CASE REPORT

Venous sinus thrombosis secondary to tuberculous meningitis: a novel cause of trigeminal neuralgia

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SUMMARY

A 33-year-old Vietnamese man with a history of superior sagittal sinus thrombosis secondary to tuberculous meningitis presented with severe recurrent trigeminal neuralgia. A complex compensatory cerebral venous drainage system developed, and a varix, in direct contact with the root entry zone of the trigeminal nerve, was the trigger for his pain. The patient's symptoms are presently controlled with a combination of medications for neuropathic pain, however he continues to experience frequent severe exacerbations and may very well require more invasive means to manage his condition in future.

BACKGROUND

Trigeminal neuralgia (TN) is a dreadful condition that results in significant physical and psychological impairment to those affected by it. It is characterised by paroxysmal attacks of severe, short, sharp, stabbing pain, affecting one or more divisions of the trigeminal nerve, most commonly the second and third (V2 and V3).[1] The aetiology is typically from root entry zone (REZ) compression either by an adjacent blood vessel (most commonly the superior cerebellar artery) or cerebellopontine angle (CPA) lesion. Alternatively it may be secondary to a demyelinating process, such as multiple sclerosis. TN caused by a venous varix has rarely been reported in the literature. Matsushige *et al*[2] described a case of TN secondary to a varix associated with a tentorial dural arteriovenous malformation, which was successfully treated with stereotactic radiosurgery. In this case the mechanism of TN was from compression of the REZ of the trigeminal nerve by a high-flow, pulsatile, arterialised varix. The unique aspect of our described case is that the varix is part of a low-flow system with passive drainage of brain parenchyma. It is therefore felt that the aetiology of the patient's symptoms is secondary to direct mass effect on the trigeminal nerve from the varix. Treatment of this case represents a major challenge, given the significant risk of acute venous drainage insufficiency, which could result in massive venous infarction should it become necessary to sacrifice the varix during surgery.

CASE PRESENTATION

A 33-year-old Vietnamese man was referred for consideration of surgical intervention for his intractable left-sided facial pain. He had contracted tuberculosis (TB) as a child, diagnosed on screening tests when he came to Australia with his family as a refugee. At the time he did not have clinical manifestations suggestive of TB meningitis. He was started on anti-TB medications but complete eradication was not achieved.

As an adolescent, he presented to hospital with severe headache and on a CT scan of his brain, including venogram, he was found to have superior sagittal sinus (SSS) thrombosis thought secondary to TB meningitis. At the time, there was no evidence of anomalous venous drainage on imaging. He was started on warfarin and treated again for his TB with resultant eradication.

Ten years later he was assessed by an ophthalmologist due to blurred vision and headaches and was found to have acute or chronic papilloedema from presumed raised intracranial pressure. He was admitted to hospital and underwent a therapeutic lumbar puncture confirming intracranial hypertension. His symptoms improved and he was discharged with planned ongoing follow-up as an outpatient.

At the age of 30, he was once again admitted to hospital with intractable left-sided facial pain and classic features of TN involving the V3 distribution. At the time, he was unable to open his mouth or speak due to severe lancinating pain and was treated with a combination of carbamazepine, gabapentin, ketamine and fentanyl, before pain relief was achieved. He was eventually discharged home on a regimen of carbamazepine and gabapentin.

Over the next few years, the patient developed recurrent episodes of severe TN, thus he was referred for a neurosurgical opinion. Invasive options have been discussed, including percutaneous glycerol rhizotomy and microvascular decompression (MVD) but given the significant risk should he have a complication, a conservative approach continues with optimisation of his medications in conjunction with the pain management unit.

INVESTIGATIONS

A cerebral angiogram and MR venogram demonstrate lack of venous drainage into the major dural venous sinuses and a resultant compensatory venous drainage system, through dilated superficial and deep cerebral vessels (figures 1A–C and 2A, B). On MRI, a left-sided varix is noted to be in contact with the REZ of the trigeminal nerve, which is the cause of the patient's TN (figure 3A, B). His most recent imaging reveals stable appearance in the area of conflict but there was an incidental finding of two small aneurysms arising from the left internal carotid artery (figure 4). These were not visible on previous vascular imaging and thus are felt to represent de novo aneurysms rather than mycotic aneurysms.

