

Role of Surgery in Cerebral Venous Sinus Thrombosis

Syed Ather Enam

Head of Neurosurgery, The Aga Khan University, Karachi.

Abstract

Cerebral venous sinus thrombosis (CVST) usually is taken care of by medical management or neuro-interventional vascular techniques. Some cases of CVST may take a malignant course if the intracranial pressure increases excessively. This increase in pressure is because of oedema in brain tissue due to impediment in venous return and/or intracerebral haemorrhage. Neurosurgical experience has shown remarkable recovery in these moribund patients if appropriate surgical intervention is done within reasonable time. Emergent decompressive craniotomy or other neurosurgical interventions are the only appropriate treatment in these malignant forms of CVST to prevent mortality and severe morbidity.

Introduction

With a worldwide availability of non-invasive imaging studies such as Computerized Tomography (CT) and Magnetic Resonance Imaging (MRI) scans, cerebral venous sinus thrombosis (CVST) has been diagnosed more frequently since the last decade. In the pre-CT era angiography with a venogram was the diagnostic test. This test is still the gold standard but due to convenience and excellent reliability magnetic resonance venogram is now the diagnostic test of choice. CT scan may be negative in a fraction of proven CVST cases and the classical "delta sign" may be seen in only a few. Depending upon the extension of the thrombosis, CT scan and MRI may also show global or focal cerebral oedema, haemorrhagic conversion of infarction, intracerebral haematoma, and signs of herniation. Antithrombotic treatments which include intravenous heparin, endovascular thrombolysis and oral anticoagulants are now well accepted treatments of CVST. In majority of the cases the medical treatment is sufficient but in a few cases the surgical treatment becomes essential to save life and prevent severe neurologic impairment.

Some cases of cvst may take a malignant course

For understanding the role of surgery in CVST, the concept of malignant CVST is being introduced.

The clinical presentation of CVST can be highly varied¹. The presentation may be insidious with headache as the most prominent symptom. Focal neurological deficits and/or seizures is the presenting symptom in about three quarter of CVST cases. An uncommon presentation is a rapidly progressive malignant form where patient quickly deteriorates neurologically ending up in coma. Patients may present with this severe form of CVST to medical attention or alternatively a mild case of CVST may develop into a

malignant form several days later regardless of appropriate medical treatment.² According to a multinational, multi-center study, ISCVT (international Study on Cerebral Vein and Dural Sinus Thrombosis), which prospectively followed 624 patients, 13.9% presented with a Glasgow Coma Score (GCS) between 9-13 and 5.2% were comatose, i.e., had a GCS less than 9 on presentation.³ Close to 9% of patients in that cohort ended up severely impaired or dead (2.2% severely impaired with a modified Rankin Scale 4 or 5 and 6.6% dead due to CVST). It is these malignant cases of CVST that can benefit most from surgical intervention.

Mortality in cvst is usually due to increased intracranial pressure

While most of the CVST cases resolve over time even if they have focal areas of haemorrhages, a transient increase in intracranial pressure (ICP) can put some on a malignant course. The pathogenesis of high ICP in these cases is a combination of oedema due to impaired venous drainage along with mass effect of intracerebral haemorrhage (ICH) that occurs due to venous infarcts and may take form of large clots. A massive swelling ensues throughout the brain which in turn leads to trans-tentorial herniation of the medial temporal lobe with compression of the midbrain or central coning. These events are clinically characterized by deep coma with or without pupillary dilatation. In a post-mortem study on 35 patients who died of CSVT, 32 showed signs of raised ICP such as cerebral swelling, trans-tentorial temporal lobe herniation, and brain stem and occipital lobe haemorrhages.⁴

Coma and intracerebral haemorrhage are poor prognostic factors in cvst

The prognosis of CVST has improved over time.^{3,5} This probably is a reflection of the ease in diagnosis of benign forms of the disease by CT and MRI as well as a wider acceptance of appropriate therapeutic interventions. A significant number of patients, however, still end up with severe morbidity or mortality. Poor prognostic factors identified by the ISCVT study were coma on presentation and haemorrhage on CT scan. Death toll due to CVST in the ISCVT study was 4.3% in the acute phase and another 2.3% during the follow up. An earlier prospective study of 59 patients and a retrospective study of 62 patients with CVST had also suggested coma and intracerebral haemorrhage as predictors of poor outcome.^{6,7} Other factors that were associated with poor outcome were papilloedema, age older than 33 years, and involvement of straight sinus.

