

# Cerebral Venous Sinus Thrombosis (CVST): A Review of the Deadly Threat



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## Abstract

Although, the frequency of cerebral venous sinus thrombosis (CVST) has dropped remarkably in recent years but unfortunately, it is still seen commonly in a developing country like Pakistan. The diagnosis of CVST needs high threshold for suspicion, especially in patients with meningitis or sinusitis who present with focal neurological deficit. Early recognition and treatment improves mortality and morbidity rates of this potentially deadly disease. Intravenous, wide spectrum, antibiotics plus anticoagulation with low molecular weight heparin and/or early surgical intervention of the primary site of infection whenever possible are crucial.

**Keywords:** Cerebral venous sinus thrombosis (CVST); Iron-deficiency anemia (IDA) Cerebral; Tuberculous meningitis (TBM); Dural; Venous; Thrombosis; Thrombophilia; Meningitis

## Introduction

CVST is the presence of blood clot in the dural vein. This happens when a thrombus forms in the brain's venous sinus that stops blood from flowing out of the brain [1]. Such patients present with a variety of symptoms [1,2]. The most severe and prognostically poor form of presentation of CVST is the venous hemorrhagic infarction (VHI) [2]. However, CVST is a rarer cause of cerebral infarction compared to arterial diseases [1].

CVST can result from a variety of congenital or acquired diseases. These may include medical conditions such as, dehydration, infection, malignancy or hematologic disorder and so forth [3]. It is interesting to note that iron-deficiency anemia (IDA) is one of common etiologies behind pediatric CVST but not of adult CVST [4]. Being a developing country, infections are a common cause of hospitalization to medical and pediatric wards in Pakistan.

Amongst the most dreadful infectious diseases, pyogenic and tuberculous meningitides are of particular concern. Such infections are dangerous on their own; however, their complications are more devastating.

Once such complication of meningitis and septic head and neck infections is CVST. This mini review will focus on the

etiology, pathogenesis and common presentation of CVST to a tertiary care hospital in Pakistan.

## Discussion

### Pathogenesis

There are no valves in the dural venous sinuses and the cerebral and emissary veins. Therefore, they permit blood to flow in either direction determined by the pressure gradients in the vascular system [3-6]. This pressure gradient dependent circulation makes them vulnerable to septic thrombosis resulting from the spread of infection from adjacent locations. Although, septic thrombophlebitis can affect any part of the cerebral venous system, but certain sinuses are more commonly thrombosed than others such as, the cavernous sinus followed by the lateral sinuses [5-9].

Dural sinus thrombosis results from a spread of infection from the surrounding area. Bacterial meningitis can spread to involve and thrombose any part of the cerebral venous system, but it commonly involves the sagittal sinus or the transverse and sigmoid sinuses. Similarly, in sphenoid and/or ethmoid sinusitis, infection spreads to the adjacent cavernous sinus resulting in cavernous sinus thrombophlebitis. 7-11 Moreover, the mastoid

sinusitis results in lateral sinus thrombophlebitis first than affecting other parts. Nevertheless, infections at other sites such as, the face, nose, tonsils, soft palate, teeth and ears may lead to thrombophlebitis in the cavernous and lateral sinuses as well as other sinuses; however, orbital infection is rarely complicated by CVST [9-13].

Infection may lead to CVST directly or indirectly by precipitating thrombosis in people who suffer from a prothrombotic illness. Such people may include those with thrombophilia or acquired causes of pro-coagulant status like chronic infection, malignancy and so forth. Hence, once study concluded that, CVST may be multi-factorial in nature [10-15].

### Etiology

Amongst the different causes of CVST, the following are of prime importance and need special attention. These include infectious agents like, pneumococcal meningitis. Other causes of meningitis such as Coccidioidomycosis, cytomegalovirus and herpes simplex as well as measles have been implicated [12-16]. However, in our part of the world, it is most commonly seen in patients with Tuberculous meningitis (TBM) followed by pyogenic meningitis and is very rare in patients with viral encephalitis [15-16]. Similarly, there have been some reports linking increased risk of CVT with HIV-AIDS. It appears that HIV infection does not play a significant role directly itself, rather it seems that, a combined opportunistic infection and/or coagulopathy and HIV infection are required to develop this serious complication [12-17].

In our experience, apart from infections, other notable causes of CVST in Pakistan include; medical illnesses like solid organ malignancy like prostatic carcinoma, hematological tumor like multiple myeloma, leukemia and lymphoma, chronic infections at sites other than the CNS such as pulmonary tuberculosis. Similarly, cerebro-vascular accidents, bed ridden patients, paraplegic or quadriplegic patients secondary to neuromuscular disorder or spinal cord disease and post-partum ladies commonly encounter CVST. Moreover, it is occasionally seen in patients who are diabetic or hypothyroid or on medications like corticosteroid or oral contraceptive pills and so forth.

### Presentation

Patients with septic sinus thrombosis are much more critically ill than those otherwise [13-18]. The illness is almost always acute in nature and patients are very sick, toxic and pyrexial. They often may have focal neurologic symptoms and signs. Moreover, they also have symptoms and signs of high intracranial pressure [15-17]. The focal symptoms and signs may vary depending on which site of the cerebral venous system is involved. Similarly, they may present with seizures, cranial nerve palsies, breathing or feeding problems, urinary retention or incontinence and altered mentation [16-18].

### Diagnosis

High level of suspicion is essential for early identification and management. The initial work up should include complete blood count, blood cultures, x-ray films of the paranasal sinuses and enhanced brain MRI plus MRV and/or Head CT scanning. Cerebrospinal fluid (CSF) analysis as well as culture is crucial in the diagnosis of the cause of CVT [17-18]. The CSF analysis in non-septic sinus thrombosis is usually normal, but occasionally, it may be bloody or xanthochromic as the result of cortical and meningeal hemorrhage. Nevertheless, in septic thrombosis, it is often abnormal and has high granulocyte and/or lymphocyte count depending on the underlying etiology such bacterial versus mycobacterial and elevated protein. The CSF culture may be positive in some of the cases. Other work may be cause directed and may include a malignancy screen, a systemic infection screen and/or thrombophilia screen and so forth [15-19].

### Management

The management includes general and specific measures and largely depends upon the etiology of CVST. Generally speaking, every patient with suspected or confirmed CVST should have his/her airways, breathing and circulation attended to [18-20]. Moreover, they should be started on sufficient amount of fluids, oxygen where needed and nutritional support. Every effort should be made to keep the metabolic and electrolyte balances in place. The caring team must ideally involve a physician, a neurologist, a neurosurgeon, a microbiologist, a hematologist and ancillary staff [13-16].

Specific measures include the early management with broad spectrum intravenous antibiotics and early surgical drainage of the primary site of infection such as that of mastoid sinus in case of mastoiditis and so forth. Similarly, most authorities recommend anticoagulation with low molecular weight heparin. However, intravenous heparin infusion and corticosteroids are of uncertain benefit, although some reports have shown favorable response rates. The need for a long term anticoagulation with an oral agent such as warfarin or dabigatran or rivaroxaban should be decided on a case to case basis as such decision largely depends on the etiologies of CVST and on the presence or absence of concomitant co-morbidities [11,15,18,19].

### Conclusion

CVST must be considered in the differential diagnoses of complicated meningitides and head and neck infections. MRI along with MRV is good enough to give a diagnosis. Treatment depends upon the cause and usually involves broad spectrum intravenous antibiotics and anticoagulation with low molecular weight heparin in the short term.

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