TREATMENT

During his most recent exacerbation, the patient was admitted and managed in conjunction with the

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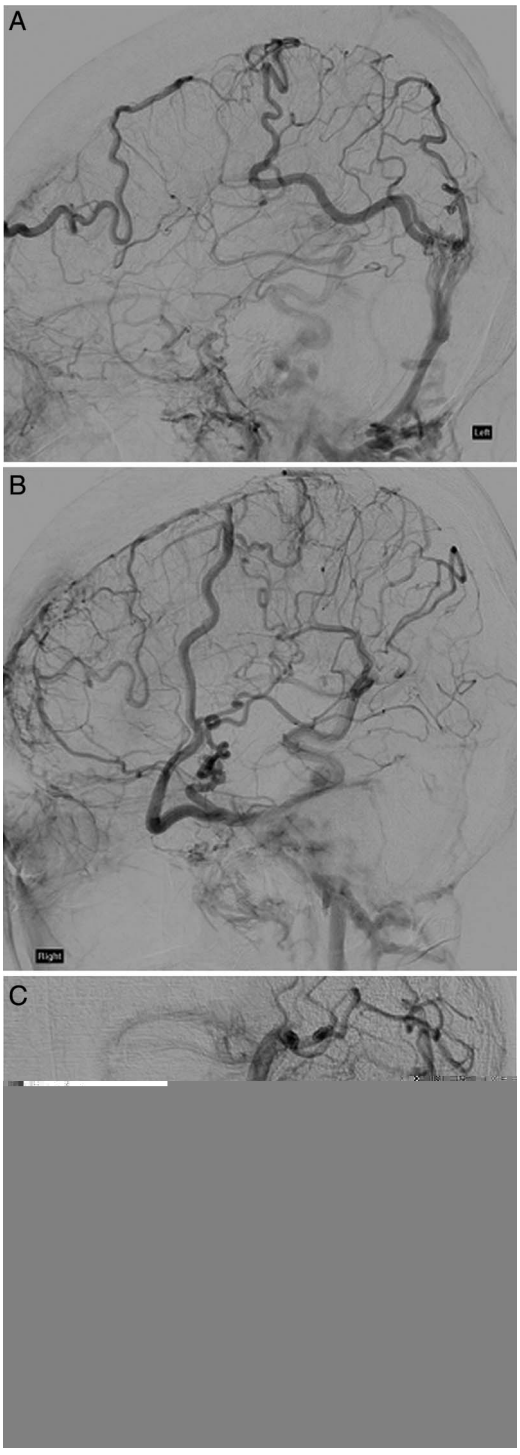

Figure 1 Digital subtraction angiogram of cerebral vessels demonstrating lack of drainage through major dural venous sinuses and resultant dilated compensatory superficial and deep venous drainage; (**A**) left internal carotid artery (ICA) injection lateral view, (B) right ICA injection lateral view, and (C) left vertebral artery injection lateral view.

pain management unit. Initially he was only able to communicate by writing on a board and he was unable to open his mouth for food or drink due to significant lancinating pain. His medications including pregabalin, baclofen and carbamazepine were up-titrated. An intravenous infusion of lignocaine was administered under close observation in the pain unit and his symptoms settled.