Surgical intervention may be the only appropriate treatment in cases of malignant cvst

CVST is primarily treated by medical intervention and by interventional radiology if needed. Most of the neurologists and internists are not very clear about the role of neurosurgery in CVST. The matter of fact is that neurosurgery plays a significant role in the treatment of CVST. There are several neurosurgical procedures that need to be considered in the context of CVST.

Decompressive craniotomy

Decompressive craniotomy is a procedure done in cases of cranial trauma, massive cerebral infarcts, and post-neurosurgical cerebral oedema when increased intracranial pressure (ICP) is a serious problem. This procedure can make a remarkable difference in a malignant case of CVST.

For CVST decompressive craniectomy was described in the 1980's.^{4,8} In one of the largest documented accounts of decompressive craniotomy for CVST, dating back to pre-CT/MRI era, 32 out of 70 CVST patients underwent this procedure.⁴ In this group of patients who had surgery survival was 54% and of those who survived neurologic improvement was observed in 89%. Since the neurological status prior to surgery was not described the value of decompressive craniotomy cannot be judged very well from that study. But in a recent study which is a series of 3 cases that underwent emergent surgery after developing bilaterally dilated and fixed pupil from CVST related cerebral swelling, decompressive craniectomy made a remarkable difference.⁹ Two of these patients returned to their normal daily life, and the third one, in which there was a delay of several hours between pupillary dilatation and surgery, was left with severe impairments but regained consciousness and some ability to communicate. The outcome of the first two cases was phenomenal. In another series of 4 cases in which patients were taken to operating room because of progressive neurological deterioration and CT signs of imminent herniation, the outcome of was very good also.¹⁰ Prior to surgery the GCS of these patients ranged from 6-13 and two of them had deteriorated despite adequate medical management. These patients underwent single or multiple procedures with or without resection of infarcted tissue and evacuation of blood clot. Due to decompressive craniotomy they were not only salvaged, they eventually recovered with no residual deficits or only mild expressive dysphasia.

Since a large cohort can not be established for a valid randomized controlled trial on the role of surgery in CVST, we need to learn lessons from other examples. In posttraumatic cerebral hypertension benefit of decompressive bifrontal craniotomy has been proven statistically.¹¹ The benefit was greatest if the intervention was done before ICP increased to 40 or above for a sustained period of time and if the intervention was done within 48 hours of the injury. In space occupying hemispheric infarction decompressive

craniotomy improved the mortality from more than 75% to less than 35%.¹² Further analysis of their data shows that if only those patients are considered who had dilated their pupils due to increased ICP decompressive craniotomy actually helped reduce mortality from 110% (16/16) to 33% (8/24). This data strongly recommends decompressive craniotomy for malignant CVST.

The decompressive procedure usually consists of removal of a large bone flap along with a dural patch either from artificial dura or autologous tissue such as pericranium, temporalis fascia, or fascia lata. In addition to the removal of bone flap the neurosurgeon may also consider resecting infarcted brain tissue and/or evacuation of haematoma.¹³ The recommended size of dural patch is 15 to 20 cm in length and 2.5 to 3 cm in width.¹² The bone flap may be replaced after 4 to 12 weeks depending upon complete resolution of swelling and medical fitness of the patient. If thrombosis is in the cerebellar veins with consequent oedematous cerebellar infarcts, suboccipital craniotomy is the appropriate procedure.

Open surgical thrombectomy

Open surgical thrombectomy consists of craniotomy over the thrombosed sinus to remove the blood clot and then repair the sinus with or without a patch. Alternatively it may consist of a small keyhole opening over the sinus to pass a catheter to pull out the clot. Technically this procedure may work well for superior sagittal sinus or transverse sinus CVST. Few published reports have documented successful use of this technique.¹⁴⁻¹⁶ Open thrombectomy with local infusion of thrombolytic agents was attempted only when these patients had deteriorated neurologically on systemic anticoagulation. With the current use of endovascular approaches by interventional neuro-radiologists, the advantage of open surgical thrombectomy is very limited.

