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Clinics in Diagnostic Imaging (30)

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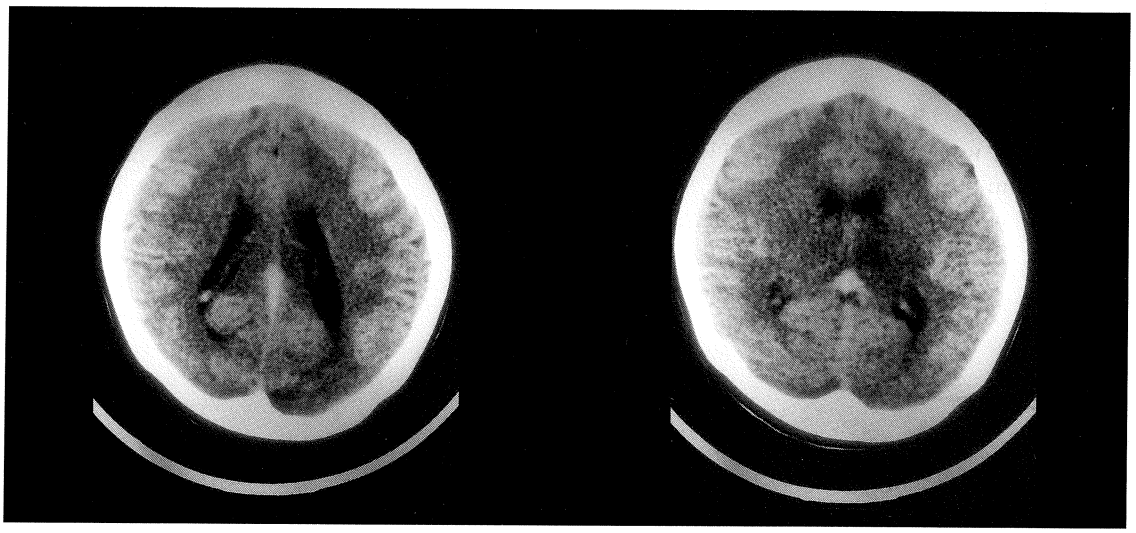

Fig 1a

Fig 1b

CASE REPORT

A 40 year-old woman presented with acute onset of confusion and disorientation. Her only significant medical history of note was that of thyrotoxicosis on follow-up. Physical examination revealed a small goitre. She appeared dazed and lethargic. Neurological examination was otherwise unremarkable, with no localising signs or weakness detected. A CT scan (Figs la & 1b) was performed. This was followed by an MRI study (Figs 2a & 2b). What do they show ?

