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Dural Venous Sinus Thrombosis in Sickle Cell Disease in a West Indian

The Editor,

Sir,

Intracranial neurovascular manifestations in sickle cell disease include cerebral infarction, parenchymal and epidural haemorrhage, subarachnoid haemorrhage with single or multiple aneurysms, Moyamoya syndrome and posterior reversible encephalopathy syndrome.

Dural venous sinus thrombosis (DVST) in sickle cell patients has been reported rarely in patients who carry the trait and the sickle cell disease (1–3). We report the first case of DVST in sickle cell disease from the West Indies, a region where this disease is endemic.

A 14-year old Afro-Trinidadian male with confirmed haemoglobin SS (HbSS) disease presented with fever, generalized body pains and right orbital swelling for 10 hours. His past medical history was significant for recurrent painful crises and a right orbital cellulitis at the age of seven years. He was not on hydroxyurea treatment. Both parents had the trait and a brother was HbSS.

On examination, he was febrile with temperature 39.5 °C, oxygen saturation (SpO2) 90% on room air with a respiratory rate of 32/minute. He was ill-looking and dehydrated. Neurological and other system examination was normal. His Hb was 7.4 g/dL and white blood cell count was 39 200 cells/uL. Treatment included oxygen, intravenous fluids, paracetamol for fever and parenteral antibiotics. Chest X-ray revealed minimal scattered infiltrates in both lung fields. Computed tomography (CT) scan of the brain showed a left cerebellar acute ischaemic infarct. Computed tomography venography or magnetic resonance imaging (MRI) with venous MR of the brain and thrombophilia screens were not obtained due to unavailability at the time. The patient was intubated, ventilated and died sixteen hours after presentation. Postmortem showed cerebral oedema (brain 1240 g) and DVST which also involved the superior sagittal region (Figure). Other postmortem findings were bilateral pulmonary thromboembolism involving medium sized and small vessels with pulmonary infarction. There was also cardiomegaly 430 g, with biven-tricular dilatation. The liver weighed 1660 g and there were features of right heart failure and iron overload. Splenic atrophy (10 g) was also noted. Death was likely due to a combination of pulmonary arterial thrombosis and dural DVST.

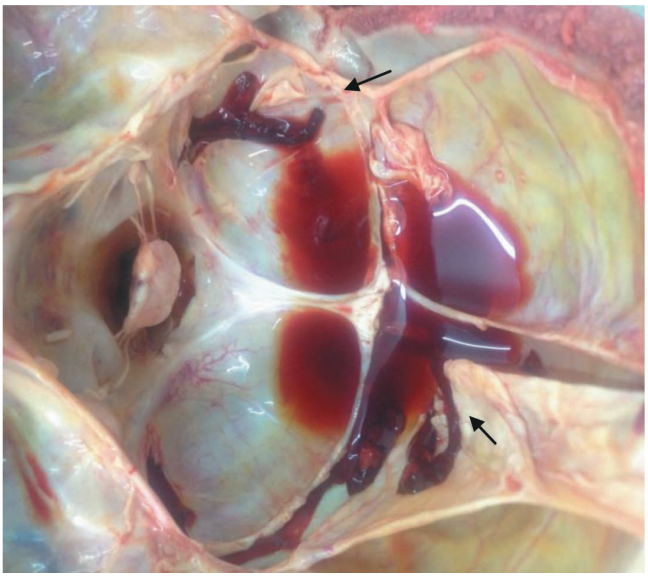

Figure: Extensive dural venous sinus thrombosis in anterior and middle cranial fossa (arrows).

Symptoms in DVST vary from only headaches, motor deficits, aphasia and seizures (4). Diagnosis is confirmed by detection of thrombus in the cerebral venous sinus system and in the brain parenchyma. Though CT scan with contrast can aid with diagnosis, the noninvasive investigation of choice is CT venography or magnetic resonance venography (MRV) which was unavailable to us at the time. Three signs seen on unenhanced CT scan are: (i) dense clot (triangle) sign, signifying acute thrombosis, present only in 55% of cases; (ii) empty delta sign, secondary to a partial recanalization of the thrombus on a contrast scan, seen in 20% of cases; (iii) cord sign: curvilinear or linear hyperdensity over the cerebral cortex caused by a thrombosed cortical vein. Computed tomography scan with contrast can miss the diagnosis in up to 40% of patients.

The initial treatment should be intravenous heparin/low molecular weight heparin with thrombolysis or thrombectomy reserved for cases undergoing secondary deterioration (5). Oral anti-coagulation with warfarin is given for three months if DVST was secondary to a transient risk factor or six to 12 months in patients with idiopathic DVST. Early recognition by appropriate imaging studies – CT venography or MRV – is necessary to begin treatment and thus prevent death.

