Case Report

Central atlantoaxial instability as a cause of syringomyelia mimic intramedullary lipoma

ABSTRACT

A case of a 32-year-old male patient is reported. He was admitted with complaints of burning dysesthesias over his right upper limb and chest and spasticity in the legs. Investigations revealed a long segment intramedullary tumor, image intensity of which matched lipoma. Imaging of craniovertebral junction suggested atlantoaxial "facetal" instability. Atlantoaxial fixation was done, and the intramedullary lipoma was not physically handled or manipulated during surgery. The patient improved in his neurological condition following surgery. The follow-up imaging showed that the intramedullary lipoma reduced significantly in its dimensions. From the case, it appears that the presence of "fat" and "water" in the intramedullary location might have similar pathogenesis.

Keywords: Atlantoaxial dislocation, intramedullary lipoma, syringomyelia

INTRODUCTION

A case of intramedullary lipoma mimicking syringomyelia in its configuration and also possibly in its pathogenetic characteristic features is reported. The management issues in this rare and surgically formidable clinical condition are reported. Our literature search did not locate any similar report.

CASE REPORT

A 32-year-old male presented with a history of dysesthesia in the form of burning pain over the entire right hand and right side of the chest and back for 2 years. The pain was progressively increasing in intensity and had worsened over the past 2 months such that he could not lift his arm or bear the touch of his clothes over his skin. He had stopped wearing his shirt and preferred to be bare-chested and was confined to his home. The pain was continuous and affected his sleep. There were no bladder or bowel-related complaints. He also noticed the wasting of his small muscles of the hand. For about 3 weeks, he had stiffness and weakness in his legs, making walking difficult. Neurological examination revealed mild spasticity in the hands and both legs. He had decreased sensations of pain and temperature below the C4 dermatome on both sides, more prominently on the right side. His skin was hypersensitive to touch on the right half of the upper chest and right upper limb (right C5-7, T1-4 dermatomes) due to which he avoided wearing a shirt. Magnetic resonance imaging (MRI) of the spine showed an intramedullary lesion occupying a significant length of the entire cord. The lesion was hyperintense on both T1-weighted and T2-weighted images [Figure 1]. The imaging features were suggestive of an intramedullary lipoma. Fat suppression images confirmed that the lesion was consistent with a lipoma. Computed tomography (CT) of the craniovertebral junction revealed

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fusion of C3-4 vertebrae and Goel Type 2 atlantoaxial facetal instability.[1] The patient was operated for a C1-C2 lateral mass distraction and fixation.[2,3] During surgery, the craniovertebral junction was found to be significantly unstable. The patient had a remarkable improvement in the burning pain sensation in his upper limb and his chest. He started to wear his shirt, and he was able to lift his arm above the head. The spasticity and power in the legs returned to normalcy. At a follow-up of 27 months, he was relieved of his major symptoms but continued to have moderate dysesthesia in the chest wall. The motor examination was normal. MRI performed at this time showed a significant regression of the intramedullary lipoma [Figure 2].

DISCUSSION

We recently presented a novel classification of atlantoaxial instability on the basis of facetal alignment and direct observation of instability by manual manipulation of bones of the region.[1] Sagittal CT scan and MRI and lateral profile plain radiographic images were observed with the head in the neutral position. Type 1 atlantoaxial instability was when the facet of the atlas was dislocated anterior to the facet of the axis. Type 2 instability was when the facet of the atlas was dislocated posterior to the facet of the axis. Type 3 atlantoaxial instability was when the facets were in alignment, but on the basis of corroborative clinical and radiological parameters, atlantoaxial instability was diagnosed by physical manipulation of bones of the region. Types 2 and 3 atlantoaxial facetal instability were identified as 'central' or "axial" atlantoaxial dislocation as there may not be any abnormal alteration in the atlantodental interval and there may not be any evidence of dural or neural compression at the craniovertebral junction by the odontoid process.[4] Central atlantoaxial instability is associated with long-standing or chronically unstable atlantoaxial joint more often identified in cases with basilar invagination, Chiari formation, and syringomyelia. Identification of central instability as a nodal point of the pathogenesis of a number of clinical entities can have a defining impact on their surgical management.[4-8] We identified that following atlantoaxial fixation, apart from recovery from neurological symptoms, there can be the reversal of secondary musculoskeletal changes and neural alterations such as short neck, torticollis, basilar invagination, Chiari formation, and syringomyelia.[5-9] In addition, the patient had C3-4 fusion. Our recent study identifies Klippel-Feil fusion as evidence of atlantoaxial instability. More often, instability in such cases is of chronic nature and is of central or axial type.[10]

