ORIGINAL ARTICLE

Cerebral Venous Thrombosis

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Abstract

Eleven patients (3 males and 8 females) in the age group 19-49 (mean 29 years), were diagnosed to have aseptic cerebral venous thrombosis over a period of four years. The presenting features were headache (11), papilledema (9), vomiting (6), altered consiousness (4), hemiparesis (4), generalized seizures (3), focal seizures (2), hemianesthesia (1) and buzzing in the ears in 1 patient. Diagnosis was established by cranial CT scan, MRI and MR Angiography. Predisposing factors identified in these patients were puerperal state (5), pregnancy (1), Behcet's syndrome (2) and sickle cell disease (1). Anticardiolipin antibodies were present in 2 patients and another patient had familial antithrombin III deficiency. All patients were treated with heparin and warfarin. In addition anticonvulsants and anticerebral edema measures were used when necessary. Eight patients completely recovered ; one patient with Behcet's syndrome and retinal vasculitis had severe visual loss ; one had residual hemiparesis and one optic atrophy.

Key Words

Cerebral Venous Thrombosis, Behcet's Syndrome, Anticardiolipin Antibodies.

Introduction

Cerebral venous sinus thrombosis (CVT) has been recognized over a century. With the advent of antibiotics, aseptic CVT has become the predominant form. Gowers (1888) recognized the occurrence of CVT during puerperium (1). Numerous clinical settings are known to favor CVT, viz. postoperative, postpartum and hypercoagulable states, cancer, cyanotic heart disease, sickle cell disease, antiphospholipid syndrome and other prothrombotic states (2). The common presenting features like headache, convulsions and altered consciousness are nonspecific and hence diagnosis demands high index of suspicion. CT scan may be suggestive but definitive diagnosis requires cerebral angiography, which is an invasive procedure. At present MRI with MRA is the investigation of choice for diagnosis and follow up (3). The mainstay of therapy is anticoagulation (4). The majority of patients show good recovery with reported mortality of 10-15% and residual sequel in 15-25% (5). Here we report eleven patients with CVT seen at King Fahd Hospital, Madina and discuss the approach to such patients.

Material and Methods

Eleven patients were diagnosed to have CVT over a period of 4 years. Detailed history was taken in all with reference to predisposing factors like pregnancy, puerperiun, hypercoagulable and hyperviscosity states and others. Complete physical and neurological examination was done in all. Laboratory evaluation included routine CBC, biochemistry, rheumatoid factor

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ESR, ANA, C3, C4, and sickling test, in all patients. Anticardiolipin antibodies, protein C, protein S and antithrombin III levels were done in 4 patients. Echocardiography, CT scan brain, MRI and MRA were carried out in all. All patients were treated with intravenous heparin to maintain the PTT levels twice the control value for two weeks followed by warfarin for six months. In addition, symptomatic therapy was instituted for seizures and or increased intracranial pressure. After discharge patients were followed every 6-8 weeks.

Results

Of 11 patients, 8 were females and 3 males. The mean age at presentation was 29 years (range 19-49).The patients' characteresties are shown in Table I. Two patients fulfilled the criteria for diagnosis of Behcet's disease. The commonest presentation was headache and papilledema (9/11). Details of clinical features at presentation are depicted in Table II. Of the five patients who presented in puerperium, one with antithrombin III deficiency had deep venous thrombosis complicated by pulmonary embolism. Of two patients with anticardiolipin antibodies, 1 had underlying Behcet's syndrome. Mitral valve prolapse was detected in 1 patient where as all others had normal echocardiograms. Cranial imaging (CT scan, MRI & MRA) showed thrombosis of superior sagittal sinus in 8; transverse sinus in 1; both the sinuses in 2 and cortical venous infarcts in 9 patients (2 hemorrhagic). Eight patients (including 2 with hemorrhagic infarct) recovered completely. Of two patients with Behcet's syndrome, one with retinal vasculitis suffered severe visual loss and another had optic atrophy. One patient with hemorrhagic infarct had residual hemiparesis.

Table I

Patients' characteristics

Total number	11
Female	8 (73%)
Male	3 (27%)
Age range	19-49 (mean 29yean)
Predisposing factors	
Puerperium	5 (45%)
Pregnancy	1 (9%)
CVT during previous pregnancy	2 (18%)
Behcet's syndrome	2 (18%)
Antithrombin III deficiency	1 (9%)
Anticardiolipin antibodies	2 (18%)
Sickle cell disease	1 (9%)

TableII

Clinical features at presentation

	Symptoms & signs	No. (%)
1.	Headache	11 (100%)
2.	Papilledema	9 (72%)
3.	Vomiting	6 (54%)
4.	Hemiparesis	4 (36%)
5.	Generalized seizures	3 (7%)
6.	Focal seizures	2 (18%)
7.	Confusion	2 (18%)
8.	Stupor	2 (18%)
9.	Hemianaesthesia	1 (9%)
10.	Buzzing in the ears	1 (9%)

Discussion

Cerebral venous thrombosis presents as acute or subacute neurological dysfunction with alteration in consciousness, focal neurological signs, seizures or increased intracranial pressure. It can occur at any age and needs to be considered in the diagnosis especially in the presence of predisposing factors.

Puerperal state was the commonest predisposing factor in five of our patients and one presented during pregnancy. Two of them had CVT during

earlier pregnancies also. Estimates of frequency of cerebral venous thrombosis in puerperium ranges from 1 in 1666-3000 pregnancies (6), and a higher incidence of 4.5 per 1000 obstetric admissions has been reported from India (7).

Cerebral venous thrombosis may be the first manifestation of Behcet's syndrome (8). Two of our patients had underlying Behcet's syndrome and one of them suffered severe visual loss due to retinal vasculitis.