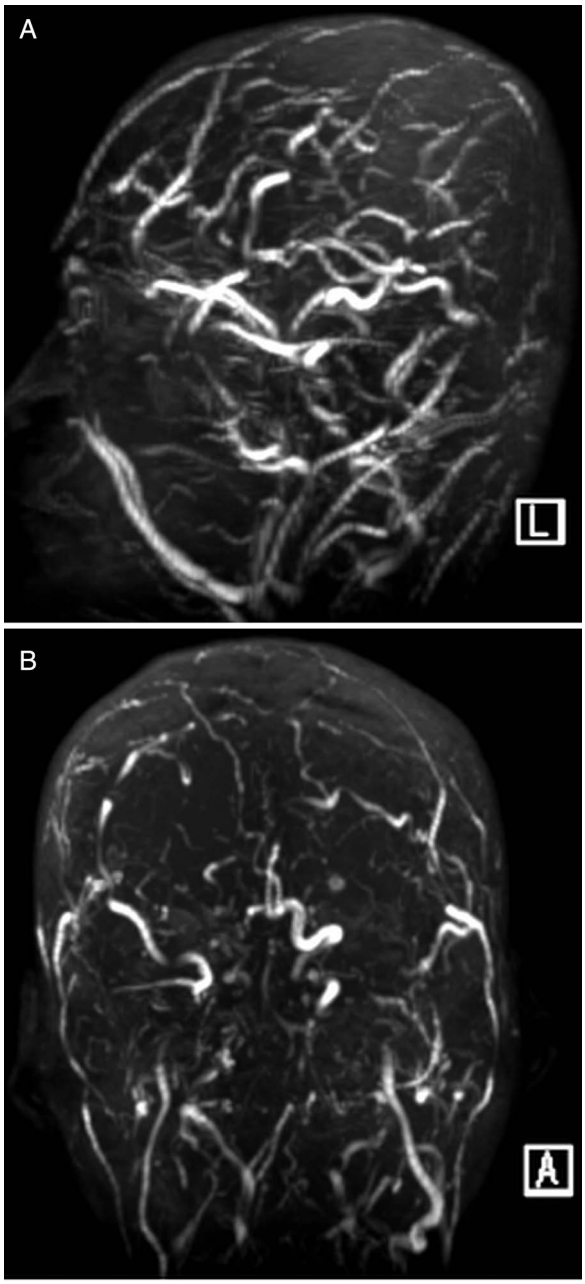

Figure 2 Three dimensional reconstruction of MR venogram again demonstrating the anomalous compensatory venous drainage system; (A) lateral view, and (B) anteroposterior view.

OUTCOME AND FOLLOW-UP

Given his exacerbations, and impact on his physical and mental well-being, the option of MVD has been considered. The concern in this case is that injury of any veins that would necessitate sacrificing the involved vessel, would likely result in massive venous infarction, given that his brain is heavily reliant on this compensatory venous drainage system. Nevertheless, should his pain no longer be responsive to maximal medical therapy, we may accept this risk and proceed to surgery. In the interim he will be followed up as an outpatient with planned annual surveillance imaging given his incidental aneurysms.

DISCUSSION

Most cases of TN are caused by compression at the REZ of the trigeminal nerve, although cases where there is no such conflict

