Headache with Increased Intracranial Pressure

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Interesting case

A 27-year-old Thai female patient, a merchant's occupation, Nakhon Ratchasima.

Right: Universal Health Coverage Service (UC)

Chief complaint

Progressive headache and blurred vision for 1 month

Present illness

2 months before admission. She had a headache in the occipital region. The pain radiated to the center of the head, persistent dull aching type in all day. The severity of headache was not related to the posture of the body such as upright or supine. The pain score was 5-6 of 10 points by her visual analog scale. She had no nausea, no vomiting, no fever, no seizure and no motor weakness.

1 month before admission. The headache had increased the intensity of the pain. The pain score was increased to 8-9 of 10 points. It was a constant pain in everyday, sometimes the pain was so intense that she felt like syncope. Her headache was provoked by cough and sneeze. She also had blurred vision but no double vision. She took some migraine painkillers, but her symptoms didn't improve, therefore she came to the hospital.

Past history

- No underlying disease, no drug allergy, no herbal medicine use, no contraceptive pill use
- No smoking and drinking alcohol
- No intake raw food

Family history

- No history of chronic headache, no malignancy in family

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Physical examinations

General appearance: A Thai young aged woman, alert and well cooperate

Vital signs: BT 37.8°C, PR 86 bpm regular, RR 20 bpm, BP 130/80 mmHg, BMI: 23.9 kg/m^2

HEENT: pink conjunctivae, anicteric sclerae, no skin lesion, no lymphadenopathy, no parotid and thyroid gland enlargement, absent carotid bruit

Heart and Lungs: unremarkable

Abdomen: soft, not tender, no hepatosplenomegaly

Extremities: no pitting edema, no rash

Neurological examination: E4V5M6, alert, orientation to time, place and person

Cranial nerves: CN I: equally sense of smell, **CN II:** pupil 2 mm RTLBE, no RAPD, normal confrontation test, Gr I papilledema (mild disc edema with a normal temporal disc margin), **CN III, IV, VI:** extraocular muscles movement were intact, **CN V:** normal mastication muscles, intact pin prick and temperature sensation, **CN VII:** no facial weakness, **CN VIII:** normal Weber and Rinne test , **CN IX, X:** equally palatal movement, positive gag reflex, **CN XI:** head turning and shoulder shrug were intact, **CN XII:** no tongue atrophy, normal tongue movement

Motor: no muscle atrophy, no fasciculation, normal muscle tone, motor power grade V/V both sides

Sensory: intact pin prick, fine touch and proprioception

DTR: 2+ all

Cerebellar signs: intact FTNTF, no dysdiadochokinesia, normal HTK test

Babinski sign: plantar flexion, **Clonus:** negative, **Meningeal sign:** negative

Problem lists: Headache with increased intracranial pressure

Discussion

A young female patient presented with a chronic headache. The temporal progression of headache associated with secondary caused from increased intracranial pressure. The neurological examinations revealed papilledema in both eyes without meningeal signs and other focal neurological deficits. The mechanism of neurological dysfunctions that cause increased intracranial pressure were increased in CSF production or decreased in CSF absorption. The possible differential diagnoses include: idiopathic intracranial hypertension, cerebral venous sinus thrombosis, reduction in size of cranial vault (craniosynostosis), tumor producing CSF formation (choroid plexus papilloma), arachnoid granulation adhesions from post CNS infection, hypervitaminosis A and drug-induced increased intracranial pressure (oral contraceptives).

Lab investigation

Complete blood count: Hb 14.5g/dL, Hct 43.9%, WBC 8,500 /uL (PMN 69%, L 25%, Mono 4%, E 1%, B 1%), Platelet count 350,000 /uL, MCV 81.9 (80-98) fL, MCH 26.6 (25.6-32.2) pg/cell, MCHC 34.1 (32.2-36.5) g/dL, RDW 12.1% (11-14), hypochromia: few anisocytosis, few microcyte

Chemistry: BS 111 mg/dL, BUN 10 mg/dL, Cr 0.54 mg%, Na 138.2 mmol/L, K 3.95 mmol/L, Cl 104 mmol/L, CO2 23 mmol/L, Ca 8.9 mg/dL, P 3.2 mg/dL, Mg 2.0 mg%

Liver function test: TP 6.9g%, Alb 4.2 g%, Glb 2.7 g/dL, TB 0.4 mg%, DB 0.2 mg%, AST 16 U/L, ALT 8 U/L, ALP 49 U/L