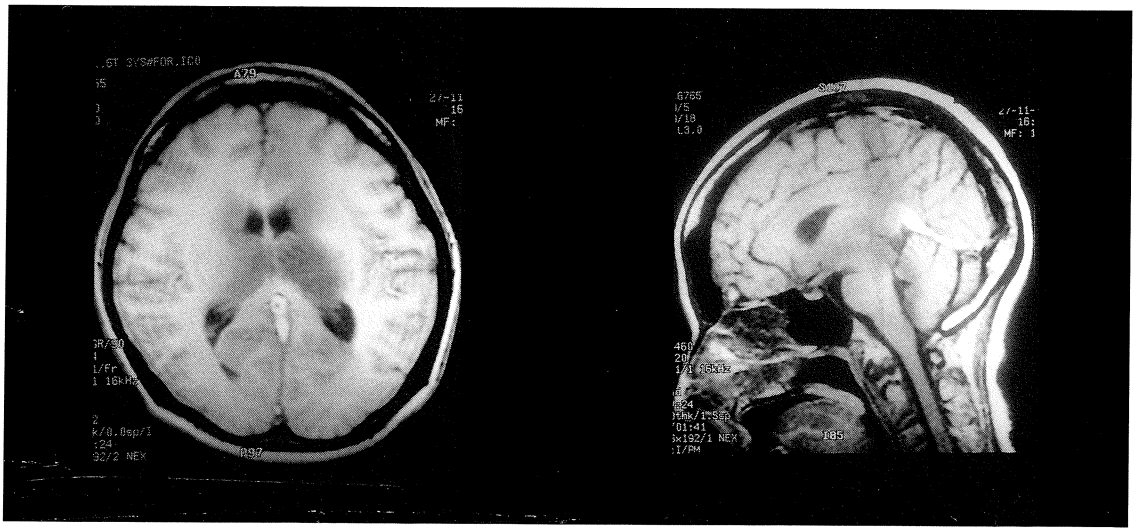

Fig 2a

Fig 2b

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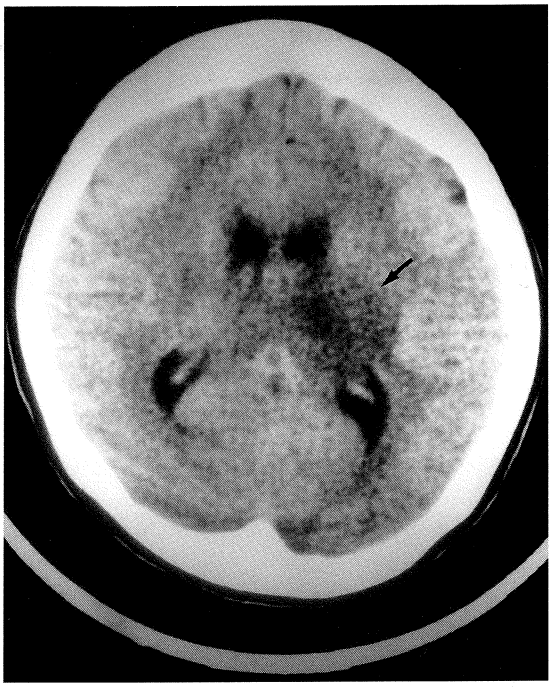

Fig 3 - Follow-up CT scan (comparable level to Fig 1b) shows resolution of the thrombus with residual left thalamic and lentiform nucleus infarcts (arrows).

IMAGE INTERPRETATION

Fig 1a demonstrates a clot in the straight sinus. There is decreased attenuation in both thalami and the left lentiform nucleus (Fig 1b) due to infarcts. The axial (Fig 2a) and sagittal (Fig 2b) T1-weighted images (T1WI) confirm the presence of a clot in the straight sinus and the vein of Galen, as well as the thalamic infarcts.

DIAGNOSIS

Vein of Galen and straight sinus thrombosis

CLINICAL COURSE

The patient was started on intravenous heparin at an initial dose of 800 units/hour and showed marked clinical improvement within 24 hours. This was subsequently converted to oral warfarin after four days. She was discharged home on a daily dose of 2 mg of warfarin. Although follow-up scans a week later (Fig 3) showed residual thalamic infarcts, she made an uneventful recovery with no significant neurological deficit. In this patient, the cause of the thrombosis is idiopathic.

DISCUSSION

For over a century following the initial description by Ribes in 1825, cerebral venous thrombosis (CVT) was mostly diagnosed at autopsy. This led to the classical description of a rare and often lethal disease characterised by headache, papilloedema, seizures, progressive coma and death. While modern imaging techniques have allowed for intravitum diagnosis, the true incidence is unknown as many patients are still underdiagnosed or misdiagnosed. It has been suggested that the incidence of CVT is higher in females and in the aged, reflecting the greater incidence of thromboembolic diseases in these categories. This was however not confirmed in recent works which showed that both sexes were equally affected and that all age groups, from the neonate to the elderly, are susceptible with possibly a slight preponderance in young adults[1].

There are many conditions that can predispose to CVT. These can generally be divided into three broad categories :-

a) local diseases such as infection, neoplasm and trauma
b) systemic conditions such as pregnancy, collagen vascular diseases, haematological disorders eg. polycythaemia, drugs especially oral contraceptives and cardiac diseases eg. heart failure and dehydration.
c) idiopathic

Of these causes, direct involvement of the dural sinus by infection or tumour, venous stasis, hypercoagulable states and dehydration are the most common.

CVT presents with a wide spectrum of symptoms and signs, the onset of which may be acute, sub-acute or even chronic. These clinical features are usually related to impairment of blood flow by the thrombus, with subsequent congestion and increased intracranial pressure. Typical signs of raised intracranial pressure such as headache, papilloedema, neck stiffness, nausea and vomiting and altered level of consciousness may then develop. Headache is the most frequent and is often the earliest symptom. The pain is often persistent, slowly progressive, does not respond to medication and may be localised or generalised. Its gradual onset often helps to differentiate it from subarachnoid haemorrhage. Lethargy and delirium are also commonly encountered. Convulsions are frequent and may accompany the headache, present days after the headache or occur in isolation. The nature of the convulsions may range from focal to generalised seizures, with status epilepticus and coma being encountered in the advanced stage. Cranial nerve involvement is usually seen only if the cavernous sinus is involved.

The diagnosis is based on a high index of suspicion aided by modern imaging techniques. Computed tomography (CT), with or without contrast, is often the first imaging examination performed. It is useful not only in confirming the diagnosis but also serves to rule out other conditions that may mimic CVT such as subarachnoid bleed, abscess and tumours. The CT findings have been well described and consist of direct and indirect signs[2].

