Case Report

Cerebellar ataxia and parkinsonism owing to a couple of intracranial arachnoid cysts in a patient with multiple skeletal deformities

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Abstract

A 16-year-old school boy with multiple skeletal deformities (in the form of severe pectus excavatum, kyphoscoliosis, and hallux valgus) was investigated for a sudden development of right hand parkinsonian-like tremor and clumsiness and for the findings of features of mild left sided cerebellar dysfunction. Magnetic resonance imaging of the brain with and without contrast were done revealing two small arachnoid cysts, one in the left anterior temporal area within the left middle intracranial fossa causing minimal focal mass effect on the adjacent parenchyma, and the other one was in the posterior intracranial fossa inferolateral to the left cerebellum causing pressure effect on it. The patient was referred to the department of neurosurgery whose decision was not to operate.

KEY WORDS: Parkinsonian tremor, arachnoid cysts, cerebellum, intracranial fossa

Introduction

Arachnoid cysts are benign congenital intra-arachnoidal cerebrospinal fluid (CSF) collections. They constitute roughly 1% of all nontraumatic intracranial masses.[1] Nearly all arachnoid cysts are benign and do not show any manifestations for the whole of life.[2] However, surgery may be warranted when they are large or are complicated by subdural hematoma or if they are causing significant neurological symptoms. In the literature, a number of cases of posterior fossa arachnoid cysts were reported to cause cerebellar ataxia, an example of which is the case reported by Aggouri et al.[3] Musella and Elyidge[4] reported a case of Parkinsonian-Like Syndrome Caused by Cyst in Posterior Fossa. To our knowledge, no case was reported to link between arachnoid cysts and the triad of cerebellar ataxia, parkinsonian-like disorder, and multiple skeletal deformities.

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Case Report

A 16-year-old right-handed male with no history of learning difficulties was brought to the hospital by his father complaining of clumsiness and resting rolling involuntary movement of the right hand, typical of parkinsonian tremor. He gave no history of head titubation or tremor elsewhere in the body. He denied any history of difficulty initiating movement, any gait difficulty, speech abnormality or weakness of any part of his body, and no history suggestive of seizures. He gave a history of intermittent moderately severe bouts of diffused bilateral headaches dated back to the last 6 months. He revealed no history of recent or previous head trauma. There is no family history of inheritable neurological diseases. On examination, abnormal facies were noted, and he was found to exhibit high-arched palate, pectus excavatum, and kyphoscoliosis [Figures 1 and 2]. Neurological examination revealed that he was fully oriented with normal speech. Upper limb examination confirmed the presence of right hand resting, rolling tremor. The tone, deep tendon reflexes and power were normal in both upper limbs. The left upper limb showed abnormal finger nose test with intention tremor and pass pointing and dysdiadokinesia. All modalities of sensation were normal in both upper limbs. The lower limbs showed bilateral hallux valgus, normal tone, and power but bilaterally brisk deep tendon reflexes and impaired heel-shin test of the left lower limb. The gait appeared to be

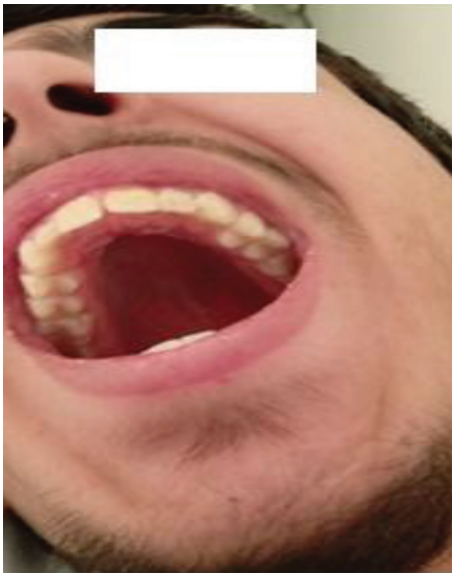

Figure 1: High-arched palate

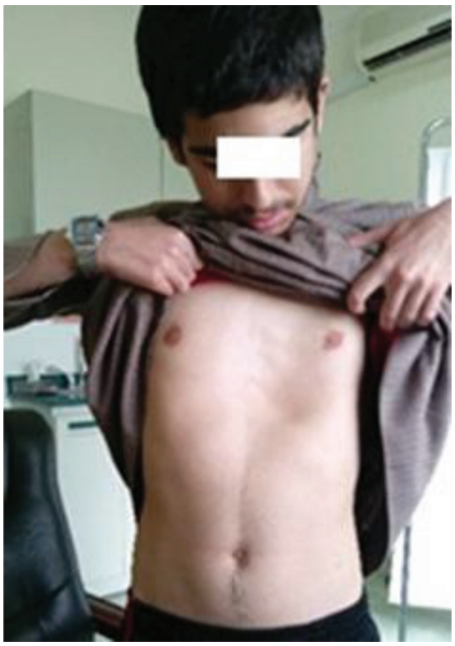

Figure 2: Severe pectus excavatum

normal, but the tandem gait test was impaired. Fundoscopy was done and was completely normal.