CSF shunt

In the ISCVT study of 624 patients shunts were performed in 1.6%.³ A need for a ventriculoperitoneal shunt usually arises when venous thrombosis in the posterior fossa causes cerebellar swelling and subsequent fourth ventricle obliteration. The hydrocephalus that ensues may be fatal and needs to be addressed emergently. A temporizing procedure, extra-ventricular drainage of cerebrospinal fluid (CSF) by an external ventriculostomy, may be considered if spontaneous resolution of the blockage in CSF flow is expected once the cerebellar swelling subsides. If hydrocephalus is severe to begin with or recurs upon closing the external ventricular drain, a ventriculoperitoneal shunt or other variations of CSF shunts may have to be inserted. Some patients with CVST develop persistent symptoms of benign intracranial hypertension even after the acute phase of the CVST is over and may need a ventriculoperitoneal or a lumboperitoneal shunt eventually.¹⁷

Venous bypass

The idea of a microvascular anastomosis of a venous channel bypassing the thrombosed sinus is interesting but very challenging. Microsurgical revascularization with a bypass graft between right transverse sinus and superficial jugular vein for a case of bilateral thrombosis of transverse sinuses and internal jugular veins has been described.¹⁸ This resulted in significant improvement. This kind of microvascular feat is hard to accomplish unless the CVST is localized to the transverse and sigmoid sinus. With recent advances in neuro-radiological interventional techniques, the value of complex procedures like microvascular bypass is probably only historical.

Surgical intervention for CVST sequelae

CVST can lead to few other problems that demand a neurosurgeon's attention. Several cases of subdural haematoma secondary to CVST have been reported that required evacuation by burr holes or by craniotomy.¹⁹⁻²¹ Dural arteriovenous fistula that may need surgery has also been documented as a rare long-term complication of lateral sinus thrombosis.⁵

Surgical conditions that may lead to CVST

While malignant form of CVST may need surgical intervention, certain surgical conditions themselves may predispose to CVST. This includes vascular malformation, particularly dural fistula. In the international study ISCVT of 624 cases, arteriovenous malformation and venous anomaly was found in one case each and dural fistula was found in 11 cases (1.6%) as the risk factors for CVST.³ In the same study, cranial trauma was identified as a risk factor for CVST in 1.1%, surgical procedure as a risk factor in 2.7%, and neurosurgical procedures as a risk factor 0.6% of cases. In another study that focused on posterior fossa surgery (suboccipital craniotomy or translabyrinthine craniectomy) 5 out of 117 (4.6%) cases developed subsequent symptomatic transverse sinus thrombosis on the operated side.¹⁷

Medical management after surgical intervention in CVST:

Anticoagulation therapy may be resumed shortly after surgical decompression, albeit at a lower level. Keller et al¹¹ in their series of 4 cases that underwent emergent decompressive craniotomy for malignant CVST describe

resumption of anticoagulation at half dosage 12 hours post-operatively and then at full dosage after 24 hours. They did not see any unwanted effect of early anticoagulation such as post-operative bleeding or enlargement of intracerebral haematoma. Their medical management included observation in the intensive care unit with continuous ICP monitoring and if needed repeat craniotomy to extend decompression or institution of barbiturate coma and hypothermia along with appropriate use of hyperventilation and osmotic therapy.

Conclusion

Current evidence, although all class III, suggests that in malignant CVST even if the patients are in their worst neurological state, neurosurgical intervention should be aggressive. One has to remember however that while surgical intervention will improve survival, some may end up severely handicapped and this should be discussed with the family.

In ISCVT, the largest study so far on CVST (624 patients), decompressive craniotomy was performed on 1.4%.³ In that study 5% presented with coma (GCS < 9), 4.3% expired during the hospitalization and another 2.3% died due to CVST during a median follow-up of 16 months. From the study it appears that the number of CVST patients who had a malignant course was 3-6 times those who had neurosurgical intervention. The ratio should have been opposite. Surgical attempts to prevent mortality should be more than the number of deaths.

With the advancement in medical management the benefits of surgical intervention has been ignored. Emphasis on the medical management and endovascular treatment for CVST should not overlook the value of surgical intervention. Decompressive craniotomy may produce remarkable recovery in patients with malignant CVST. Some of the factors that impose a poor prognosis to the CVST should be the indications for neurosurgical intervention. These include coma, anisocoria, CT / MRI showing intracerebral haemorrhage, signs of imminent herniation and severe cerebral oedema. When patients develop signs of herniation, emergent decompressive craniotomy may be the only appropriate treatment. The case series of Stefini et al⁹ and Keller et al¹¹ clearly show that if decompressive craniotomy is done in a timely fashion, those patients who are at the brink of death may have excellent recovery and may return to their normal life.

