use in clinical decision making can lead to cognitive errors. Most common cognitive errors arise from faulty data collection, defective interpretation or flawed reasoning. Doctors may miss important clues in history or misjudge the significance of a symptom/physical finding and hence, may not arrive at the correct diagnosis. In our patient, the salience of new-onset daily headaches, nausea and aggravation of headache by routine physical activity was not appreciated during the initial consultations.
One common cognitive error is 'anchoring' i.e., hastily and firmly latching on to a diagnosis that quickly comes to mind. Once a diagnosis is 'anchored', then the physician will have a tendency to cherry pick the information that fits the presumptive diagnosis (confirmation bias). Such cognitive error can frequently lead to a subsequent mistake e.g., failure to timely refer the patient to a specialist. Too much relevance was given to the stressful time that the patient was going through and based solely on this 'circumstantial evidence', a cause-effect relationship was (wrongly) established and a diagnosis of tension headaches made. This led to a premature closure of the diagnostic process, so that the differential diagnosis was never considered. We feel that physicians should keep an open mind by compiling a comprehensive differential diagnosis and should always take time to critically analyze and reflect on their working diagnosis. Though the first CT-scan in the private hospital did not reveal a space-occupying lesion, the physician obviously had inadequate level of comfort in diagnosing the headache and hence the patient was rightly referred for specialist's advice.

As compared to hospital-based doctors, the general practitioners are more likely to see patients early in the course of the disease; at a stage when the signs and symptoms of the disease may not be pronounced. It is therefore important to maintain suspicion for early disease and instruct the patient to return if the symptoms do not settle, get worse or new symptoms develop. In most cases, headache due to CVST is associated with signs of intracranial hypertension or other neurological features. However, the headache can precede the development of the neurologic signs by days or weeks.[2,12] Absence of seizures/mental status disorder in patients with CVST is known to lead to admission delay.[13] Given the absence of specific characteristics, any recent and persistent headache should lead to a thorough neuro-ophthalmological examination and neuroimaging to rule out CVST, particularly when the patient has a concomitant prothrombotic condition. This systematic approach led to the diagnosis of not only CVST but also the associated risk factor (nephrotic syndrome) and the underlying etiology (SLE) in our patient.

CONCLUSION

Technology has made fantastic strides in the diagnosis and treatment of many serious illnesses but to make the best use of these technological advances, the fundamental principles of clinical medicine must be followed. This should be coupled with efforts to improve cognitive awareness in order to reduce diagnostic failures.

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