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CASE REPORT

Fatal cerebral venous sinus thrombosis after lumbar puncture: a case report and review of the literature

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Abstract - Cerebral venous sinus thrombosis (CVST) is a rare disorder with multiple known risk factors. Sometimes a rare and unusual cause is found. We describe a patient in whom fatal CVST occurred following a lumbar puncture with post lumbar puncture headache. A nineteen-year-old man was treated with high dose corticosteroids (methylprednisolone) and underwent a lumbar puncture after demyelating lesions were seen on cerebral magnetic resonance imaging. He developed post lumbar puncture headache and after four days generalised seizures. A computed tomography scan showed CVST with diffuse swelling of the brain and no contrast enhancement of the straight sinus, superior sagittal sinus and transverse sinus. Despite thrombosuction and placement of a stent, bilateral decompression craniotomy, deep sedation and hypertonic saline the patient died two days after the diagnosis of CVST. The cause of the CVST was probably due to cerebrospinal fluid leakage after lumbar puncture in combination with high dose corticosteroids (methylprednisolone). Intracranial hypotension after a lumbar puncture may cause intracranial veins to dilate, causing reduced blood flow, stasis and subsequent CVST. High dose intravenous corticosteroids may cause a hypercoagulable state and therefore might contribute to CVST. Up to date information of the different treatment options of CVST is given. We would like to emphasize that CVST could occur after a lumbar puncture in combination with high dose corticosteroids.

Keywords - **lumbar puncture, cerebral venous sinus thrombosis, treatment**

Introduction

Cerebral venous sinus thrombosis (CVST) is a rare disorder with an incidence of 3-4 cases per million population affecting more females than males. Multiple causes and risk factors are known. In 70-80 percent of cases a cause can be found with the most frequent ones being medication (oral contraceptives, prednisolone), postpartum state, infection, malignancy or coagulopathy. Sometimes a rare and unusual cause is found. We describe a patient in whom fatal cerebral venous thrombosis occurred following a lumbar puncture with post lumbar puncture headache.

Case Report

A nineteen-year-old man with a history of traumatic skull base fracture fourteen years previously developed visual loss of the right eye, with slow pupillary response to light. No papilloedema was seen and magnetic resonance imaging (MRI) was done because of suspected optic neuritis. MRI showed multiple infra and supra-tentorial white matter lesions (also juxtacortical and corpus callosum) suggestive of demyelination (but no signs of cerebral venous sinus thrombosis). Blood examination showed no abnormalities, with normal blood cell count, thrombocytes, electrolytes and normal liver and kidney functions. Lyme disease (borrelia IgG/IgM) and lues (VDRL/TPPA) were negative and ruled out. Based on a suspicion of multiple sclerosis, the patient underwent a lumbar puncture which showed slight pleiocytosis (23/3 leucocytes), normal glucose (5.0 mmol/L) and protein count (0.44 G/L). The IgG index was elevated (0.93) and oligoclonal bands were found. High dose methylprednisolone was given (1000 mg a day for three days). He developed post lumbar puncture headache, which was treated with bed rest. After four days the patient developed tonic - clonic generalised seizures and somnolence. Computed tomography showed swelling of the brain and a dense clot sign of the straight sinus (Figure 1). However, at first, the dense clot sign was mistaken for a subarachnoid haemorrhage and he was transferred to the intensive care unit of our hospital. Soon after this, further impairment of consciousness occurred (Glascow coma scale E1M1V1). A repeated computed tomography scan showed diffuse swelling of the brain and haemorrhage of the left frontal and right parietal and temporal lobes. There was no contrast enhancement of the straight sinus, superior sagittal sinus and transverse sinus (Figure 2). A quickly performed digital subtraction angiography (DSA) confirmed the diagnosis of CVST. During DSA procedure, local medication with thrombolytic drugs (urokinase), trombosuction and placement of a stent was performed, but no obvious recanalization was obtained. Treatment with anticoagulant drugs (heparin) was initiated. Further severe swelling of the brain was seen on repeated imaging. Placement of an intracranial pressure device and bilateral decompression craniotomy was performed to facilitate the swelling of the brain. However, persistent high intracranial pressure was seen, despite treatment with severe sedation and hypertonic saline. A computed tomography scan

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showed increasing brain herniation (Figure 3) and the patient died two days after the diagnosis CVST had been made.

