CME Article

Clinics in diagnostic imaging (103)

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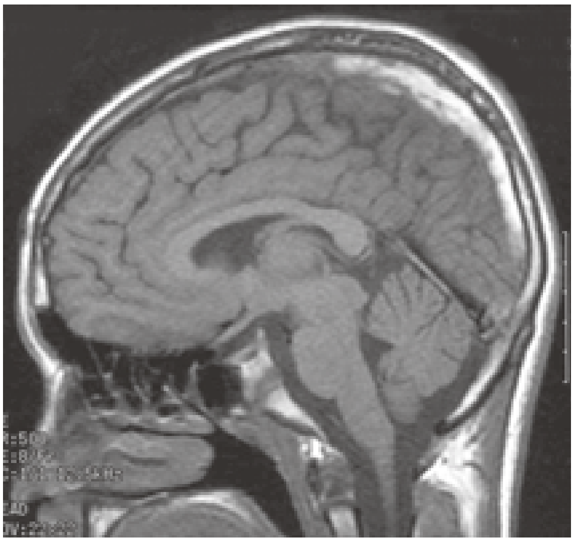

Fig. 1a Unenhanced sagittal spin-echo T1-W MR image.

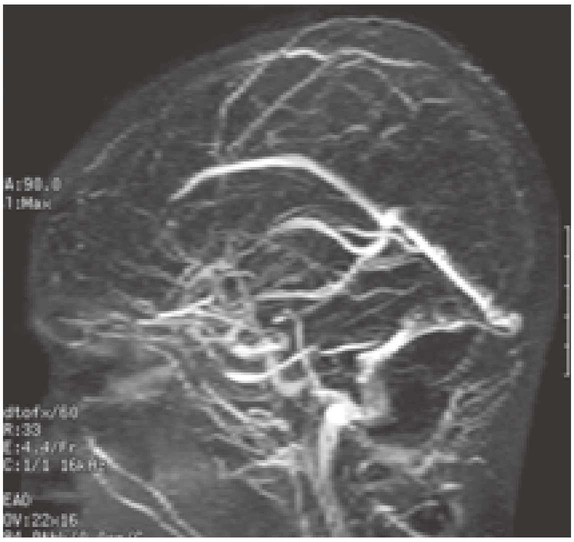

Fig. 1b Sagittal 2D time-of-flight (TOF) MR venography image.

CASE PRESENTATION

A 13-year-old boy was referred from another hospital with a history of severe headache for two weeks. He had also vomited many times. While he was being admitted at the other hospital, he had a seizure. Neurological examination was normal except for bilateral papilloedema. What do the magnetic resonance (MR) imaging scan of his brain (Fig. 1a) and MR venography (Fig. 1b) show? What is the diagnosis?

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Fig. 2 Coronal 2D TOF MR venography image shows no flow signal in superior sagittal, right transverse, and right sigmoid sinuses. Flow signal is seen in medial portion of left transverse sinus (arrow) and in left sigmoid sinus (S).

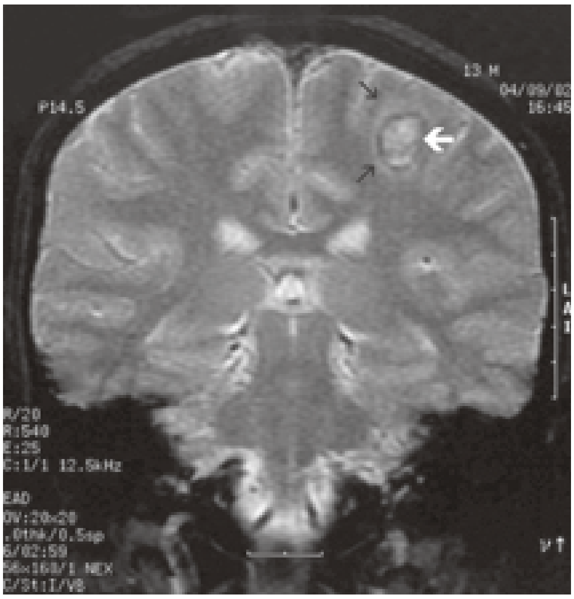

Fig. 3 Coronal gradient-echo MR image shows a small subacute haematoma in left parietal lobe as oval shaped hyperintense lesion surrounded by haemosiderin rim (white arrow). Note minimal brain oedema (dark arrows).

