Neurology Asia 2024; 29(1) : 277 – 281

IMAGING HIGHLIGHTS

Unilateral thrombosis of dominant internal jugular vein presenting with benign intracranial hypertension

[1]Ranchini Murgan *MS*, [1]Sutharshan Sockalingam *MRCS*, [2]Khairul Azmi Abd Kadir *MRad*, [3]Khean Jin Goh *FRCP*, [4]Visvaraja Subrayan *FRCS*, [5]Bee Ping Chong *MMed*, [1]Dharmendra Ganesan *FRCS(Neusosurg)*

[1]Division of Neurosurgery, University of Malaya Medical Centre, Kuala Lumpur, Malaysia; [2]Department of Biomedical Imaging, University of Malaya Medical Centre, Kuala Lumpur, Malaysia; [3]Division of Neurology, University of Malaya Medical Centre, Kuala Lumpur, Malaysia; [4]Department of Ophthalmology, University of Malaya Medical Centre, Kuala Lumpur, Malaysia; [5]Division of Haematology, University of Malaya Medical Centre, Kuala Lumpur, Malaysia

Benign intracranial hypertension (BIH) is defined as a chronic elevation of intracranial pressure (ICP) divided into primary, idiopathic intracranial hypertension, and secondary forms.

Idiopathic intracranial hypertension is defined as an elevation of intracranial pressure (ICP) with no identifiable aetiology.[1] The term 'pseudotumor cerebri' or BIH, was commonly used in the past for idiopathic intracranial hypertension, but is now used to describe the chronic elevation of ICP regardless of its aetiology and is further divided into primary (idiopathic intracranial hypertension) and secondary forms.

We report a rare case of BIH secondary to unilateral dominant internal jugular vein thrombosis.

CASE REPORT

A 33-year-old lady with a history of polycystic ovarian syndrome (PCOS) on oral contraceptive pills (OCP) presented with worsening neck pain for 2 months. After about 10 weeks of enduring the pain, she developed blurring of vision. There was bilateral papilledema. Her weight was 85 kg, and her BMI was 29kg/m^2, which was overweight, bordering on obesity. The blood pressure was 152/98mmHg. The neurological examinations were unremarkable. She had no symptoms or signs of connective tissue disease.

Computed tomography (CT) and magnetic resonance imaging (MRI) of the brain and spine did not detect any overt abnormalities. The opening pressure was 55cmH_2O on the lumbar puncture. The cerebrospinal fluid biochemistry was normal, and the culture had no infection. The blurring of vision resolved following the first lumbar puncture, only to recur after one week. She was commenced on Oral Acetazolamide 1g QID, a carbonic anhydrase inhibitor, which did not help her symptoms much.

Magnetic resonance venography (MRV) brain with contrast was done for further investigation, which revealed a filling defect in the proximal part of a dominant right internal jugular vein (IJV) at the flexure of the sigmoid sinus to the IJV, as seen in Figure 1-4. She had thrombosed the dominant right IJV. The other intracranial venous sinuses were patent.

In view of the progressive blurring of vision and florid papilledema, which did not respond to medical management, she required a cerebrospinal fluid diversion procedure. She had a lumbar peritoneal shunt using a programmable shunt *(MEDTRONIC Strata valve*) which was set at 1.0. All the symptoms improved a few days after the shunt.

The hematologist detected she had heterozygous Factor V Leiden mutation (R506Q). Lupus anticoagulant screening was mildly positive. Serum homocysteine level was mildly elevated. Antiphospholipid antibodies and cardiolipin antibodies were all negative. She was started on anticoagulant therapy, oral Rivaroxaban one week following surgery.

Repeated MRV (Figure 5 – 7) with contrast after 3 months on anticoagulant treatment showed a smaller filling defect in the right proximal internal jugular vein with the rest of the deep venous sinuses preserved.