Deep venous thrombosis and pulmonary embolism complicated the course of one patient with familial antithrombin III deficiency and CVT. Apart from antithrombin III deficiency (8,9) Factor V Leiden mutation (10) has also been reported to be an inherited risk factor for CVT.

Two of our patients had anticardiolipin antibodies. Anticardiolipin antibodies have been detected in 26% of patients with CVT compared to 3.2% in controls (11). Anticardiolipin antibodies may be an important factor contributing to cerebral venous thrombosis even in patients with other underlying disorders (12).

In the majority of patients, multiple dural sinuses are involved with superior sagittal sinus being the most frequent (8) as observed in ten of our patients. Two patients had lateral sinus thrombosis in addition. Two patients with isolated superior sagittal sinus thrombosis had features of intracranial hypertension without focal signs. Biousse *et. al.* have reported similar observation in 37% of patients with CVT (13). Cortical venous thrombosis, which frequently accompanies dural sinus thrombosis, increases the morbidity and mortality (2). Nine of our patients had cortical venous infarcts causing seizures, focal symptoms and signs.

Although some characteristic and highly specific CT findings strongly suggest the diagnosis of sinovenous thrombosis, cerebral angiography is the definitive method of confirmation (14, 15). However MRI with MRA, a noninvasive procedure is preferred for diagnosis as well as demonstration of recanalization of the dural sinuses (31).

At present anticoagulation is the favoured modality of therapy (16) along with anticonvulsants and measures to reduce the intracranial pressure when indicated. Recanalization of the venous system has been demonstrated in 36 of 42 patients treated with intravenous heparin for 3 weeks followed by oral anticoagulation (17). Low dose heparin was found to be efficacious in puerperal CVT even in patients with hemorrhagic infarction (18). Although there are no definite guidelines, six months of oral anticoagulation seems to be appropriate.

Eight out of eleven patients in the present study completely recovered and three had residual deficit. Prognosis is generally good in majority of patients despite extensive neurological deficit at presentation (5). Hence anticoagulation should be performed even in the presence of hemorrhagic infarction.

References

1. Gowers VJR (1888) Quoted by : D. Hilton Jones. Cerebral venous and dural sinus disorders. In : Michael Swash, John Oxbury, eds: Clinical Neurology. Churchill Livingstone New York 1991 : 973-85.
2. Adams MP, Jr., Biller J. Ischemic cerebrovascular disease. In: Bradley WG, Daroff RB, Fenichel GM, Marsden CD eds: Neurology in Clinical Practice. 2nd edition, Butterworth-Heienemann Boston 1996 : 993-1031.
3. Laffite F, Boukobza M, Guichard JP *et. al.* MRI and MRA for the diagnosis and follow up of cerebral thrombosis. *Clin Radiol* 1997 ; 52(9) : 672-79.
4. Villringer A, Einhaupl KM. Dural sinus and cerebral venous thrombosis. *New Horiz* 1997 ; 5(4) : 332-41
5. Beltran-Blasco Cerebral venous thrombosis : *Rev Neurol* 1997 ; 25(140) : 569-75.
6. Carroll JD, Leak D, Lee HA. Cerebral thrombophlebitis in pregnancy and puerperium. *Quarterly Journal of Medicine* 1996 ; 35 :347-56.

7. Srinivasan K. Cerebral venous and arterial thrombosis in pregnancy and puerperium. *Angiology* 1983 ; 34 : 731-46.
8. Bousser MG, Chiras J, Sautron B *et. al.* Cerebral venous thrombosis. A review of 38 cases. *Stroke* 1985 ; 16 : 199-213.
9. Kobayashi S, Hino H, Tazaki Y. Superior sagittal sinus thrombosis due to familial antithrombin III deficiency: case report of two families. *Rinsho Shinkeisaku* 1980 ; 20 : 904-10.
10. Ludeman P, Nabavi DG, Junker R *et. al.* Factor V Leiden mutation is a risk factor for cerebral venous thrombosis: a case control study of 55 patients. *Stroke* 1998 ; 29(12) : 2507-10.
11. Christopher R, Nagaraja D, Dixit NS *et. al.* Anticardiolipin antibodies: a study in cerebral veous thrombosis. *Acta Neurol Scand* 1999 ; 99(2) : 121-4.
12. Carhuapoma JR, Mitseas P, Leivine SR. Cerebral venous thrombosis and anticardiolipin antibodies. *Stroke* 1997 ; 28(12) : 2363-69.
13. Biousse V, Ameri A, Bousser MG. Isolated intracranial hypertension as the only sign of cerebral venous thrombosis. *Neurology* 1999 ; 53(7) : 1537-42.
14. Vines FS, Davis DO. Clinical - radiological correlation in cerebral venous occlusive disease. *Radiology* 1971 ; 98 : 9-22.
15. Gabrielson TO, Seeger JF, Knake JE *et. al.* Radiology of cerebral vein occlusion without dural sinus occlusion. *Radiology* 1981 ; 140 : 403-408.
16. Einhaupl KM, Villringer A, Meister W *et. al.* Heparin treatment in sinovenous thrombosis. *Lancet* 1991 ; 338 : 597-600.
17. Brucker AB, Vollert-Rogenhofer H, Wagner M. Heparin treatment in acute sinus venous thrombosis: a retrospective clinical and MR analysis of 42 cases. *Cerebrovasc Dis* 1998 ; 8(6) : 331-37.
18. Nagaraja D, Haridas T, Taly B. Puerperal venous thrombosis: therapeutic benefit of low dose heparin. *Neurol India* 1999 ; 47(1) : 43-46.