

Short Reports

nerve pareses. Fundoscopy revealed papilledema. A magnetic resonance angiogram (MRA) showed a block in the anterior third of the superior sagittal sinus and a thrombus partly occluding the confluence of sinuses (Figure 2). The patient was placed on anticonvulsants and anti-edema drugs and heparin with which his symptoms improved over five days. Heparin was then changed to oral anticoagulants and he was discharged when he was asymptomatic.

Discussion

Thrombosis of the superior sagittal sinus (SSST) and torcula is rare and is usually attributed to scalp or skull infections, oral contraception, dehydration, pregnancy, blood dyscrasias and metabolic derangements.^{1,2} Although post-traumatic SSST has been reported, it still remains an unusual complication of traumatic brain injury. The high mortality rate (40%-80%)³ stresses the need for early diagnosis and treatment.

The diagnosis is based on a high index of clinical suspicion, confirmed by imaging. General symptoms and signs are headache and seizures due to cerebral venous infarcts.⁴ Buonanno et al³ reported 11 cases of SSST diagnosed with CT and later confirmed by angiography or autopsy. A reliable finding on CT is the "empty triangle" sign seen on contrast films in the cuts slicing perpendicularly across the posterior aspect of the sinus. The "empty triangle" is due to the presence of an isodense clot within the sinus enclosed by an area of engorged vessels. Less specific findings include small ventricles, gyral enhancement in a distribution of venous infarct and multiple focal bilateral parasagittal hemorrhages.

The pathogenesis of SSST is not well established. Carrie and Jaffe⁵ state that abnormalities in the clotting mechanism, disturbances in blood flow or damage to the capillary endothelium may predispose and lead to thrombosis. Alteration in coagulation after head injury has been documented supporting this hypothesis.⁶

Treatment includes maintaining good hydration, and the administration of anti-edema agents like dexamethasone and mannitol. Anticonvulsants are necessary to prevent seizures. There are reports of the successful management of SSST with



Figure 2: Magnetic resonance angiogram showing block in the anterior part of the superior sagittal sinus with multiple collaterals and a partial thrombus at the torcula

heparin and urokinase, including the restoration of the patency of the sagittal sinus with continuous urokinase infusion directly into the sinuses via a transfemoral transvenous microcatheter.^{7,8}

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Atlantoaxial dislocation in idiopathic cervical dystonia

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We report a case of severe cervical spondylosis and atlantoaxial dislocation (AAD) in association with idiopathic

cervical dystonia (ICD) in a middle-aged male. To our knowledge, this is the first case of ICD reported in association

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with AAD.

Key Words: Idiopathic cervical dystonia, Atlantoaxial dislocation.

Introduction

Dystonia is a type of movement disorder, which results in a twisting movement of the parts involved.¹ It may be focal or generalized. ICD is the most common form of adult onset focal dystonia.² When dystonia involves the cervical spine for a long time, it can result in degenerative changes leading to myeloradiculopathies.²⁻⁴ A rare case with ICD is reported where, in addition to the gross degenerative changes in the cervical spine the patient had AAD.

Case Report

A 40-year-old male was admitted with a history of progressive difficulty in walking and stiffness of both lower limbs for two months. He had repeated falls whilst walking during this period. He was able to perform all his routine work without assistance until 1 month prior to admission, at which time he needed a walking stick to move around. Since childhood, he had suffered from involuntary twisting movements involving the muscles of the neck and face, which later extended to the limbs. His family history was unremarkable.

On examination, he had dystonic movements involving predominantly the neck and trunk and, to a lesser extent the face and the limbs. He had spastic Grade 4 quadriparesis. Kinesthetic sensations were impaired in all four limbs. The rest of the neurological and systemic examination was normal. Routine blood investigations were normal. Screening for Wilson's disease was negative.

Investigations revealed degenerative changes in the cervical spine and AAD (Figure 1). However, there was no obvious compression noted at the cervicomedullary junction. The patient refused any surgical treatment.

Discussion

Oppenheim⁵ in 1911 coined the term dystonia, a condition characterized by uncontrollable muscular contractions that produce abnormal posture. Dystonia is defined as a syndrome of sustained muscle contractions, frequently causing twisting and repetitive movements or abnormal posture. Focal (cervical) dystonia is defined as an involuntary twisting and turning of the neck caused by abnormal involuntary muscle contracture.^{3,6} Abnormal postures frequently seen in cervical dystonia are torticollis, laterocollis, anterocollis, retrocollis or any combination of the above. In 1970, Levin and associates⁷ noted that abnormal neck movements and posture caused premature spondylotic changes and degenerative changes were greater on the side of neck flexion. Nishihara and colleagues,⁸ in their study of athet-

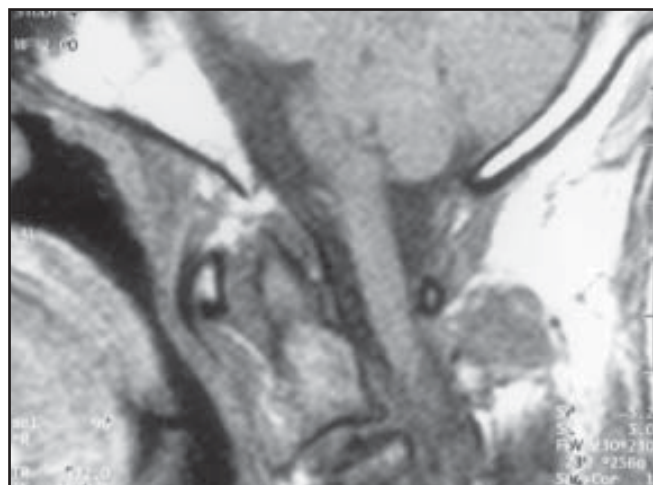


Figure 1: T1 weighted (TR532ms / TE 15ms) sagittal image shows AAD

oid patients with cervical spondylotic myelopathy, found that C₃-C₄ and C₄-C₅ were the most frequently affected sites. It was suggested that patients with cervical movement disorders have a higher velocity during flexion or extension and an increased range of neck movements. This causes greater shearing forces and bending movement of the cervical structures, which predisposes the cervical spine to premature degenerative changes. In addition to these changes our patient also had AAD, which has not been described in the literature before. Cervical spondylotic changes with atlantoaxial instability in patients with abnormal movements have been previously described by Yashioka.⁹ Greenberg et al¹⁰ also reported AAI in patients with abnormal movement disorder.

The management of AAD in movement disorders is controversial and difficult as postoperative immobilization of the cervical spine is difficult due to the involuntary movements.

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