IMAGE INTERPRETATION

The sagittal T1-weighted MR image (Fig. 1a) shows replacement of the normal flow void in the superior sagittal sinus by a hyperintense subacute thrombosis. MR venography (Fig. 1b) confirms that there is no flow signal in the superior sagittal sinus. There is also thrombosis of the right transverse, right sigmoid, and left transverse sinuses (Fig. 2). Haemorrhagic infarction is seen in the right frontal and left parietal regions (Fig. 3).

DIAGNOSIS

Dural sinus thrombosis with cerebral venous infarction.

CLINICAL COURSE

The patient had many blood tests to identify the cause of sinus thrombosis, including protein C, protein S, and antithrombin levels, but all the results were negative. He was treated with anticoagulants. The patient underwent a follow-up MR venography seven months later, which showed restoration of flow signal in the left transverse sinus and some flow signal in the superior sagittal sinus (Fig. 4), but there was still no flow in the right transverse or right sigmoid sinuses (Fig. 5).

DISCUSSION

Cerebral venous thrombosis in childhood is a serious disease, the incidence of which is increasing, mainly because of more sensitive diagnostic methods and increasing awareness of this entity[1]. Cerebral venous thrombosis usually involves the cerebral venous sinuses, such as the sagittal and transverse sinuses, and may extend to the cerebral veins[2]. The signs and symptoms are extremely varied and nonspecific, ranging in severity from mild headache to progressive neurological deficit related to intracranial haemorrhagic infarction.

Neuroimaging techniques, including computed tomography (CT), MR imaging, MR venography, and CT venography, are crucial for the diagnosis of cerebral venous thrombosis[2-5]. This article briefly reviews the causes, signs, symptoms, the imaging findings, treatment, and outcomes of cerebral venous thrombosis in children. Because cerebral catheter angiography is rarely used for diagnosis of cerebral venous thrombosis[3] and Doppler ultrasonography is only done in infants[6], neither of these will be discussed here.

Causes, clinical signs and symptoms

Cerebral venous thrombosis in children is a multifactorial disease that results in the majority of cases from a combination of prothrombotic risk factors and/or an underlying disorder. Local or systemic infections, vascular trauma, cancer, acute lymphoblastic leukaemia, drug toxicity, lupus erythematosus, nephrotic syndrome, dehydration, asphyxia, maternal problems during pregnancy, Behcet's disease, and metabolic disorders have been reported as predisposing factors[1]. Cerebral venous thrombosis caused by head and neck infections, particularly common in preschool children, is related to otitis media and mastoiditis[7].

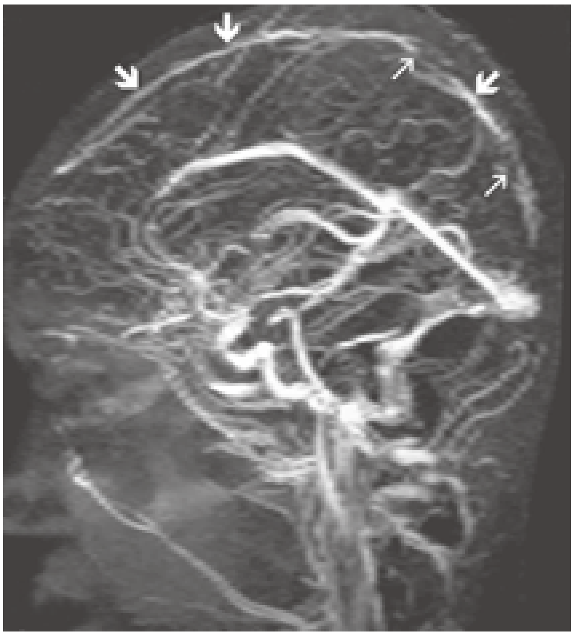

Fig. 4 Follow-up sagittal 2D TOF MR venography image shows some flow signal in superior sagittal sinus (large arrows). Note irregular filling defects because of residual thrombi (small arrows) in sinus.