Discussion

We describe a young patient with fatal massive cerebral venous sinus thrombosis. We hypothesized that the cause of the thrombosis might have been cerebrospinal fluid leakage following lumbar puncture in combination with prior intravenous high dose corticosteroids (methylprednisolone).

CVST may result in raised intracranial pressure with headache, nausea and vomiting. Eventually papilloedema, focal deficits, seizures and impairment of consciousness occurs, but not all patients become comatose. CVST may occur without severe symptoms in some patients. CVST may be due to several different causes, such as prothrombotic states, pregnancy or puerperium. Other causes are hereditary (factor V Leiden, protein S deficiency or Von Willebrand disease) or secondary coagulopathy. Secondary coagulopathies due to systemic or local infection, systemic disease or drugs (oral contraceptives, prednisolone) is the major cause of CVST. Less frequent causes are trauma or lumbar puncture. However, in 20-30% of cases no cause is found [1]. CVST in multiple sclerosis patients has been described earlier [2-5] and high doses of intravenous methylprednisolone might cause a hypercoagulable state. In Cushing syndrome the hypercoagulable state is due to an increase in plasma levels of clotting factors such as factor VIII-complex. A high level of fast acting plasminogen activator inhibitor and decreased levels of tissue type plasminogen activator might give impaired fibrinolytic capacity [6]. The sequence of lumbar puncture followed by high dose intravenous corticosteroids has been considered a possible cause in patients with multiple sclerosis who developed CVST.

Lumbar puncture itself could cause CVST, especially when post lumbar puncture headache has occurred. Post lumbar puncture headache is a typical headache which arises when the patient rises and disappears when supine. This is due to leakage of CSF through a dural rent, resulting in low CSF pressure (CSF hypotension) in the subarachnoid space [7]. In a study done by Canhao [8], a lumbar puncture resulted in a decrease of 47% of the mean blood flow velocity in the straight sinus. The decrease of blood flow sustained for more than six hours after the lumbar puncture and this mechanism might induce thrombosis. Otherwise intracranial hypotension after a lumbar puncture might cause intracranial veins to dilate, leading to reduced blood flow, stasis and subsequent thrombosis. A substantial number of patients described in the literature with a cerebral venous thrombosis after a lumbar puncture had a prothrombotic state [9-13]. One might hypothesize that a prothrombotic state could contribute to a cerebral venous thrombosis after a lumbar puncture.

In the literature between 1985 and 2010, only a small number of cases of spinal dural injury followed by CVST are described. Aidi [6] and Miglis [14] found a total of 29 cases of CVST after lumbar puncture. Lumbar puncture was performed for various reasons, like spinal anaesthesia, myelography, intrathecal administration of drugs and after lumbar puncture for diagnosis. In 22 of the 29 case reports other risk factors for CVST were present like congenital thrombophilia, postpartum state, haematological malignancy, oral contraceptives and high dose corticosteroid use. In our patient there was no infection or prothrombotic state, nor was a coagulopathy present. However, the administration of intravenous methylprednisolone in combination with post lumbar puncture headache one week prior to the CVST might have been the cause of CVST.

A complication from a lumbar puncture is rare. Complications included post lumbar puncture headache, meningitis, nerve root involvement, brain stem herniation, subdural hematoma and CVST. Hence CVST must always be suspected when post lumbar

Figure 1. Thrombus in the straight sinus called dense clot sign (green arrow), and diffuse swelling of the brain.