The direct signs are:-

a) the cord sign - a rare sign due to the visualisation of thrombosed cortical veins on the unenhanced CT scan. A similar phenomenon has been reported[3] involving the straight sinus although viewed in length rather than in cross-section due to the orientation of the sinus to the plane of scan.
b) the dense triangle sign - which represents superior sagittal sinus (SSS) opacification by freshly congealed blood (Fig 4a)
c) the empty delta sign - where there is a filling defect in the affected vessel following contrast

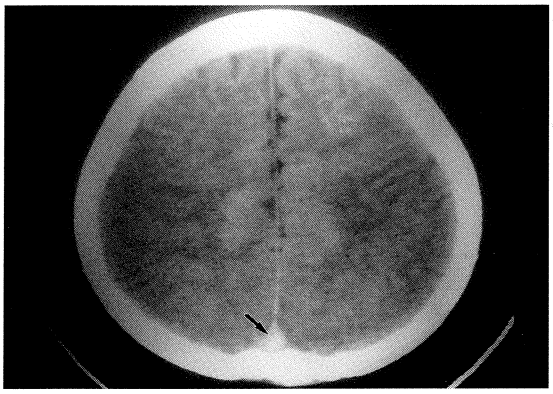

Fig 4a - Unenhanced CT scan shows the 'dense triangle sign' due to a clot in the posterior aspect of the superior sagittal sinus (arrow).

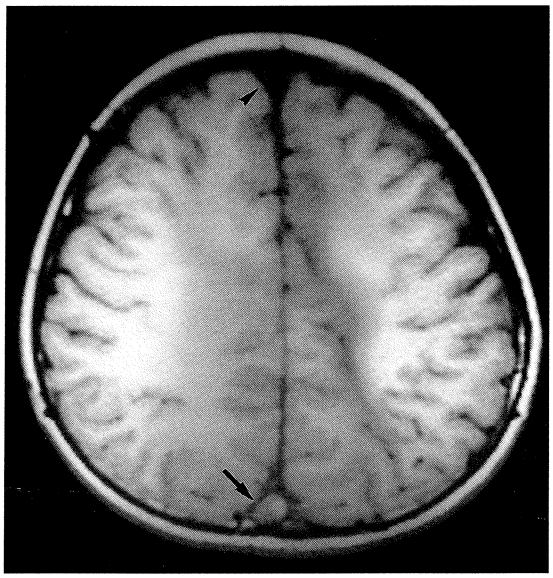

Fig 4b - MRI T1WI axial scan of the same patient as Fig 4a confirms the presence of the clot (arrow). Compare this to the normal flow void seen in the anterior aspect of the superior sagittal sinus (arrowhead).

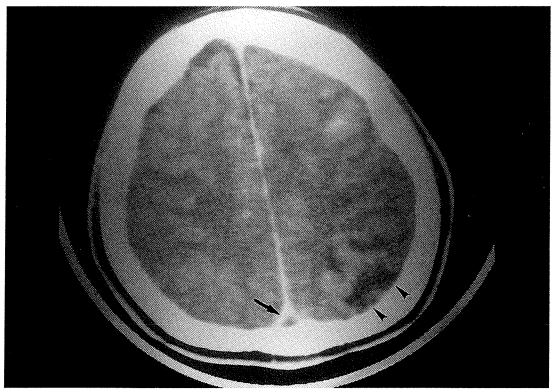

Fig 5 - CT scan of the brain. The 'empty delta sign' seen following contrast enhancement due to a clot (the filling defect) within the sinus with contrast outlining it (arrow). Note the venous infarct in the left parietal lobe posteriorly (arrowheads).

administration, due to the presence of the thrombus. Although the most frequent direct sign, it is estimated to be present in only about 30% - 50% of the cases (Fig 5).

Indirect and non-specific imaging abnormalities are more frequently encountered. These include the presence of small ventricles resulting from cerebral swelling, although this may be difficult to detect in young adults where the ventricles are normally small. Moreover, the presence of large ventricles does not rule out the possibility of CVT. Intense enhancement of the falx and tentorium is another recognised sign but again this is not an easy sign to detect. A more useful sign is that of venous infarcts which appear as areas of focal hypodensities. Although classically described as multiple haemorrhagic infarcts, usually in a parasagittal location, they can have a more varied appearance and may be single or multiple, unilateral or bilateral, and haemorrhagic or non-haemorrhagic. They usually occur superficially in the hemispheres in the case of SSS thrombosis or within the basal ganglia in deep venous thrombosis.