Address correspondence to: Dr Dharmendra Ganesan, Department of Surgery, University of Malaya. Kuala Lumpur, Malaysia. Email: dharmendra@um.edu.my

Date of Submission: 4 March 2023; Date of Acceptance: 21 June 2023

https://doi.org/10.54029/2024jwr

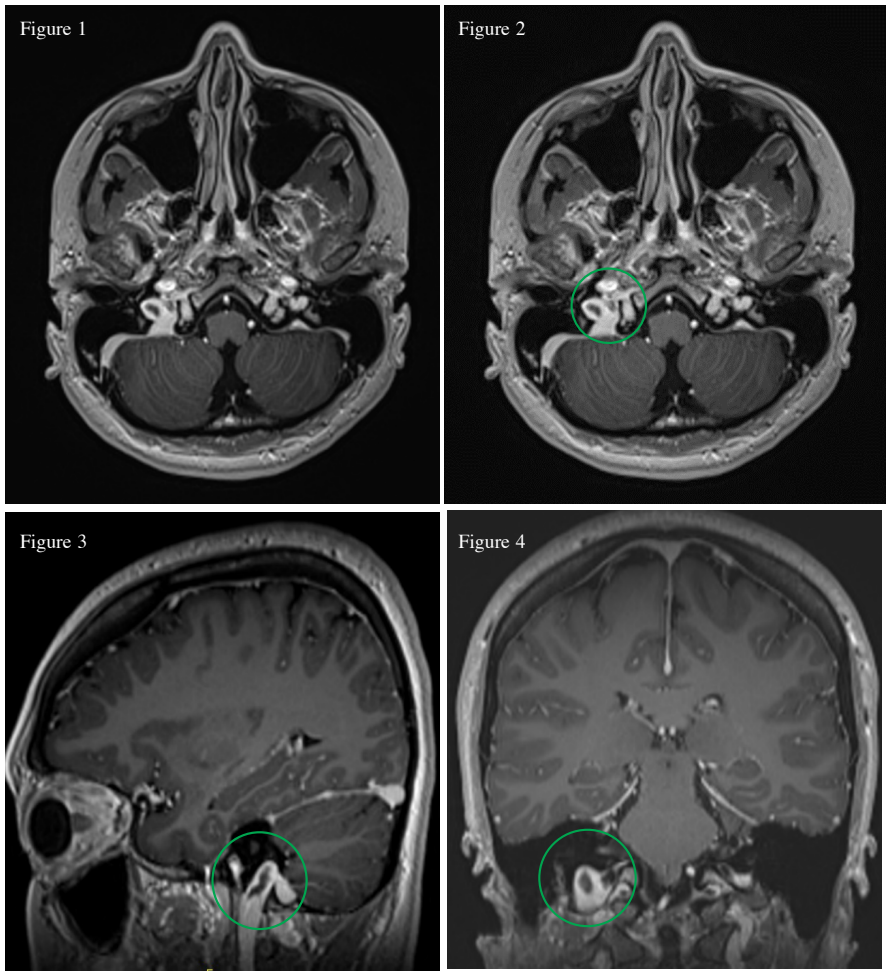

Figure 1-4. MRV brain with contrast showing long segment filling defect in the right proximal internal jugular vein which indicates thrombosis

MRV with contrast (Figure 8 – 10) after 9 months showed complete resolution of the thrombosis. MRV with contrast in the 3D image depicts the stenosis of the right proximal IJV at presentation (Figure 11) and resolution of stenosis 9 months later after treatment (Figure 12).

DISCUSSION

This case illustrates that the initial standard MRI screening missed the secondary cause of BIH. The reason was that the thrombosis was very close to the skull base as the sigmoid sinus turned to become the IJV. Once the index of suspicion was cast, the scrutiny of the MRV brain with contrast at the proximal part of the dominant right IJV showed a distinct thrombosis at the junction of the sigmoid sinus and IJV. The clinical symptom was pronounced as the thrombosis involved the larger IJV, which was the dominant cerebral venous drainage. About 68-76% of the right IJV is dominant.[2-4]

The presence of Leiden mutation has a 1.4 to 1.6-fold increase in the risk of recurrent venous thromboembolism compared to patients without thrombophilia.[5]

In patients with threatened vision, a cerebrospinal fluid diversion will be required.[3,6,7] Lumbar peritoneal drain insertion was preferred in this patient because the ventricles were narrow. The literature has also reported the lumbar peritoneal shunt as a safe and effective mode of treatment for idiopathic intracranial hypertension.[6,8] We chose a programmable shunt rather than a fixed pressure shunt; to enable adjustment of the pressure to customise to the patient's cerebrospinal pressure dynamics. It has been reported that rapid reduction of cerebrospinal fluid in patients with chronic IIH cases can lead to encephalopathic changes and an altered conscious state.[9]