It was recently identified that atlantoaxial fixation can be the treatment for "idiopathic" syringomyelia and for

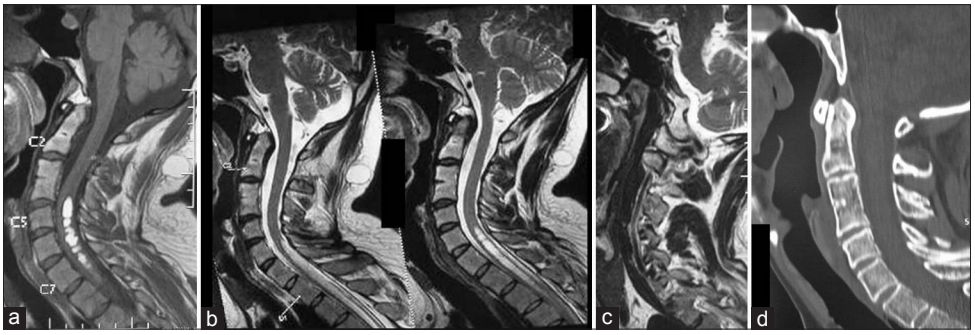

Figure 1: Preoperative images. (a) T1-weighted sagittal magnetic resonance imaging showing the hyperintense fat globulets occupying the central canal in the cervical region. (b) T2-weighted sagittal image showing the hyperintense fat streaks in the center of the cervical spinal cord and extending caudally. (c) Sagittal cut of magnetic resonance imaging passing through the facets showing the Goel Type 2 facetal instability. (d) Computed tomography scan showing C3-4 fusion. No abnormality in the atlantodental interval is seen

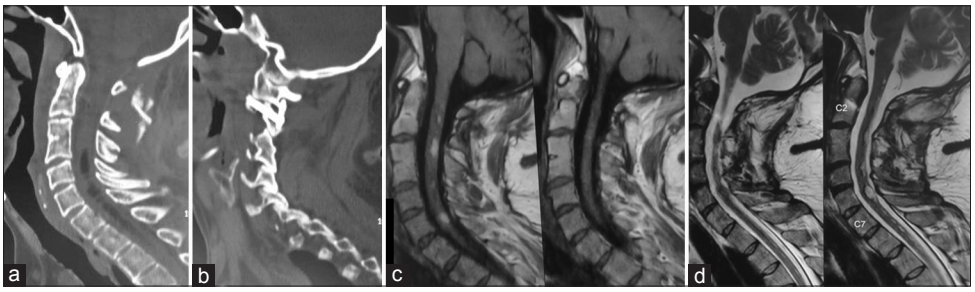

Figure 2: Postoperative images. (a) Postoperative computed tomography image. (b) Postoperative computed tomography image showing the fixation. (c) Delayed postoperative (after 27 months) T1 weighted image showing regression in the fat globulets in the central canal. (d) Delayed postoperative (after 27 months) T2 weighted image showing significant regression in the hyperintense signal in the central canal

syringomyelia associated with Chiari formation.[5,7,8] It was reported that following atlantoaxial fixation, the clinical symptoms recovered dramatically, and the syringomyelia cavity regressed in size in a significant number of cases.[5,7,8] Our current observation is that when the follow-up imaging is done after more than a year, the syringomyelia cavity regressed in 100 percent cases. Our studies identify syringomyelia as a secondary and probably "protective" entity, the nodal point of pathogenesis of which was atlantoaxial instability.

In the presented case, there was an intramedullary tumor, image characteristic of which favored lipoma. Considering its long segment and location in the center of the spinal cord, direct and safe surgical resection was impossible. The lipoma mimicked in its shape and extension a more common clinical entity of syringomyelia. Even though there was no associated Chiari formation, there was C3-4 fusion and a clear presence of Type 2 atlantoaxial facetal instability. Atlantoaxial fixation was done to treat atlantoaxial instability, and no physical alteration in the intramedullary lipoma was expected. The patient had a remarkable recovery from all his symptoms in the immediate postoperative phase. The reduction of lipoma on delayed postoperative imaging, similar to the reduction in the syringomyelic cavity, was a surprising observation.

Although on the basis of a single clinical report no definite conclusions can be made regarding the pathogenesis and the treatment, our observation suggests that both "fat" in the intramedullary lipoma like in the presented case and "water" in the syringomyelia cavity could have a similar origin and executed similar function.[11] More importantly, surgical management is similar.

CONCLUSION

The case presents hitherto unreported observation of intramedullary long segment lipoma that mimicked syringomyelia and was associated with atlantoaxial instability. Atlantoaxial stabilization resulted in clinical recovery and "regression" of lipoma.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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