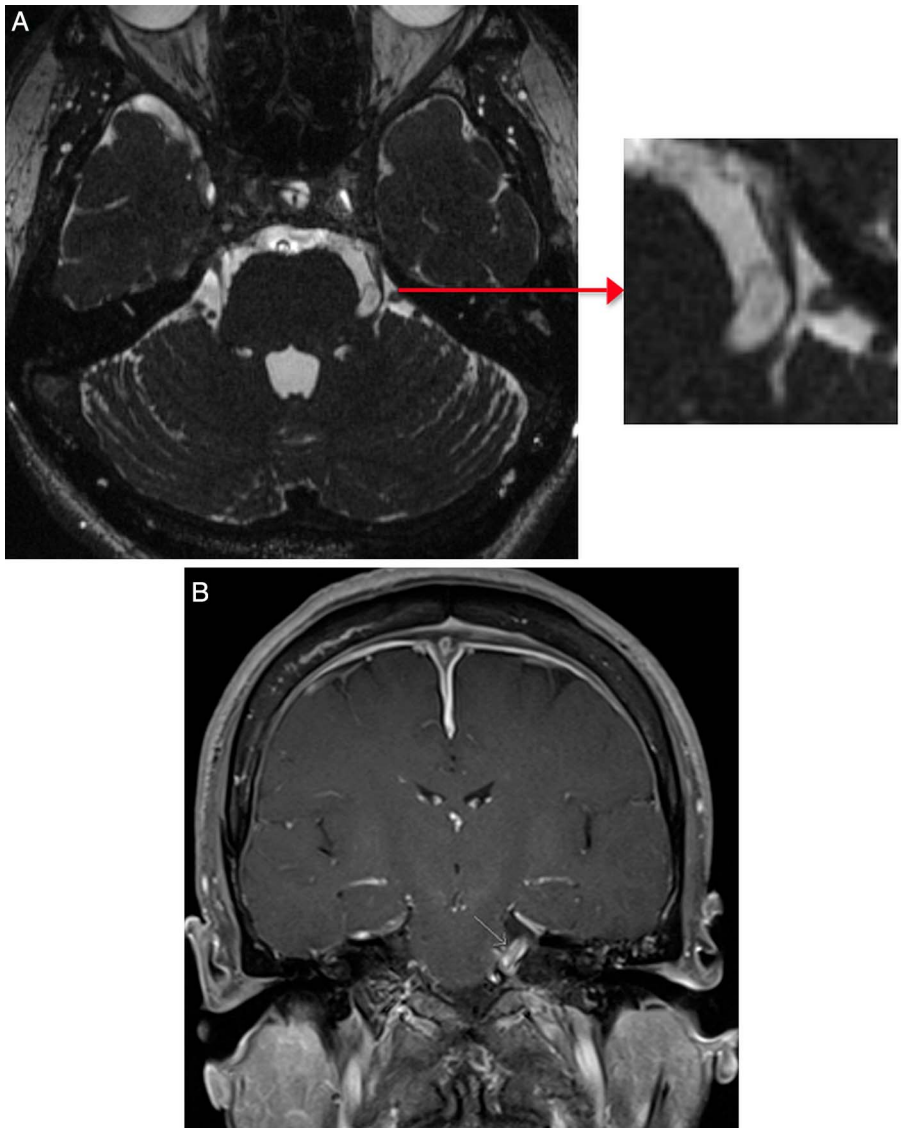

Figure 3 (A) T2-weighted fine slice axial MRI demonstrating a varix compressing and causing lateral bowing of the left trigeminal nerve at its root entry zone (including magnified view), and (B) T1-weighted coronal MRI once again demonstrating the area of conflict at the left trigeminal nerve.

are frequently encountered. Discussed here is a case where the cause is direct compression of the trigeminal nerve by a varix. The varix formed as part of a dilated venous drainage system, a sequelae of SSS thrombosis secondary to TB meningitis. To the best of our knowledge, no other such case has been reported in the literature, however two cases of TN secondary to a TB of the CPA have been described.[3 4] Also, lower back pain and sciatica from spinal nerve root compression by epidural venous enlargement secondary to venous thrombosis, was described by Dudeck *et al*.[5] Although this is in a different location to our case, they both demonstrate low flow systems causing nerve root compression with resultant pain.

There are three clinical entities of central nervous system TB, namely meningitis, intracranial tuberculoma and spinal TB arachnoiditis. These conditions are encountered frequently in regions of the world where TB is endemic.[6] TB meningitis accounts for about 1% of all cases of TB and 6% of all extrapulmonary infections in immunocompetent individuals.[7] One of the uncommon sequelae of TB meningitis is septic dural sinus thrombosis as previously reported in a case by Verma *et al*.[8] In another case, extensive calvarial TB causing compression and occlusion of the SSS was reported in a man presenting with a non-healing scalp sinus.[9]

Given the large size of the SSS, septic thrombotic occlusion is very rare and in fact the most common infection associated with this condition is bacterial meningitis.[10] If thrombosis is acute and complete occlusion of the sinus occurs, the outcome is universally fatal due to massive brain infarction and cerebral oedema. In the case described here, the condition must have occurred as a chronic process as the patient developed compensatory drainage via a dilated venous system.