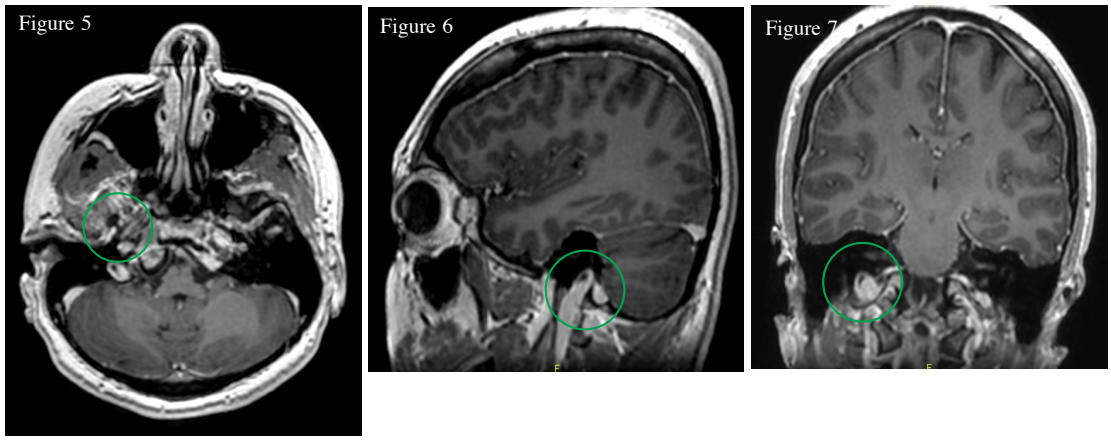

Figure 5-7. MRV brain with contrast showing smaller residual right proximal internal jugular vein thrombosis

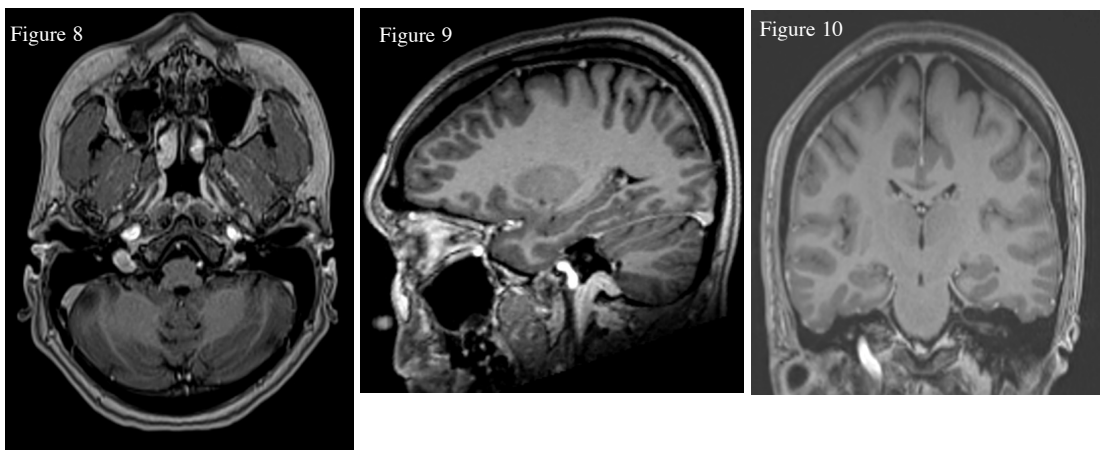

Figure 8-10. MRV brain with contrast showing resolved right internal jugular vein thrombosis

Endovascular stentings have been reported in a few series as definitive treatment for IJV or cerebral venous sinus stenosis.[10-13] It was not employed in this case as the interventional radiologist was concerned about the flexure at the sigmoid sinus – IJV junction.

The definite management of internal jugular vein thrombosis with associated heterogenous Factor V Leiden mutation was the commencement of anticoagulant.[3,5] One week after the lumbar peritoneal drain insertion, she was on a novel oral anticoagulant (NOAC), Rivaroxaban.