Lipid profile: CHO 180 mg%, TG 94 mg%, HDL 48 mg%, LDL 113.2 mg%

Pregnancy test negative, Anti-HIV: non-reactive, VDRL: non-reactive

Protein C 122% (70-140%), Protein S activity 87% (59-118%), Factor V Leiden PCR: negative, Anti-Beta 2 glycoprotein, IgM, IgG: negative, Lupus (1:50) 1.21 (0.44-1.40), Lupus (1:500) 1.09 (0.48-1.32), dRVVT ratio 1.09 (<1.31), PT 11.8 second (9.90-12.20), PTT 22.3 second (22.0-29.3), INR 1.11

CSF profiles (at admission)

Color	Clear colorless
OP/LP	32/18 mmH2O
Clot formation	Absence
pH	8
Specific gravity	1.006
WBC	5 cell/mm^3 (L 100%)
RBC	0
Protein	25 mg/dL (50-70 mg/dL)
Sugar ratio	52.6%
Gram's stain	Not found organism
Aerobic culture	No growth

EKG: normal sinus rhythm, rate 82/min regular, no ischemic pattern, no chamber enlargement

CXR: normal cardio-thoracic ratio, normal parenchymal of both lungs

MRI brain: MRI Brain (Figure 1): Axial T2-weighted MR images (a-c) demonstrated an absence of normal flow void on the dural sinuses at superior sagittal sinus, left sigmoid and left internal jugular veins. Axial gadolinium enhanced T1-weighted MR images (d-f) showed heterogeneous enhancement of intraluminal clots.

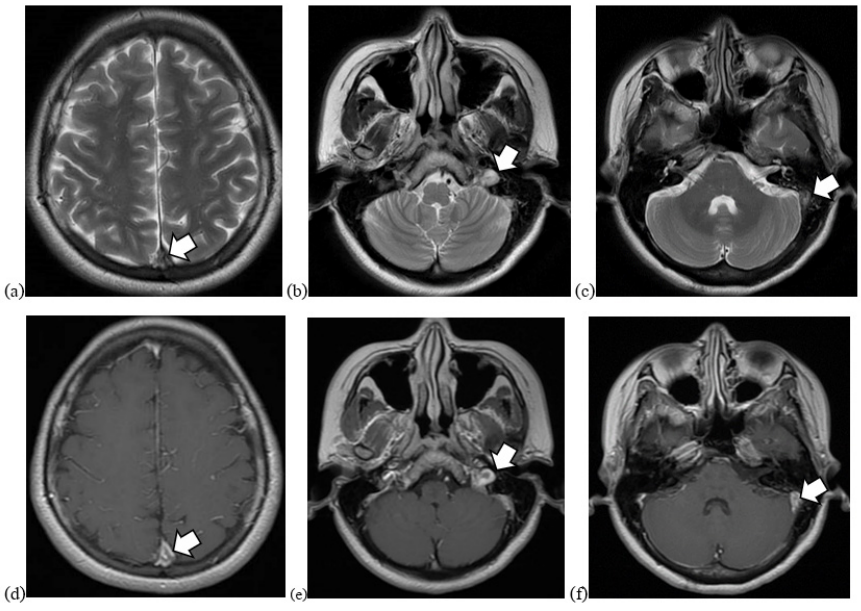

Figure 1 Axial T2-weighted (a-c) and gadolinium enhanced T1-weighted MR images (d-f) (back cover page)

MRV Brain (figure 2): Sagittal gadolinium enhanced MRV images (a-d) revealed multiple intraluminal filling defect along posterior superior sagittal sinus to torcular herophili (a), straight sinus (b), bilateral transverse sinuses (c), left sigmoid sinus (d) down to bilateral internal jugular veins (c).