Magnetic resonance imaging (MRI) of the brain with and without contrast were done, the report of which came showing a small left anterior temporal CSF structure seen within the left middle intracranial fossa causing minimal focal mass on the adjacent brain parenchyma, measuring about 3 × 1.6 × 2 cm suggestive of small left temporal arachnoid cyst. Another small

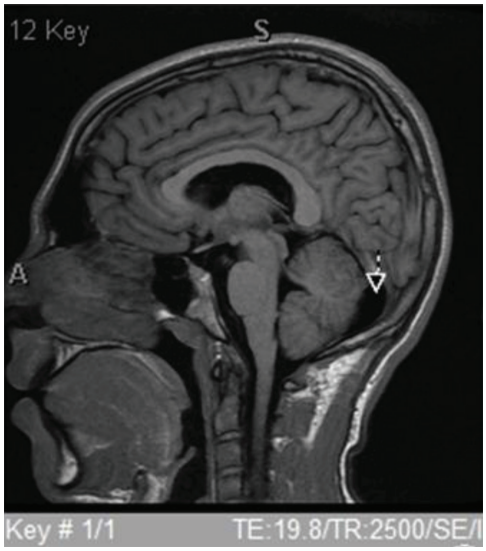

Figure 3: Sagittal T1 fluid-attenuated inversion recovery MRI without contrast showing a posterior fossa arachnoid cyst (PFAC; arrow)

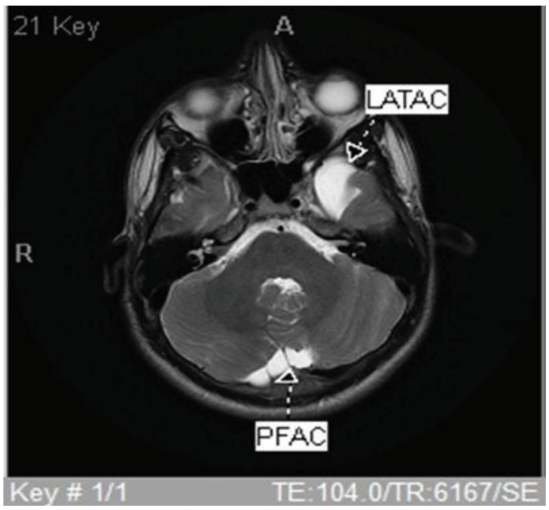

Figure 4: T2-weighted axial MRI showing left anterior temporal arachnoid cyst (LATAC) and PFAC

CSF structure was seen posterior inferolateral to the left cerebellar hemisphere within the posterior intracranial fossa measuring about 2 × 1.6 × 1.2 cm also suggestive of an arachnoid cyst. All laboratory studies done were normal including renal function test, thyroid function test, and parathyroid hormone and calcium studies.

The patient was diagnosed with cerebellar ataxia and parkinsonism secondary to posterior fossa and left middle fossa arachnoid cysts, respectively. The patient's resting tremor responded well to antiparkinsonian drugs and was referred to the neurosurgery department whose decision was against surgery.

Discussion

Intracranial arachnoid cysts were first described by Bright[5] in 1831 as "serous cysts forming in connection with the arachnoid and apparently lying between its layers."

The incidence of the diagnosis of arachnoid cysts has increased significantly owing to the recent advance in neuroimaging. These cysts are usually benign and cause no symptoms. Those involving the middle cranial fossa are associated with temporal lobe hypogenesis and often cause erosion and expansion of the overlying cranium.[6] When clinical features are present, the most common indications are headache, dizziness, and convulsive episodes.[7] Although the vast majority of arachnoid cysts are congenital in origin, some were found to be secondary to trauma, an example of which is the case reported by Valluru and Raj[8] of a 22-year-old man with an incidental finding of grossly dilated occipital and temporal horns of the right lateral ventricle following a head trauma owing to road traffic accident. Our case is a male adolescent with multiple skeletal deformities with no family history of inherited neurological diseases who presented with a sudden development of parkinsonian tremor and was later found to have mild cerebellar ataxia as well. The differential diagnosis should include multiple sclerosis, spinocerebellar degeneration, and secondary causes of parkinsonism-like Wilson's disease and drug intoxication. Extensive laboratory studies including thyroid function test and other hormonal profile were normal. MRI of the brain was requested and came back to show a couple of arachnoid cysts involving the left anterior temporal lobe and the posterior fossa causing compression of the cerebellum.

Conclusion

This is a case of two intracranial arachnoid cysts, one in the left middle fossa and the other one in the posterior intracranial fossa, which have led to cerebellar ataxia and secondary parkinsonism. Initially, a differential diagnosis of multiple sclerosis, benign essential tremor, spinocerebellar degeneration (in view of the multiple skeletal deformities), and metabolic causes were considered. An MRI was diagnostic of the couple of the arachnoid cysts. The patient was referred for a neurosurgical consultation to be assessed for surgical intervention, but the decision was in favor of conservative management. The patient's parkinsonian-like tremor responded very well to antiparkinsonian drugs.

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