IJV thrombosis is a potentially life-threatening condition. The incidence of IJV thrombosis was reported with a very low frequency (1.7%).[3,14] The incidence of the dominant IJV thrombosis leading to benign intracranial hypertension (BIH) is unclear, apart from a few case reports.[6,7,15] About 20% of IJV thrombosis is primary, with the common secondary causes being cancer, central venous catheter and ovarian hyperstimulation syndrome.[16,17] With regard to treatment, trials are limited, and many studies have indicated that in the majority of such cases, treatment options have been directed towards central vein catheterisation and anticoagulation treatment.[3]

In conclusion, this case illustrates a rare cause of BIH, which is unilateral thrombosis of the proximal part of a dominant internal jugular vein. It is essential to scrutinize the sigmoid sinus – IJV junction, particularly in cases with a dominant IJV. The best imaging modality to detect this condition at this location would be an MRV brain with contrast.

DISCLOSURE

Financial support: None

Conflict of interest: None

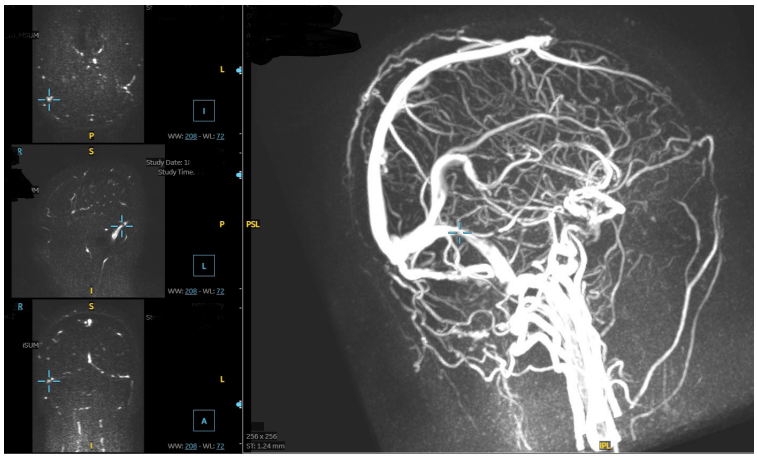

Figure 11. 3D venogram derived from MRV brain with contrast showing right internal jugular vein thrombosis (cross mark)

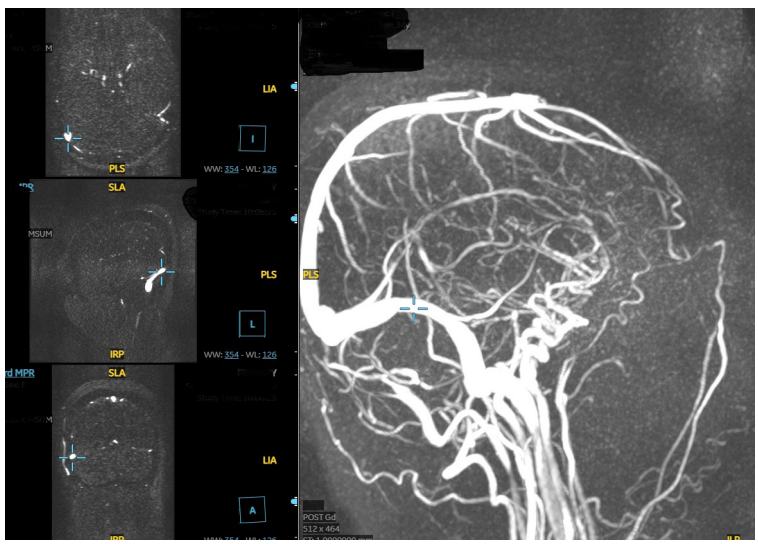

Figure 12. 3D venogram derived from MRV brain with contrast showing resolution of the stenosis secondary to the thrombosis at the right internal jugular vein thrombosis (cross mark)