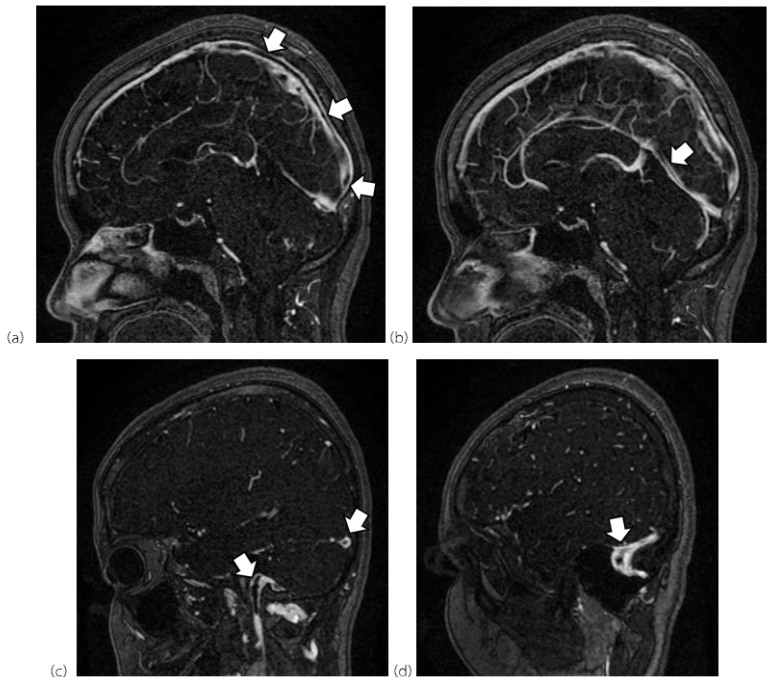

Figure 2 Sagittal gadolinium enhanced MRV images (cover page)

MRI Brain (Figure 3): A small hyperintense lesion at subcortical white matter of left temporal lobe on axial T1-weighted (a) and T2 FLAIR weighted (b) MR images with perilesional edema and heterogeneous dark foci on axial SWI (c), representing small late subacute hemorrhage. There are also irregular dark foci at right (d) and left (e) parasagittal high frontal lobes.

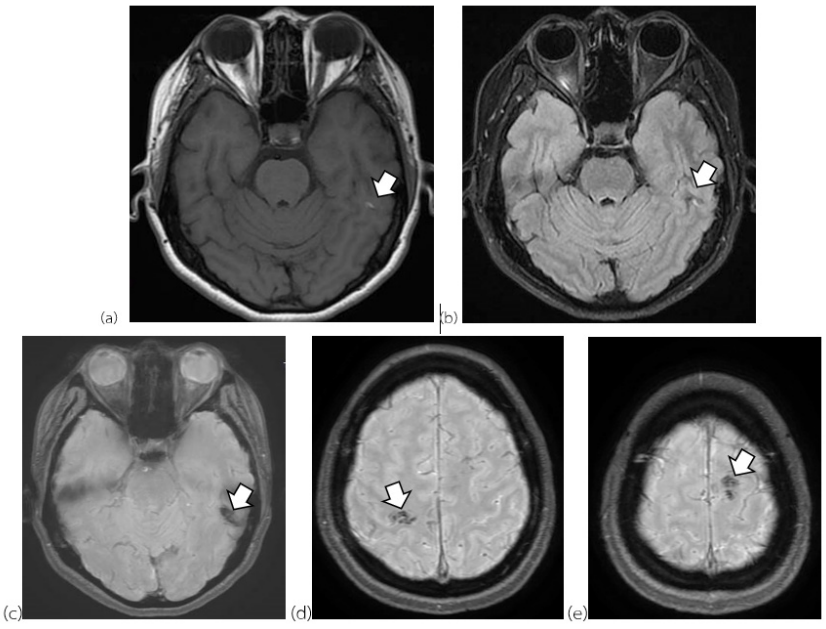

Figure 3 T1-weighted, T2 FLAIR weighted and SWI MR images (back cover page)

MRI Brain (Figure 4): Axial T2-weighted MR image show flattening of the posterior sclera, minimal protrusions of the bilateral optic nerve heads and mild tortuosity of the optic nerves, corresponding with papilledema insult from increase intracranial pressure.

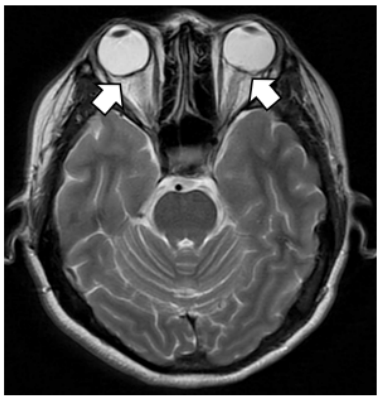

Figure 4 Axial T2-weighted MR image

Diagnosis: Chronic dural venous sinus thrombosis

Progress note:

From the results of the blood and CSF analysis, it was found that the patient had no evidence of infection. She was treated with initial anticoagulation with LMWH in full anticoagulant doses, followed by oral vitamin K antagonist with a target INR of 2.0 to 3.0. She was initially given acetazolamide to alleviate the effects of increased intracranial pressure. No complications were observed during treatment such as seizures, progressive intracranial bleeding and focal neurological deficit.