Fig. 5 Follow-up coronal 2D TOF MR venography image shows no flow signal in right transverse or right sigmoid sinuses. Flow signal in left transverse sinus (large arrow) and some flow signal in superior sagittal sinus (arrowhead) are seen. Note multiple collateral veins (small arrows) near superior sagittal and right transverse sinuses.

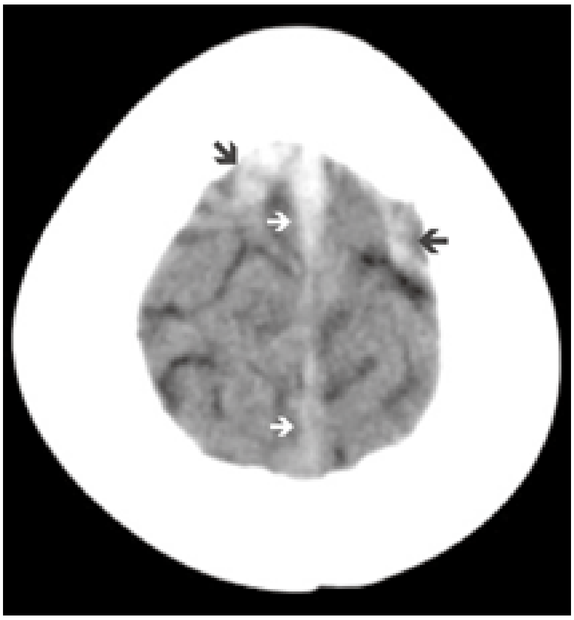

Fig. 6 Cord sign. Unenhanced axial CT image of the brain shows linear areas of high density (black arrows). Note hyperdense thrombus in superior sagittal sinus (white arrows).

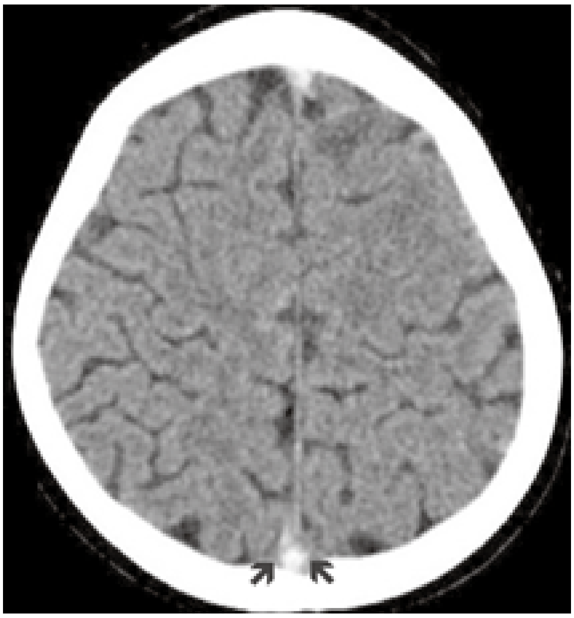

Fig. 7 Dense triangle sign. Unenhanced axial CT image of the brain shows a hyperdense thrombus in posterior part of superior sagittal sinus (arrows).

Signs and symptoms of cerebral venous thrombosis in children are age-dependent, ranging from minimal and nonspecific ones such as decreased oral intake and irritability to more ominous states such as lethargy and coma. Seizures, fever, lethargy or irritability and respiratory distress are common signs of cerebral venous thrombosis in neonates. Older children commonly present with fever and lethargy, often associated with signs of intracranial hypertension such as vomiting, headache, papilloedema, and abducens nerve palsy[2].

