Letter to Editor

Figure 2a: Magnetic resonance venography (Coronal) images show non-visualization of the superior sagittal sinus and only partial visualization of the right transverse and sigmoid sinuses.

Figure 2b: Magnetic resonance venography (Sagittal) images show non-visualization of the superior sagittal sinus and only partial visualization of the right transverse and sigmoid sinuses.

arterial thrombosis simultaneously is an uncommon event. D Nagaraja et al reported three patients (two below 40 years of age) in whom simultaneous thrombosis of cortical veins and sinuses and major cerebral arteries was observed. The coagulation profile was not assessed and the diagnosis was based on partial autopsies. Our patient did not respond to conventional heparin but responded to low molecular weight heparin, suggesting an antithrombin III deficiency which we were unfortunately unable to confirm by laboratory tests. She was found to be heterozygous for methylenetetrahydrofolate reductase C667T mutation, which causes homocysteinemia. Elevated levels of homocysteine could explain the patient’s thrombophilic state and the apparent antithrombin III deficiency. Since she was positive for the C667T mutation, we concluded that homocysteinemia was the probable cause of her thrombophilia.

Stopping anticoagulants and replacing them with anti-platelet drugs could be harmful in the absence of a definitive diagnosis, as in our patient. Thrombophilic disorders of unknown etiology should be managed only with anticoagulants.

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References


Accepted on 18.10.2003

Cervical dystonia responsive to levodopa

Sir,

Spasmodic torticollis (ST) or idiopathic cervical dystonia (ICD), the most common adult segmental dystonia,1 is characterized by recurrent or sustained abnormal head postures. Drugs like trihexiphenydyl, clozapine,2 and mexiletine3 have been tried for treatment with an unfavorable response or with intolerable side-effects. Local botulinum toxin injections are currently the treatment of choice for ST.4 We report a case with cervical dystonia showing favorable response to levodopa.

A 23-year-old male started having intermittent turning of his neck to the right for two years, when he was 21 years old. It was observed at rest and disappeared while doing farm work in the field. Over the next month he had to abandon his work as his condition aggravated and his head remained turned to the right for most of the day. There were no other abnormal movements, or diurnal variation in his symptom. He did not report any geste antagonistic. He was prescribed propanolol and trihexiphenydyl without appreciable benefit. Levodopa with carbidopa (100+25-mg), one tablet twice daily improved his torticollis completely, but the effects continued
only for two to three hours after the administration. He visited our center after discontinuing the drugs. He was born of a non-consanguineous marriage and there was no family history of dystonia, significant psychological stress, or trauma to the neck or head. The past medical history was not significant.

The torticollis made his head turn to the right by 70-80 degrees. The rotation persisted for 75% of the waking time of the day. Even forceful straightening of his neck was unsuccessful. There was no head tilt or shoulder elevation. His speech and cranial nerves including the ocular fundii were normal. Keyser Fleischer ring was absent on slit lamp examination. There was no muscle wasting or muscle weakness and muscle tone was normal in all the muscles except the left sternocleidomastoideus. The deep tendon reflexes were normal bilaterally, with a flexor plantar response. There were no tremors on his extremities and no other involuntary movements were discernible apart from the torticollis.

The routine hemogram, serum biochemistry, serum copper and ceruloplasmin levels were normal. MRIs of the brain and the cervical spine were normal. Diagnosis of idiopathic cervical dystonia was made and he was put on 6mg/day of trihexiphenyldyl, without any favorable response. A further increase in the doses was associated with intolerable side-effects. A therapeutic trial with levodopa was considered based on his past history and 300 mg of levodopa with decarboxylase inhibitor per day was started in three divided doses. In one week twist of his neck decreased by more than 75% and he could easily maintain a straight position of his neck. Levodopa was stopped to confirm that the clinical improvement was indeed due to the drug. In 24 hours his symptoms recurred to the pre-treatment level. Controlled release levodopa (levodopa 200 mg + Carbidopa 50 mg) was started to sustain the clinical response for a longer period of time. The patient became symptom-free and was discharged on 600 mg per day of levodopa (controlled release). He has been symptom-free until now.

It appears that our patient was an atypical case of dopa-responsive dystonia, however, it is difficult to exclude the possibility of a particular type of ICD responding to levodopa, based on clinical evaluation. Younger age of onset, absence of geste antagonistic and responsiveness to levodopa were features against the diagnosis of ICD. In the presence of normal serum biochemistry and normal neuroimaging studies it is unlikely that the patient had secondary dystonia. Small, open-label trials of levodopa and dopamine agonists, in patients with ST have been without success.

Dopa-responsive dystonia (DRD) occurs typically in childhood, starts in the lower limbs and exhibits marked diurnal variation, and patients eventually develop parkinsonian features. These patients respond to low doses of levodopa. Involvement of the neck muscles is very rare in patients with DRD and has only been reported as minimal retrocollis. There were reports of DRD that showed torticollis associated with generalized dystonia, but there has been no report of DRD starting with torticollis or showing torticollis as a predominant feature.