Conclusion

Although cerebral venous sinus thrombosis is an uncommon but potentially serious and life-threatening cause of severe headache and stroke. On the basis of findings for history taking and neurological examination are reasonable for the initial investigation and management of disease to optimize care and minimize complications. Additional specific treatment input as needed to provide therapeutic anticoagulation is appropriate.

Dural venous sinus thrombosis

Pathomechanisms[1-3]:

The current knowledge is based on available animal model studies. There were shown that sinus thrombosis leads to sinus occlusion and backflow of blood into venules and capillaries, resulting in increased local pressure, eventually resulting in massive cerebral edema. Extensive anastomoses found within the cerebral venous system often allow the development of collateral circulation if there is an occlusion to venous flow. Cortical vein occlusion can result in increased small vessel pressure, depending on the extent of thrombosis and availability of collaterals. This can further lead to disruption of the blood-brain barrier, leakage of blood components into the interstitial space causing vasogenic edema, and parenchymal tissue damage. Decrease in the CSF drainage due to sinus occlusion can result from the dysfunction of arachnoid granulations, causing an increase in intracranial pressure. A continuous rise in pressure results in capillary hypertension, cerebral edema, and venous hemorrhage as reported by the patient.

Clinical Features[4,5]:

The clinical manifestation is depending on the site and extent of thrombosis, patient's age, and the underlying etiological factors. The most common presentation includes signs of increased intracranial pressure such as headache, reduced visual acuity, and papilledema, focal neurological deficits, seizures, and diffuse encephalopathy. Other

uncommon presentations include subarachnoid hemorrhage, thunderclap headache, recurrent transient ischemic attacks, tinnitus, isolated headache, and multiple cranial nerve palsies. The patient's symptoms in this report were no different from the previous studies.[5,6]

Risk factors[6-9]:

These risk factors are usually always associated with thrombogenic triad of Virchow (vessel wall injury, blood stasis, and hypercoagulability). The other important causes including:

- Prothrombotic conditions: Hereditary (factor V Leiden mutation, antithrombin III deficiency, protein C-S deficiency, G20210A prothrombin gene mutation) and acquired: female sex, oral contraceptive pill, hormone replacement therapy
- Infections: previous meningitis, HIV, tuberculosis, otitis media, mastoiditis
- Inflammations: vasculitis, SLE, Sjogren's syndrome, temporal arteritis, antiphospholipid syndrome, Wegener's granulomatosis, Bechet's disease
- Trauma: traumatic brain injury, jugular vein catherization
- Intracranial abnormality: tumor, arteriovenous malformation, dural venous fistula, venous anomalies
- Hematologic problems: essential thrombocytopenia, polycythemia, paroxysmal nocturnal hemoglobinuria
- Systemic diseases: dehydration, malignancies, sarcoidosis, obesity
- Drugs: tamoxifen, glucocorticoids, hemostatic therapy, cyclosporine

Treatments[5, 9-11]:

The initial treatment will consider anticoagulant therapy with the goal of treatment including to prevent thrombus growth, to facilitate recanalization, and to prevent deep vein thrombosis or pulmonary embolism. Although there are other treatment methods. However, it was found that the results of treatment may be different in outcomes.[2,5] The complications that may be encountered include: seizure, hydrocephalus and intracranial hypertension.

Anatomy of the cerebral venous system (Figure 5) Cerebral venous system is divided into the superficial and deep venous system. Cerebral veins lack valves and do not follow cerebral arterial territory. The superior sagittal sinus, also drains the CSF from the subarachnoid space. The superficial system comprises the dural sinuses and cortical veins. It drains the cerebral cortex and superficial white matter. The two major dural sinuses include the superior sagittal sinus that drain dorsolateral area and the cavernous sinus that drain the anteroventral region. The superior sagittal sinus drains into the transverse sinus which then drains into the straight sinus. The cavernous sinus drains into the transverse sinus posterolaterally and sigmoid sinus inferolaterally, along the superior and inferior petrosal sinuses, respectively. The superficial cortical veins are the superiorly draining veins and the inferiorly draining veins (vein of Labbe and sylvian or superficial middle cerebral veins). The deep system includes straight, lateral, and sigmoid sinus, as well as drain the deeper cortical veins (vein of Galen, internal cerebral veins, Rosenthal or basal vein, medullary, and subependymal veins). These vessels drain the basal ganglia, thalamus, the upper brain stem, and deep brain white matter. Both the superficial and deep venous systems eventually drain into the internal jugular veins.