Imaging findings

Imaging findings of cerebral venous thrombosis can be categorised as direct, when there is visualisation of the cortical or dural sinus thrombus, or indirect, when there are ischaemic cerebral changes related to venous outflow disturbances[3].

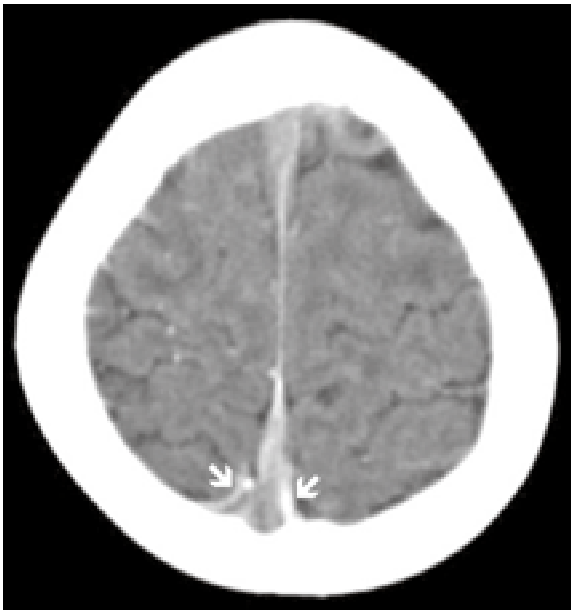

Fig. 8 Empty delta sign. Enhanced axial CT image of the brain shows triangular filling defect in the posterior part of superior sagittal sinus (arrows).

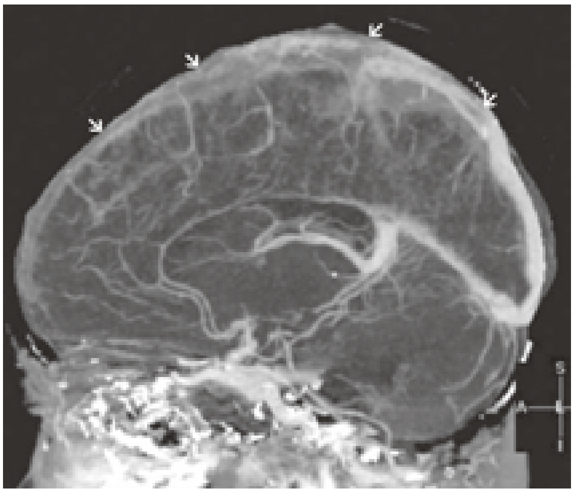

Fig. 9 Sagittal reconstructed CT venography image shows irregular filling defects in superior sagittal sinus (arrows).

Cranial CT

In the first two weeks, thrombotic blood is usually hyperdense on unenhanced CT compared to brain parenchyma. If there is a thrombus in a cerebral vein, it is seen as a linear area of high density, called the "cord" sign[2-4,8] (Fig. 6). If there is a hyperdense thrombus in the posterior aspect of the superior sagittal sinus, it is called the "dense triangle" sign[2,3] (Fig. 7). After two weeks, a thrombus will become isodense to brain parenchyma, and therefore will only be visible on enhanced CT as a hypodense area or a filling defect in the venous sinus surrounded by enhanced dura. If the thrombus is located in the posterior part of the superior sagittal sinus, a triangular filling defect (the "empty delta" sign)[2-4,8] (Fig. 8) is sometimes seen on enhanced CT. CT findings may be false positive, particularly in neonates, as the higher haematocrit and predominantly unmyelinated brain in this age group produce a hyperdense posterior falx/torcula, mimicking sinus thrombosis[9].