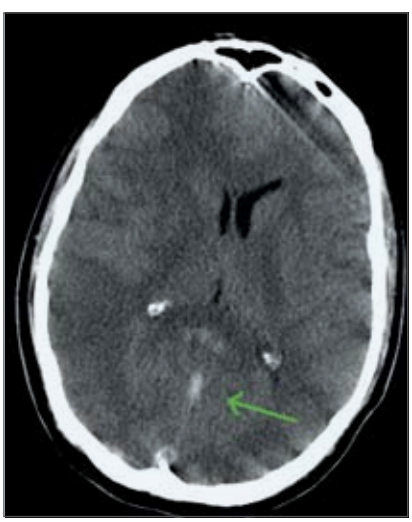
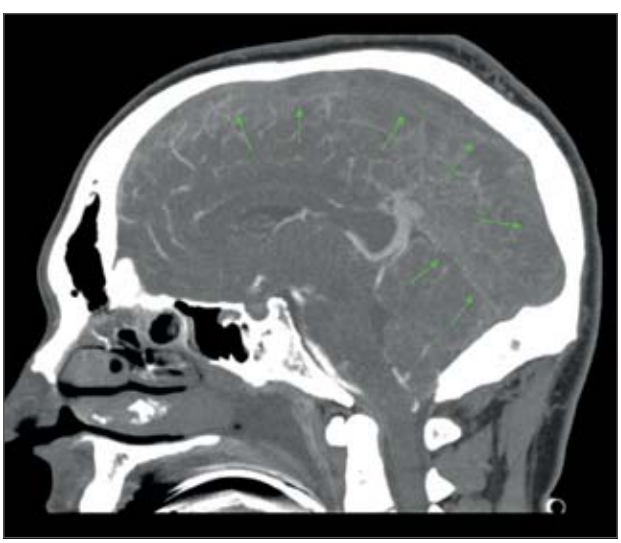


Figure 2. Computed tomography angiography with no contrast enhancement of the straight sinus and superior sagital sinus (green arrows).

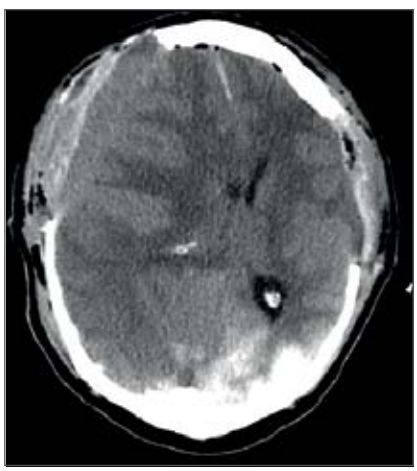


puncture headache evolves into a continuous severe headache [6].

Normally a post lumbar puncture headache disappears within a few days after bed rest. When bed rest doesn't improve the headache, one could consider a blood patch to solve the CSF leakage, however, it is very doubtful if a blood patch in our case would have prevented CVST.

Anticoagulant drugs are a generally accepted treatment for CVST [15]. The decision to perform additional invasive treatments remains up to the individual judgement of the treating physician, since evidence from randomized controlled trails is lacking for those treatments. We knew that despite anticoagulants moderate to severe disability occurred in 5% and death in 8% of the patients with sinus thrombosis. In those cases other treatment options could be considered. In our patient with massive thrombosis we chose to try mechanical and chemical thrombolysis, eventually followed by decompressive surgery. Transtentorial herniation by mass effect and midline shift is the most frequent cause of death in CVST and emergency decompressive surgery has been suggested as a life-saving procedure, however, data from randomized controlled trials are not available. Theaudin [16] and Coutinho [17] report that patients with already impaired consciousness or coma and/or dilated pupils could still have a good outcome. Besides removing the threat of fatal herniation and the relief of mass effect, decompressive surgery may improve cortical collateral vein drainage and thereby heparin therapy.

Figure 3. Status after bilateral decompressive craniotomy with brain herniation and severe midline shift to the left side.



There are no randomized controlled trials on the use of thrombolytics, although they appear to be safe (intracranial haemorrhage in 17% and in 5% clinical deterioration), but their efficacy cannot be assessed from the published literature [18]. The use of locally infused thrombolytics gives favourable outcomes with individualized procedures [19], but less successful cases have also been reported [20]. Mechanical thrombectomy is a promising therapy modality but randomized controlled trials for this intervention are also lacking. The available literature tells us that its use alone as well along with fibrinolytics gives various results [21].

In conclusion, we present a young man in whom CVST occurred following a lumbar puncture with post lumbar puncture headache and high dose intravenous methylprednisolone. Despite aggressive treatment of CVST with thrombolytic drugs, thrombosuction, placement of a stent and bilateral craniotomy, the patient died. We would like to emphasize that CVST could occur after a lumbar puncture in combination with high dose corticosteroids. We would also like to emphasize that a post lumbar puncture headache that changes into a constant headache is a warning sign and should prompt physicians to consider CVST.

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