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Case Report

Tuberous Sclerosis in Combination with Ossification of the Posterior Longitudinal Ligament: Association or Incidental Finding?

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ABSTRACT

Tuberous Sclerosis and Ossification of posterior longitudinal ligament (OPLL) are two different disorders of different etiology. Their association has not been reported till date. We report a 39 year old male patient with Tuberous Sclerosis complex who presented with quadriparesis due to compressive cervical myelopathy etiology being OPLL. Patient was managed conservatively.We propose that disturbances in growth hormone associated with tuberous sclerosis may be cause of OPLL in our patient.

KEYWORDS: Cervical Myelopathy, Ossification of Posterior Longitudinal Ligament, Seizures, Tuberous sclerosis

INTRODUCTION

Tuberous sclerosis (TS) or Epiloia Or Bournervlle's Disease is neuro-cutaneous syndrome characterized by abnormalities of both the integument and centre nervous system (CNS) with an estimated frequency of 1/6000. TS is an extremely heterogeneous disease with a wide clinical spectrum varying from severe mental retardation and incapacitating seizures to normal intelligence and a lack of seizures, often within the same family.[1] Ossification of the posterior longitudinal ligament (OPLL) is a condition of abnormal calcification of the posterior longitudinal ligament. The most common location is at the cervical spine region. Compression of spinal cord caused by OPLL may lead to neurologic symptoms and in the cases with severe neurologic deficit, surgical treatments are required. However, the exact pathogenesis and natural history of OPLL remain unclear.[2] There has been no case reported with a combination of both conditions. We here report a case of this unusual combination of tuberous sclerosis with OPLL.

CASE REPORT

39 year old male patient was admitted in our institute under department of neurology with history of trauma sustained due to fall over stairs followed by weakness of all four limbs two days prior to admission. Patient also complained of decreased pain and temperature sensation below lower part of neck. Patient also had history of urinary retention for which he was catheterised. There was no history of head injury. There was no history of seizure preceding the fall. Patient had history of traumatic quadriparesis 5 years back and had near complete recovery with conservative management. He was not investigated that time. He had past history of childhood seizures but was not on treatment.

On examination patient was conscious oriented, vitals were stable. He had facial angiofibromas (Fig.1), multiple hypomelanotic macules and Shagreens patch over back (Fig.2). Higher mental function examination was normal. He had grade 3 power in upper limbs and grade 2 in lower limbs. DTRs were brisk with extensor plantar on both sides. All modalities of sensations were decreased below T2 dermatome.

Haematological and biochemical workup was normal. Growth hormone level was 13.3 ng/ml (N=1-9 ng/ml). Other pituitary hormone levels were normal (ACTH-12.2pg/ml (N=6-76pg/ml), FH-5mIU/ml (N=1-12mIU/ml), LH-6.7mIU/ml (N=2-12mIU/ml), TSH-0.56uIU/ml (N=0.34-4.25uIU/ml)). Patient MRI of cervical spine showed diffuse ossification of posterior longitudinal ligament (OPLL) from C2 to C7 level leading to cervical cord compression and cervical stenosis. (Fig. 3 and Fig. 4)

CT scan of head revealed subependymal tiny calcified nodule abutting bilateral lateral ventricle. (Fig. 5)

As patient refused surgical intervention he was treated conservatively with steroids and neck immobilisation. Patient had minimal improvement of grade in muscle power after 7 days and was discharged.

DISCUSSION

The clinical and radiographic features of tuberous sclerosis complex have now been divided into major and minor categories based on the apparent degree of specificity for tuberous sclerosis complex of each feature. Major features include Facial angiofibromas or

Fig. 1- Patient showing multiple facial angiofibromas.

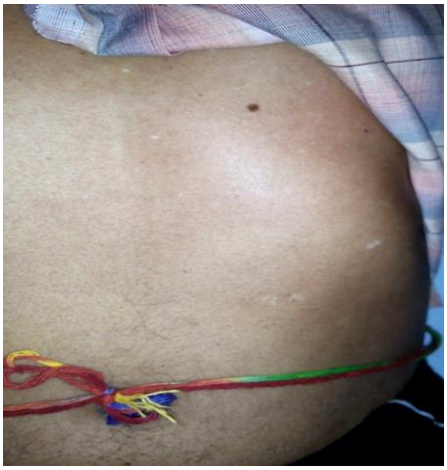

Fig 2: Multiple hypomelanotic macules over back

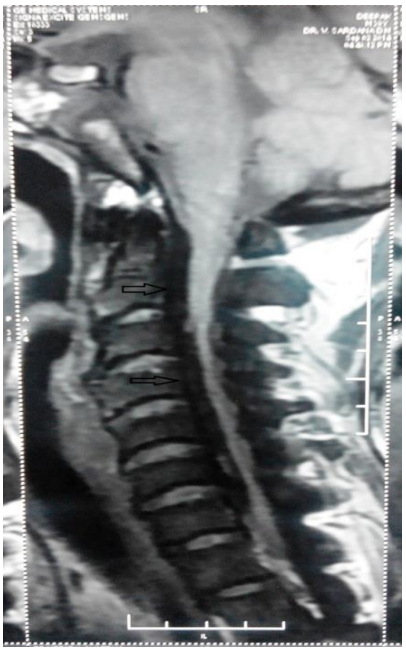

Fig 3: T1W sagittal image showing hypointense lesion extending from C2-C7 in posterior Part of vertebral body s/o OPLL