Venous infarction may be evident on CT as a global or focal lesion of hypodensity. Infarction occurs when cerebral venous thrombosis results in increased intracranial pressure and increased retrograde venous pressure. Decreased cerebral blood flow leads to decreased cerebral perfusion pressure, and finally to venous infarction which is often accompanied by haemorrhage[3]. These hypodense lesions do not conform to the pattern of an arterial distribution, and mass effect is common[3,4]. In superficial sinus thrombosis, infarctions are seen in the cortex and subcortical white matter. Associated haemorrhage is also more commonly found in subcortical locations[10]. Bilateral parasagittal hypodense lesions on CT are a common feature of venous thrombosis of the superior sagittal sinus. Bilateral thalamic hypodense lesions may be seen in deep sinus thrombosis, and on unenhanced CT, the thrombus may be seen in the straight sinus[3].

MR imaging and MR venography

MR imaging combined with MR venography is usually the modality of choice for diagnosis of venous sinus thrombosis and also for the follow-up[11]. Conventional T1- and T2-weighted MR images can directly show the thrombus in the dural sinuses, as well as the secondary parenchymal changes that result from venous outflow obstruction. The signal intensity of the venous thrombus depends on the degree of residual flow and its age. In the early or acute stages of venous thrombosis (the first 3 to 5 days), blood is in the deoxyhaemoglobin state, so the occluded vein will appear isointense on T1-weighted images and hypointense on T2-weighted images. Thus, hypointensity of T2-weighted images can be misinterpreted as the normal flow void, and patients with suspected occlusion must be carefully evaluated[4,12].

In the subacute phase, a thrombus is hyperintense on T1- and T2-weighted MR images because deoxyhaemoglobin has been converted to methaemoglobin. This signal change is seen five to 30 days after the start of thrombosis. Later changes are generally related to the degree of flow and recanalisation. Flow void may return with recanalisation, although a high percentage of patients continue to have abnormal signals in the dural sinus for months or years after thrombosis[12]. Chronically-thrombosed sinuses undergo fibrosis, and extensive venous collaterals may develop[4] (Fig. 5). Since the signal intensity of the thrombus varies over time, and because signals from slow flowing blood may confuse interpretation, MR imaging should be done together with MR venography[12].

On MR venography, a thrombosed dural sinus does not show the normal flow signal of the sinus nor an irregular filling defect of the sinus, as in this patient. Sometimes, arachnoid granulation can cause a filling defect of the sinus, but it is regular[13]. A variety of parenchymal changes are seen on MR imaging of venous thrombosis. These vary greatly, depending on whether venous sinuses, superficial cortical veins, or the deep venous system are thrombosed. Parenchymal changes can include oedematous and haemorrhagic lesions[12].

CT venography

CT venography is a technique that aids in the diagnosis of venous thrombosis. It is similar to CT angiography except that optimal venous imaging requires a longer delay after the administration of contrast medium than for CT angiography. With this delay, there is enough time for the contrast medium to reach and opacify the veins. After venous images have been obtained, the bony details are subtracted and three-dimensional (3D) reconstruction methods are used, similar to CT angiography[5]. CT venography is at least as good as, if not better than, MR venography for the detection of dural sinus and deep cerebral venous thrombosis[11,14]. However radiation exposure in CT venography is a comparative disadvantage.

CT venography can be performed immediately after unenhanced CT in patients with signs and symptoms suspicious of cerebrovascular disease. If imaging of the venous structures is possible at the same time without moving the patient to the MR imaging suite, diagnosis and treatment are expedited[5]. On CT venography, a thrombosed dural sinus is revealed as an irregular filling defect in the sinus (Fig. 9) or an absence of contrast medium in the sinus. Before making a diagnosis of cerebral venous thrombosis, the radiologist should consider variations in the anatomy of the cerebral veins and dural sinuses, such as unilateral hypoplastic transverse and sigmoid sinuses with compensation via the contralateral transverse sinus[15], which may mimic a thrombosed sinus, leading to a false-positive diagnosis.