REFERENCES

1. Mollan SP, Ali F, Hassan-Smith G, Botfield H, Friedman DI, Sinclair AJ. Evolving evidence in adult idiopathic intracranial hypertension: pathophysiology and management. *J Neurol Neurosurg Psychiatry* 2016; 87(9):982-92. DOI: 10.1136/jnnp-2015-311302
2. Lichtenstein D, Saïfi R, Augarde R, *et al*. The internal jugular veins are asymmetric. Usefulness of ultrasound before catheterization. *Intensive Care Med* 2001; (1):301-5. DOI: 10.1007/s001340000792
3. Scerrati A, Menegatti E, Zamboni M, *et al*. Internal jugular vein thrombosis: Etiology, symptomatology, diagnosis and current treatment, *Diagnostics (Basel)* 2021; 11(2):378. DOI: 10.3390/diagnostics11020378
4. Tenenbein P, Godhke B, Britz G, Sekhar L, Lam A. Internal jugular vein diameter on ultrasound predicts jugular bulb dominance. *J Neurosurg Anesthesiol* 2006;18:304.
5. White CW, Thomason AR, Prince V. Recurrent venous thromboembolism in a patient with heterozygous factor V Leiden mutation. *Hosp Pharm* 2014; 49(8):748-51. DOI: 10.1310/hpj4908-748
6. Konrad J, Vogt R, Helbig H, Oberacher-Velten I. Intrakranielle Hypertension und Jugularvenenthrombose. *Der Ophthalmologe* 2015;112(12):1002-5. DOI: 10.1007/s00347-015-3248-8.
7. Thandra A, Jun B, Chuquilin M. Papilloedema and increased intracranial pressure as a result of unilateral jugular vein thrombosis. *Neuroophthalmology* 2015; 39(4):179-82. DOI: 10.3109/01658107.2015.1044541
8. Yadav YR, Parihar V, Agarwal M, Bhatele PR, Saxena N. Lumbar peritoneal shunt in idiopathic intracranial hypertension. *Turk Neurosurg* 2012; 22(1):21-6. DOI: 10.5137/1019-5149.JTN.4307-11.1
9. Yahanda AT, Shah AS, Hacker C, Akbari SH, Keyrouz

S, Osbun J. Custom shunt system for increased baseline intracranial pressure in a patient with idiopathic intracranial hypertension. *World Neurosurg* 2020; 136:318-22. DOI: 10.1016/j.wneu.2020.01.142
10. Huang PY, Yang YH, Lin WC, Chen CH, Lin RT. Successful treatment of cerebral venous thrombosis associated with bilateral internal jugular vein stenosis using direct thrombolysis and stenting: A case report. *Kaohsiung J Med Sci* 2005; (11):527-31. DOI: 10.1016/S1607-551X(09)70162-7
11. Liu X, Di H, Wang J, *et al*. Endovascular stenting for idiopathic intracranial hypertension with venous sinus stenosis. *Brain Behavior* 2019; 9(5):e01279. DOI: 10.1002/brb3.1279
12. Yan F, Rajah G, Ding YC, *et al*. Safety and efficacy of intravascular ultrasound as an adjunct to stenting for cerebral venous sinus stenosis–induced idiopathic intracranial hypertension: a pilot study. *J Neurosurg* 2020; 132(3):749-54. DOI: 10.3171/2018.11.JNS181885
13. Zhou D, Meng R, Zhang X, *et al*. Intracranial hypertension induced by internal jugular vein stenosis can be resolved by stenting. *Eur J Neurol* 2018; 25(2):365-e13. DOI: 10.1111/ene.13512
14. Diebold J, Löhrs U. Venous thrombosis and pulmonary embolism. *Pathol Res Pract* 1991;187(2-3):260-6. DOI: 10.1016/S0344-0338(11)80781-8
15. Park G, Fleifel M, Kesserwani HN. Pseudotumor cerebri secondary to jugular bulb thrombosis: A case report and a review of the diagnostic steps. *Cureus* 2022; 14(8):e27557. DOI: 10.7759/cureus.27557
16. Gbaguidi X, Janvresse A, Benichou J, Cailleux N, Levesque H, Marie I. Internal jugular vein thrombosis: outcome and risk factors. *QJM* 2011; 104(3):209-19. DOI: 10.1093/qjmed/hcq179
17. Matsuda W, Noguchi S, Fujiyama F. Pseudotumor cerebri and lung cancer-associated jugular vein thrombosis: Role of anatomical variations of torcular herophili. e*NeurologicalSci* 2018; 13:18-20. DOI: 10.1016/j.ensci.2018.11.002