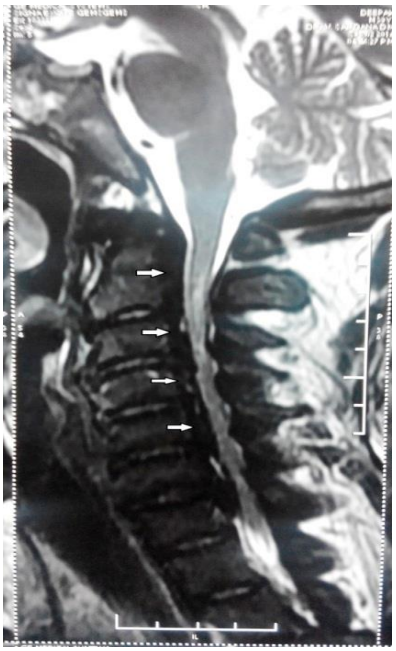

Fig 4: T2W sagittal image showing hypointense lesion extending from C2-C7 in posterior Part of vertebral body s/o OPLL and segment of intramedullary altered signal intensity Seen in C2-C3 s/o cord edema due to compressive Myelopathy.

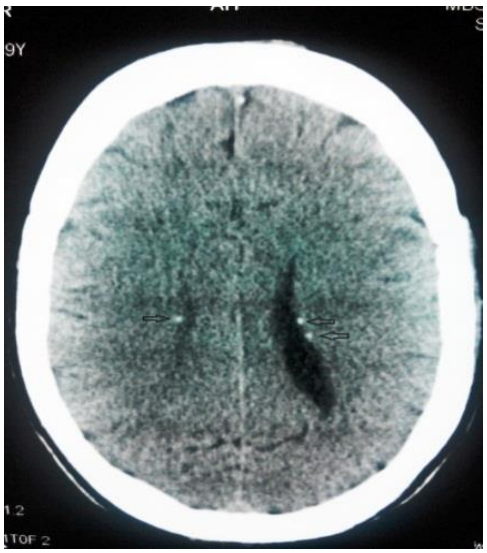

Fig 5: Axial CT brain image showing multiple hyperdense nodules seen in subependymal region s/o subependymal calcified nodules.

forehead plaque, Non traumatic ungual or periungual fibroma, Hypomelanotic macules (>3), Shagreen patch (connective tissue naevus), Cortical tuber, Subependymal nodule, Subependymal giant cell astrocytoma, Multiple retinal nodular hamartomas, Cardiac rhabdomyoma, single or multiple Lymphangiomyomatosis, Renal angiomyolipoma.

Minor features include Multiple randomly distributed pits in dental enamel hamartomatous rectal polyps, Bone cysts, Cerebral white matter migration tract, Gingival fibromas, Non renal hamartoma, Retinal achromic patch, Confetti skin lesions, Multiple renal cysts. Definite tuberous sclerosis complex diagnosis need 2 major features or1 major feature + 2 minor features.[3] Our patient had facial angiofibromas, multiple hypomelanotic macules, Shagrens patch and subependymal calcifications satisfying criteria for definite diagnosis of tuberous sclerosis complex.

OPLL is a hyperostotic condition of the spine that causes severe neurologic symptoms induced by spinal cord compression. OPLL isa multifactorial disease in which complex genetic and environmental factors interact. The polymorphisms in collagen11A2 gene (Col11A2) and collagen 6A1 gene (Col6A1) may be associated with the disease. OPLL was significantly correlated with thickness of each posterior longitudinal ligament in cervical spine involving mid-cervical vertebra. In Caucasian Americans and Germans, C6 was the most common site. About 80-85% of OPLL patients experience slow progression, but the symptoms become suddenly aggravated or even quadriplegia may appear by mild injuries.[4] Our patient presented with neurological deficit precipitated by trauma.

There are published case reports of elevated growth hormone levels in tuberous sclerosis which may be of pituitary origin or may possibly be secreted ectopically by a hamartoma.[5] Elevated growth hormone/IGF1 is one of proposed mechanism for OPLL.[6] We propose that disturbances in growth hormone associated with tuberous sclerosis may be cause of OPLL in our patient. Though the association may be incidental, vigilant observation is needed to assess the correlation between two.

CONFLICT OF INTEREST: None declared.

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