MRI offers major advantages in the diagnosis of CVT because of its sensitivity to blood flow, its better demonstration of the thrombus because of its multiplanar capability, and its ability to predict the age of the thrombus. In the early stage, there is absence of the normal flow void (Fig 4b), and the occluded vessel appears isointense on T1WI and hypointense on T2WI. With time, the thrombus becomes hyperintense, initially on T1WI (Fig 6) and then on T2WI. These changes in signal characteristics with ageing of the thrombus reflect the biochemical conversion of oxyhaemoglobin to methaemoglobin with time. Besides its ability in detecting thrombosis, MRI also offers the advantage of sometimes showing early parenchymal changes that may not be visible on CT scan and occasionally demonstrating an unsuspected underlying cause eg. mastoiditis. In addition, MR venography may also be performed and this can provide a good overview of the status of the intracranial veins and show the existence of collateral circulation patterns[4].

Angiography was previously a key procedure in the diagnosis of CVT. The partial or complete lack of filling of the venous sinuses is the best angiographic sign of CVT. This is most easily recognised when it involves the posterior third of the SSS, the lateral sinuses or the deep venous system, but may be more difficult to interpret in other locations such as the anterior third of the SSS or the left lateral sinus where it may be confused with hypoplasia. Although less frequently used nowadays with the advent of CT and MRI, it is still performed occasionally especially in cases where direct thrombolysis of the clot is contemplated. Isotope brain scanning with ^{99m}Tc has been shown to be useful in the diagnosis of SSS and lateral sinus thrombosis[5]. SSS thrombosis has also been demonstrated using ^{111}Indium platelet scintigram. However these techniques are of limited use in the acute setting and the false negative rates in the case of ^{99m}Tc have been reported to be higher than the other methods used.

The treatment of CVT is still controversial, and methods include surgical thrombectomy and anti-coagulation. The reluctance to use heparin in the past stemmed from the frequent occurrence of

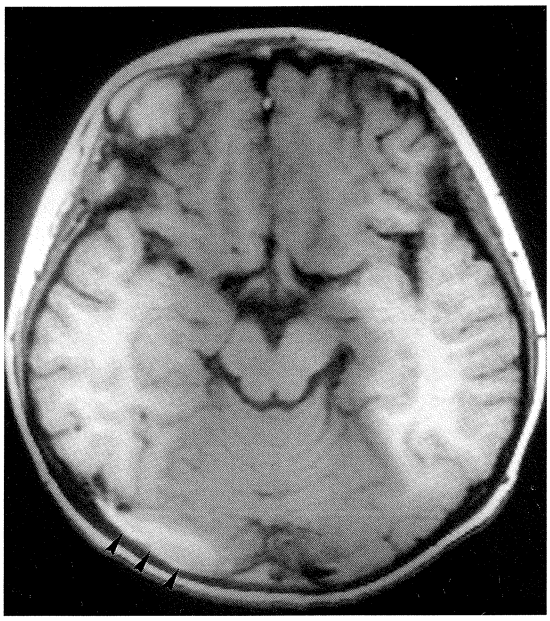

Fig 6 - MRI T1W1 axial scan shows a sub-acute thrombus in the right lateral (transverse) sinus (arrowheads).

haemorrhagic infarcts and fear of exacerbating such haemorrhage. Anti-coagulant therapy was thus mainly reserved for patients where the thrombosis was early, partial or actively propagating. Nowadays, however, anti-coagulation is generally accepted as the mainstay of treatment[6]. This may involve either systemic or localised anti-coagulation delivered by a catheter introduced into the affected sinus, usually via a femoral vein puncture. The latter method is gaining increasing popularity in view of the lower reported incidence of haemorrhagic complications[7]. Dramatic improvement usually occurs soon after institution of such treatment. Anti-convulsants and antibiotics (in cases where the underlying cause is an infective process) may also be useful as adjuvant therapy.

Previously diagnosed mainly at autopsy and frequently thought to be a lethal condition, the mortality from CVT has now dropped to 10% - 15% in most recent series. Poor prognostic indicators include a rapid rate of evolution of the thrombus, its occurrence in the very young and the aged, and the presence of haemorrhagic infarcts or coma. The single most important prognostic factor is however, thought to be determined by the underlying cause, with septic causes carrying a higher mortality rate. However, it is well recognised that if the patient survives, the prognosis for functional recovery is much better than in arterial thrombosis, with significant neurological sequelae seen only in a small number of patients[8]. This emphasises the importance of early diagnosis through an increased awareness of the condition, the use of appropriate imaging techniques and early institution of treatment.

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ABSTRACT

A 40-year-old lady presented with acute onset of confusion and disorientation. CT and MRI scans showed vein of Galen and straight sinus thrombosis. The clinical and imaging features of venous sinus thrombosis are described.

Keywords: thrombosis, venous sinus, cerebrovascular disease