TN is a condition that often requires multidisciplinary management with the combined efforts of a patient's general practitioner, neurologist, chronic pain specialist and if intractable, a neurosurgeon. It is also important that the patients' mental health is considered given that the condition can leave them incapacitated, and often unable to work. Medical therapy is the first-line treatment and is effective in most cases. The typical agent chosen initially is carbamazepine, which results in complete or near complete pain control in 58–100% of patients.[11]

More invasive measures are considered when the pain is intractable despite maximal medical treatment. These include percutaneous lesional techniques, such as glycerol rhizotomy, radiofrequency thermocoagulation or mechanical balloon compression, MVD and gamma knife radiosurgery in selected cases.

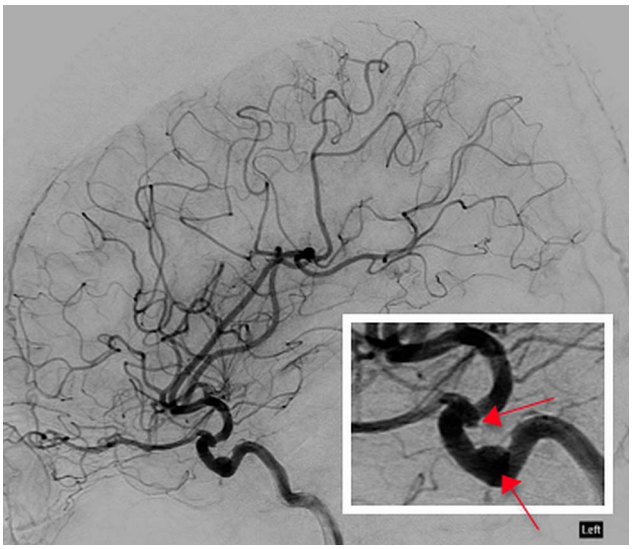

Figure 4 Left internal carotid artery injection cerebral digital subtraction angiogram showing two small incidental aneurysms (magnified view inset).

Learning points

- Trigeminal neuralgia results in physical and psychological impairment and often requires a multidisciplinary approach to manage the patient.
- Low-flow compression of the trigeminal nerve root entry zone by a venous varix, although rarely described, is a potential aetiology of trigeminal neuralgia.
- Invasive measures to manage trigeminal neuralgia, including microvascular decompression, are only considered once the pain is intractable despite maximal medical therapy.
- In cases where a patient is dependent on an anomalous cerebral venous drainage system, there is the added risk of massive venous infarction from surgery should it be necessary to sacrifice the conflicting vein.

These are not without risks however, and careful consideration of the options is required prior to embarking down the path of an invasive approach. In our case, the specific risk of concern is that of massive venous infarction should injury to the conflicting or surrounding vessels occur intraoperatively.

There is good evidence supporting the effectiveness of MVD for the treatment of TN with up to 90% of patients attaining initial pain relief.[11] The pain-free rates do tend to decline with time although up to 73% can still have good relief up to 5 years.[11] Although the reported mortality rate is only of the order of 0.2%, here we have a unique situation with a patient heavily reliant on his anomalous venous drainage pathways.

Percutaneous rhizotomy procedures are less invasive and although initial effectiveness is high, the pain-free rates decline significantly with time and only approximately 50% of patients are pain-free at 5 years.[11] Gamma knife radiosurgery is another option although there is typically a lag time of about 1 month before pain relief is achieved.[12] The main concern however, would be the potential for delayed damage to the varix by the marginal dose of radiotherapy given the intimate relationship with the trigeminal nerve.

Overall, given that MVD is most likely to provide our patient with lasting pain relief, it would be the preferred option should he require an invasive approach. At present, despite occasional exacerbations, the patient's symptoms are controlled by medical means and thus a conservative approach continues.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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