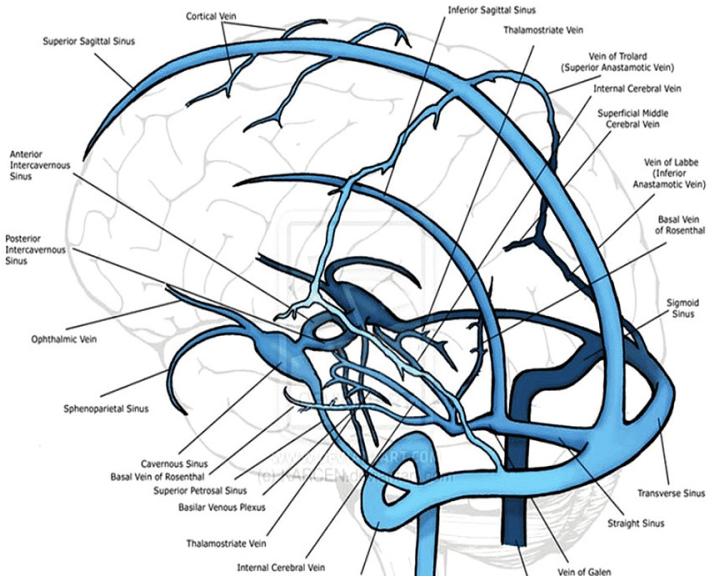

Figure 5 Dural sinuses and encephalic veins anatomy
(https://www.researchgate.net/figure/Dural-sinuses-and-encephalic-veins-anatomy)

References

1. Idiculla PS, Gurala D, Palanisamy M, Vijayakumar R, Dhandapani S, Nagarajan E. Cerebral venous thrombosis: A comprehensive review. Eur Neurol 2020;83:369-79. doi: 10.1159/000509802
2. Ferro JM, Canhão P. Cerebral venous sinus thrombosis: update on diagnosis and management. Curr Cardiol Rep 2014;16:523.
3. Alvis-Miranda HR, Milena Castellar-Leones S, Alcala-Cerra G, Rafael Moscote-Salazar L. Cerebral sinus venous thrombosis. J Neurosci Rural Pract 2013;4: 427-38
4. Bousser MG, Ferro JM. Cerebral venous thrombosis: an update. Lancet Neurol 2007;6:162-70.
5. Saposnik G, Barinagarrementeria F, Brown RD Jr, Bushnell CD, Cucchiara B, Cushman M, et al. Diagnosis and management of cerebral venous thrombosis: a statement for healthcare professionals from the American Heart Association/American Stroke Association. Stroke 2011;42:1158-92.
6. Dmytriw AA, Song JSA, Yu E, Poon CS. Cerebral venous thrombosis: state of the art diagnosis and management. Neuroradiology 2018 ;60:669-85.
7. Ferro JM, Canhão P, Aguiar de Sousa D. Cerebral venous thrombosis. Presse Med 2016 ;45:e429-50.
8. Coutinho JM, Ferro JM, Canhão P, Barinagarrementeria F, Cantú C, Bousser MG, et al. Cerebral venous and sinus thrombosis in women. Stroke 2009;40:2356-61.
9. Zuurbier SM, Arnold M, Middeldorp S, Broeg-Morvay A, Silvis SM, Heldner MR, et al. Risk of cerebral venous thrombosis in obese women. JAMA Neurol 2016;73: 579-84.
10. Korathanakhun P, Petpichetchian W, Sathirapanya P, Geater SL. Cerebral venous thrombosis: comparing characteristics of infective and non-infective aetiologies: a 12-year retrospective study. Postgrad Med J 2015;91:670-4.
11. Ferro JM. Causes, predictors of death, and antithrombotic treatment in cerebral venous thrombosis. Clin Adv Hematol Oncol 2006; 4:732-3.