References

1. Allroggen H, Abbott RJ. Cerebral venous sinus thrombosis. *Postgrad Med J*. 2000;76:12-5.
2. Weber J, Spring A. Unilateral decompressive craniectomy in left transverse and sigmoid sinus thrombosis. *Zentralbl Neurochir*. 2004;65:135-40.
3. Ferro JM, Canhao P, Stam J, Bousser MG, Barinagarrementeria F; ISCVT Investigators. Prognosis of cerebral vein and dural sinus thrombosis: results of the International Study on Cerebral Vein and Dural Sinus Thrombosis (ISCVT). *Stroke*. 2004;35:664-70.
4. Nagpal RD. Dural sinus and cerebral venous thrombosis. *Neurosurg Rev*. 1983;6:155-60.
5. Preter M, Tzourio C, Ameri A, Bousser M. Long-term Prognosis in Cerebral Venous Thrombosis: Follow-up of 77 Patients. *Stroke*. 1996;27:243-46.
6. Bienfait HP, Stam J, Lensing AW, van Hilten JJ. Thrombosis of the cerebral veins and sinuses in 62 patients. *Ned Tijdschr Geneesk*. 1995; 139:1286-91.
7. de Bruijn SF, de Haan RJ, Stam J. Clinical features and prognostic factors of cerebral venous sinus thrombosis in a prospective series of 59 patients. For The Cerebral Venous Sinus Thrombosis Study Group. *J Neurol Neurosurg Psychiatry*. 2001;70:115-8
8. Ohya M, Sato O. A case of cerebral venous and superior sagittal sinus thrombosis, treated with decompressive craniectomy (author's transl). *No Shinkei Geka*. 1980;8:803-11.
9. Stefini R, Latronico N, Cornali C, Rasulo F, Bollati A. Emergent decompressive craniectomy in patients with fixed dilated pupils due to cerebral venous and dural sinus thrombosis: report of three cases. *Neurosurgery*. 1999;45:626-9.
11. Keller E, Pangalu A, Fandino J, Konu D, Yonekawa Y. Decompressive craniectomy in severe cerebral venous and dural sinus thrombosis. *Acta Neurochir Suppl*. 2005;94:177-83.
11. Polin RS, Shaffrey ME, Bogaev CA, Tisdale N, Germanson T, Bocchicchio B, Jane JA. Decompressive bifrontal craniectomy in the treatment of severe refractory posttraumatic cerebral edema. *Neurosurgery*. 1997;41:84-92.
12. Rieke K, Schwab S, Krieger D, von Kummer R, Aschoff A, Schuchardt V, Hacke W. Decompressive surgery in space-occupying hemispheric infarction: results of an open, prospective trial. *Crit Care Med*. 1995;23:1576-87.
13. Nguyen JP, Lepresle E, Homs JB, Djindjian M, Goujon C. The role of surgery and low molecular weight heparin in the treatment of cerebral venous thrombosis. *Apropos of 2 cases Agressologie*. 1989;30:351-2.
14. Persson L, Lilja A. Extensive dural sinus thrombosis treated by surgical removal and local streptokinase infusion. *Neurosurgery*. 1990;26:117-21.
15. Kourtopoulos H, Christie M, Rath B. Open thrombectomy combined with thrombolysis in massive intracranial sinus thrombosis. *Acta Neurochir (Wien)*. 1994;128:171-3
16. Ekseth K, Boström S, Vegfors M. Reversibility of severe sagittal sinus thrombosis with open surgical thrombectomy combined with local infusion of tissue plasminogen activator: technical case report. *Neurosurgery*. 1998;43:960-5.
17. Keiper GL Jr, Sherman JD, Tomsick TA, Tew JM Jr. Dural sinus thrombosis and pseudotumor cerebri: unexpected complications of suboccipital craniotomy and translabyrinthine craniectomy. *J Neurosurg*. 1999;91:192-7.
18. Sindou M, Mercier P, Bokor J, Brunon J. Bilateral thrombosis of the transverse sinuses: microsurgical revascularization with venous bypass. *Surg Neurol*. 1980;13:215-20.
19. Marquardt G, Weidauer S, Lanfermann H, Seifert V. Cerebral venous sinus thrombosis manifesting as bilateral subdural effusion. *Acta Neurol Scand*. 2004;119:425-8.
20. Matsuda M, Matsuda I, Sato M, Handa J. Superior sagittal sinus thrombosis followed by subdural hematoma. *Surg Neurol*. 1982;18:206-11.
21. Takamura Y, Morimoto S, Uede T, Yamaki T, Minamida Y, Yamamura A, Nakagawa T. Cerebral venous sinus thrombosis associated with systemic multiple hemangiomas manifesting as chronic subdural hematoma--case report. *Neurol Med Chir (Tokyo)*. 1996;36:650-3.