Onset at an ‘older’ age, absence of diurnal variation and features of parkinsonism were against the diagnosis of DRD in this case. DRD is characterized by an age-dependent occurrence of symptoms and diurnal variation of symptoms might be absent in about 25% patients, particularly in patients with onset in the older age. Our patient could be a representative of such a group of patients of DRD. On close evaluation it appears that ‘dystonia’ and ‘dopa responsiveness’ are the key features of DRD, both of which were present in our case.

CSF neopterin assay, oral phenylalanine loading test, cultured lymphoectytic GTP cyclohydrolase (GCH1) activity and direct GCH1 gene sequence analysis can help in the diagnosis of DRD, but none of these tests is confirmatory. The GCH1 gene (located on the chromosome 14q 22.1-q 22.2) is the candidate gene for DRD. Genetic analyses can confirm the diagnosis of DRD in 60% of cases and up to 40% of cases can be sporadic. The availability of these tests could confirm the diagnosis in our case, however, as none of the above mentioned tests are confirmatory, our patient could still have DRD with an atypical presentation. While typical cases of DRD rarely elude diagnosis, in atypical cases of dystonia a significant response to levodopa could be a pointer to the disease. We conclude that in atypical cases of dystonia, a therapeutic trial of levodopa could be attempted. The need for diagnostic tests, which can help in the confirmation of the clinical diagnosis in a reliable and reproducible manner in such atypical cases, cannot be therefore, overemphasized.

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References

Calvarial tuberculosis

Sir

Calvarial tuberculosis is a rare manifestation of extrapulmonary tuberculosis with few reports in the literature.¹,²

A 16-year-old girl from a middle class family presented with a left parietal scalp swelling noticed incidentally three months prior to admission after a minor trauma. The swelling was 5 cm x 4cm in size, was non-tender and there were no signs of inflammation. The scalp over the swelling was normal. There was an impulse on coughing and a palpable calvarial defect with everted bony edges. General physical and routine hematological examination revealed no abnormality. Erythrocyte sedimentation rate was 22 mms in the first hour. X-ray of the skull revealed a lytic area in the left parietal bone with sclerosis of the edges.

Computerized tomogram of the head showed an encapsulated lesion with calcified foci communicating between the intracranial and extracranial compartments eroding the inner and outer tables of the skull near the left parietal eminence (Figure 1). The margins of the lesion exhibited minimal contrast enhancement. Magnetic resonance imaging of the brain showed a hypointense and intermediate signal lesion over the left parietal lobe extradurally in T1 weighted images with intermediate to hyperintense signals on T2 weighted images, extending medially up to the superior sagittal sinus.

The lesion was exposed after taking an incision around the swelling. The whole lesion was excised piecemeal, laterally up to the normal bone margin. The contents of the lesion resembled the cheesy material of an epidermoid cyst. The capsule of the lesion was peeled off completely from the dura. The defect in the calvarial bone was repaired with the help of iliac bone graft. The histopathological examination confirmed that the lesion was tuberculous osteomyelitis. The patient was treated with antituberculous drugs. A repeat CT head scan confirmed that the grafts had fused. HIV status was checked and was found to be negative. The patient recovered clinically and gained weight.

Isolated calvarial tuberculosis is rare³ but can be seen in association with pulmonary tuberculosis, tuberculous osteomyelitis involving other bones, cervical lymphadenitis, renal and intestinal tuberculosis. The frontal and parietal bones, having greater area of diploic space and cancellous bone are more vulnerable.⁴ The tubercular process reaches the vault bones possibly through the blood stream. Concentrically-placed fibroblasts proliferate and encircle the tubercular granulation tissue and prevent its extension through the bone; which when deficient, extension occurs through the inner or outer or both tables.⁵ Sutures are not a strong barrier to spread but the dura usually prevents intradural extension. An extensive area of destruction occurs before clinical presentation.

The radiological picture can vary, with either an osteolytic or sclerotic variety being seen; the osteolytic type itself can be circumscribed or of the spreading type.⁵ The CT picture of tuberculous osteomyelitis is not very specific, with pyogenic osteomyelitis, calvarial metastases, myeloma, hemangioma, giant cell tumor or even an aneurysmal bone cyst and Langherans cell histiocytosis forming the differential diagnoses.⁵

The criterion for diagnosis is isolation of tubercle bacilli, which most of the time is not possible, as in our case. In the absence of this, histological features of caseous granuloma are often the only clue, along with the radiological features.⁶ On many occasions, there may not be supporting evidence of extracalvarial tuberculosis or raised erythrocyte sedimentation rate.⁵ Clinical and radiological response to specific antituberculous chemotherapy is usually good.³

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