Keywords: Dural venous sinus thrombus, haemoglobin, sickle cell disease

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Right Ovarian Mucinous Cystadenoma Coexisting with an Incidental Left Ovarian Haemangioma in a Postmenopausal Patient

The Editor,

Sir,

Haemangiomas were first described by Payne in 1869 (1). In the literature, their coexistence with various diseases of the female genital tract has been reported (2–5). Our case differs in that a postmenopausal woman had a mucinous cystadenoma in one ovary and an incidentally discovered cavernous haemangioma with focal positivity for progesterone receptor (PR) in the other ovary.

An 81-year old woman had right lower quadrant pain. Transabdominal sonography revealed a 10 cm right ovarian cystic and septated mass. No left adnexal lesion was reported. There was no sign of ascites. The serum cancer antigen (CA) 125 level of the patient was 144.9 IU/mL (ref 0–35), serum Ca-19-9 level was 24.9 IU/mL (ref 0–39).

Magnetic resonance images revealed a well circumscribed, slightly lobulated contoured, T1 SE hypo-, T2 SE hyperintense, multilocular cystic lesion in the right half of the pelvis (Figs. 1A–D). The left ovary measured 34 x 14 x 35 mm with slight spiculations and was isointense to myometrium on T1 SE and T2 SE weighted sequences (Fig. 1A–B). With injection of contrast material, slightly heterogeneous parenchymal contrast enhancement less than the myometrium was observed (Fig. 1A–D).

The result of the right ovary was reported as a mucinous cystadenoma on frozen section (Fig. 2).

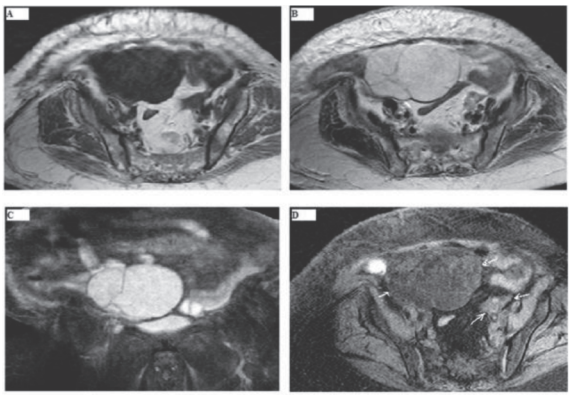

Fig. 1: Magnetic resonance imaging shows right ovarian lesion to be a multilocular cyst; (A) homogeneous hypo; signal intensity on T1-weighted images and (B) high signal intensity on T2-weighted images. Left ovary has slight spiculations isointense on T1 and T2 weighed images. (C) T2 SPAIR axial image (D) with contrast material; T1-weighted WATS CE axial images show the presence of a slightly heterogeneous enhancement in the left ovary (right lower arrow).

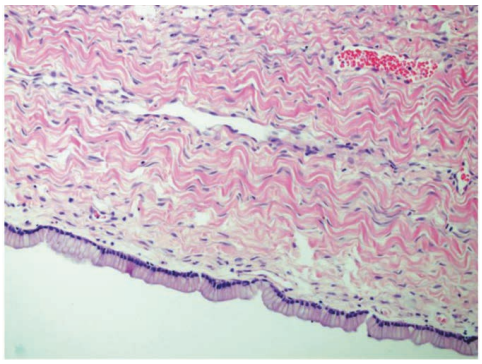

Fig. 2: Histologically, the right ovarian cyst was lined by mucinous epithelium (H&E; x 20).

On the cut surface of the left ovary (4 x 3 x 1.5 cm in size), a haemorrhagic nodular lesion, 2.5 x 1.5 cm in size, was noted. Microscopic sections revealed dilated, thin-walled vascular structures, variable in diameter, and the walls of which were comprised a single layer of endothelial cells not showing atypia (Fig. 3).

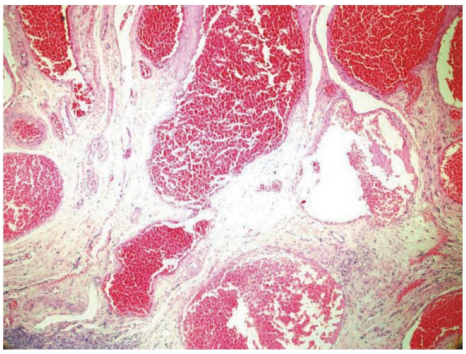

Fig. 3: Near the ovarian stroma, dilated thin walled vessels were lined by a single layer of endothelial cells, containing red blood cells in their lumen (H&E; x 4).