Treatment and outcome

In children with cerebral venous thrombosis, extent and location of the thrombosis, as well as complications, final outcome, and therapy, depend on the aetiology[16]. Conservative general medical care and supportive neurological care are the mainstays of treatment. Adequate hydration is crucial. Aggressive antibiotic therapy and antiepileptic medications should be used when appropriate[2]. The roles of anticoagulant therapy and thrombolytic therapy in children with cerebral venous thrombosis remain controversial[2,7]. In the absence of major cerebral nervous system haemorrhage, anticoagulation is appropriate. CT venography or MR venography is used for monitoring the response. If no anticoagulants are given, e.g. when there is significant haemorrhage, repeat CT venography or MR venography should be obtained one week after diagnosis to assess for propagation of the initial thrombosis[7].

The majority of children (90%) with cerebral venous thrombosis survive the initial illness. For approximately one-fourth of the patients who die, deaths are attributed to the cerebral venous thrombosis[7]. In the Canadian Registry, outcomes were assessed for 82 older infants and children with a mean interval from thrombosis to the last follow-up visit of 1.6 (range 0.52-5.2) years. There were 42 (51%) with normal outcome, 32 (39%) with neurological deficits, and eight (10%) who died, two from cerebral venous thrombosis. Other outcomes included seizures (11%) and recurrent venous thromboembolism (17%). Predictors of adverse neurological outcome or death included seizures at onset and presence of a venous infarct[17].

ABSTRACT

A 13-year-old boy had a history of severe headache for two weeks. He also had seizures and vomited many times. Neurological examination was normal except for bilateral papilloedema. Sagittal T1-weighted magnetic resonance (MR) images showed loss of the normal flow void in the superior sagittal sinus. There was hyperintensity, which came from subacute thrombosis in the posterior half of the superior sagittal sinus. MR venography confirmed loss of flow signal in the superior sagittal sinus. There was also thrombosis of the right transverse, right sigmoid, and left transverse sinuses, and haemorrhagic infarctions in the right frontal and left parietal regions. A diagnosis of dural sinus thrombosis with cerebral venous infarction was made. CT, MR imaging, MR venography, and CT venography findings are discussed in patients with cerebral venous thrombosis.

Keywords: cerebral infarction, computed tomography, cranial sinus thrombosis, magnetic resonance imaging

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SINGAPORE MEDICAL COUNCIL CATEGORY 3B CME PROGRAMME

Multiple Choice Questions (Code SMJ 200505A)

	True	False
Question 1. On CT and MR imaging, for patients with cerebral venous thrombosis:		
(a) Cerebral oedema does not conform to the pattern of an arterial distribution.	❑	❑
(b) Mass effect is uncommon.	❑	❑
(c) Haemorrhage is uncommon.	❑	❑
(d) There may be global or focal parenchymal change.	❑	❑
Question 2. In cerebral venous thrombosis:		
(a) Signs and symptoms are nonspecific.	❑	❑
(b) Cerebral catheter angiography is necessary for diagnosis.	❑	❑
(c) CT venography is better than MR venography for diagnosis.	❑	❑
(d) MR venography shows an absence of normal flow signal.	❑	❑
Question 3. The following may sometimes result in a false-positive diagnosis of cerebral venous thrombosis:		
(a) A high haematocrit count.	❑	❑
(b) An unmyelinated brain in neonates.	❑	❑
(c) A unilateral hypoplastic transverse sigmoid sinus.	❑	❑
(d) Subacute stage of venous thrombosis on MR imaging.	❑	❑
Question 4. For the imaging findings of cerebral venous thrombosis:		
(a) The cord sign is seen on enhanced CT.	❑	❑
(b) The empty delta sign is seen on unenhanced CT.	❑	❑
(c) The dense triangular sign is seen on enhanced CT.	❑	❑
(d) Bilateral parasagittal hypodense lesions are a common CT feature of superior sagittal sinus thrombosis.	❑	❑
Question 5. The following are predisposing factors of cerebral venous thrombosis in children:		
(a) Dehydration.	❑	❑
(b) Otitis media and mastoiditis.	❑	❑
(c) Lupus erythematosis.	❑	❑
(d) Nephrotic syndrome.	❑	❑

Doctor's particulars:

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