of the C1/C2 facet capsules after rupture of the tectorial membrane and alar ligaments.

We cannot discuss the treatment of vertical AAD, as our patient died relatively early from cardio-vascular decompensation. The vertical AAD of 16 millimeters probably led to cardio-vascular decompensation from progressive oedema at the cervico-medullary junction or from progressive oedema in the posterior fossa due to dissection, thrombosis or spasm of the vertebral arteries. The other reported patient with vertical AAD survived with no spinal cord or brainstem injury. In this patient immediate external reduction by applying axial pressure on the head under fluoroscopy and secondary posterior occipito-cervical fixation were performed.[9] We therefore think that after establishing the diagnosis on plain radiographs and CT, immediate reduction by applying axial pressure on the head under fluoroscopy followed by internal fixation and fusion probably offers the best chances for survival with this pathology.

CONCLUSION

This is the second case of a purely traumatic vertical AAD without associated cranio-cervical junction pathology reported in the English literature. We hypothesize that after rupture of the tectorial membrane and the alar ligaments, the C1/C2 articular capsules become less resistant to axial distraction than the atlanto-occipital membranes. Due to the rarity of this pathology, no treatment guidelines can be established, but applying axial pressure on the head under fluoroscopy to reduce the AAD followed by internal fixation and fusion is appropriate.

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A case of presumptive primary lateral sclerosis with upper and lower motor neurone pathology

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Summary Motor Neurone Disease (MND) is one of the commonest neurodegenerative disorders of adulthood. MND characteristically presents with a combination of both upper and lower motor neurone features. Primary Lateral Sclerosis (PLS) is thought to be a variant of MND presenting with purely upper motor neurone signs. Debate continues over whether PLS constitutes a distinct pathological entity or whether it is part of the spectrum of motor neurone diseases that present as an upper motor neurone-predominant form of MND. We present a case of MND with purely upper motor neurone features and a prominent pain component. A pre-mortem diagnosis of PLS was made, however autopsy findings demonstrated both upper and lower motor neurone involvement. We believe these findings support the view that PLS is not a discrete pathological entity, but that it is a part of the range of motor neurone diseases that present with predominant but not exclusive upper motor neurone involvement. This case also highlights the feature that pain may be associated with MND even though it is not appreciated to have a sensory pathology.

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INTRODUCTION

Motor Neurone Disease (MND) or Amyotrophic Lateral Sclerosis (ALS) is one of the most common neurodegenerative disorders occurring in adult life.[1] The clinical hallmarks of this disease are a combination of both upper and lower motor neurone features including muscle wasting and weakness, fasciculations, spasticity, brisk reflexes and extensor plantar responses.[2] Although the diagnosis is essentially a clinical one, a variety of investigations are usually carried out to exclude other potentially reversible causes of such a presentation. Electrophysiological tests can be undertaken to help support the diagnosis of MND. In particular, electro-

myography should demonstrate denervation in at least three limbs to confirm the lower motor neurone abnormalities.[3]

In one sense, the term MND (or ALS) is used to describe the 'classic' presentation of the disease as described above, with the combination of upper and lower motor neurone features. Alternatively, MND has been used as an umbrella term to describe the range of degenerative motor neurone diseases. The spectrum of motor neurone diseases includes those that present with only upper motor neurone signs where the condition is referred to as primary lateral sclerosis (PLS), and those that present with pure lower motor neurone features, namely progressive spinal muscular atrophy.

Whether or not primary lateral sclerosis is a distinct entity (both clinically and pathologically) from MND or merely represents one end of the spectrum of the motor neurone diseases, has been a subject of controversy for many years. We present a case of MND that presented with pure upper motor neurone features clinically, but pathologically was consistent with a 'typical' MND picture of both upper and lower motor neurone degeneration. We believe that this case report and the post-mortem findings provide support for the view that PLS is not a discrete clinicopathological entity. It may be more appropriate to consider PLS as part of the range of degenerative motor neurone diseases that present predominantly but not exclusively with upper motor neurone involvement.

CASE REPORT

A 56 year old woman presented with an eight month history of progressive dysarthria. Her past medical history included hypertension and excision of a melanoma from her left leg five years earlier. There was no family history of neurodegenerative disease.

The initial neurological examination revealed a spastic dysarthria with an increased jaw jerk. There were no fasciculations or wasting seen in the tongue or limb musculature. There was hypertonia, more so on the left than right in the limbs. The upper limbs were notably more spastic than her lower limbs and her left arm was mildly weak. She was hyper-reflexic generally and a left Babinski response was present. Her gait was spastic. There were no cerebellar signs and sensation was intact.

Blood screens including a complete blood picture, electrolytes, liver and renal function was within normal limits. ESR was 10 mm/Hr and anti-neuronal antibodies were negative in CSF and serum. CSF examination demonstrated normal protein, glucose levels and no cells. MRI of the brain and spinal cord did not reveal any focal lesions or areas of increased signal. Nerve conduction studies were normal and there was no electromyographic evidence of denervation.

Over the next several months there was a gradual decline. Pseudobulbar features became more prominent with facial grimacing and crying. Initially swallowing was preserved, however she later became dysphagic. The degree of spasticity increased bilaterally with posturing of the left hand and foot and some associated cramps. This resulted in a deterioration of her gait with an increased number of falls. She used a walking stick to mobilise initially and then progressed to a walking frame. Oral baclofen was commenced for spasticity with limited success. She also described a burning sensation around her left elbow, left knee and both ankles. This responded partially to gabapentin. Upper motor neurone features worsened on examination with bilateral Babinski responses but there were no lower motor neurone signs. A repeat MRI scan of the brain and spinal cord performed approximately six months after the initial scan showed preferential volume loss in the region of the motor cortex and subtle high signal in the corticospinal tracts.

She was admitted to hospital for management of severe spasticity and pain nineteen months after her initial presentation. At this stage she had a spastic quadriparesis involving the legs more than arms and was unable to walk. She was anarthric and dysphagia was a significant management issue. Various aids to communication were implemented. Diet was modified to reduce aspiration risk but as swallowing deteriorated she required a percutaneous endoscopic gastrostomy (PEG) to maintain adequate nutrition.

Initial management of the spasticity and pain was with a subcutaneous patient-controlled analgesia (PCA) midazolam infusion. This was used for symptomatic control until the oral baclofen could be titrated up to adequate doses. Three weeks after admission there had only been limited improvement in spasticity and pain on baclofen therapy. Dantrolene was added to the regime with only minor success. She had trials of splints and botulinum toxin injections in an attempt to improve spasticity. The spasticity and pain were difficult to control. The oral baclofen dose was temporarily increased but was cut back after she developed confusion and drowsiness.

In addition to her general spasticity and pain, she also developed severe pain in the anal region. No local lesion was found but there was some spasm felt on rectal examination. Local anaesthetic and steroid creams were of limited benefit and antispasmodic agents gave little relief to this symptom.

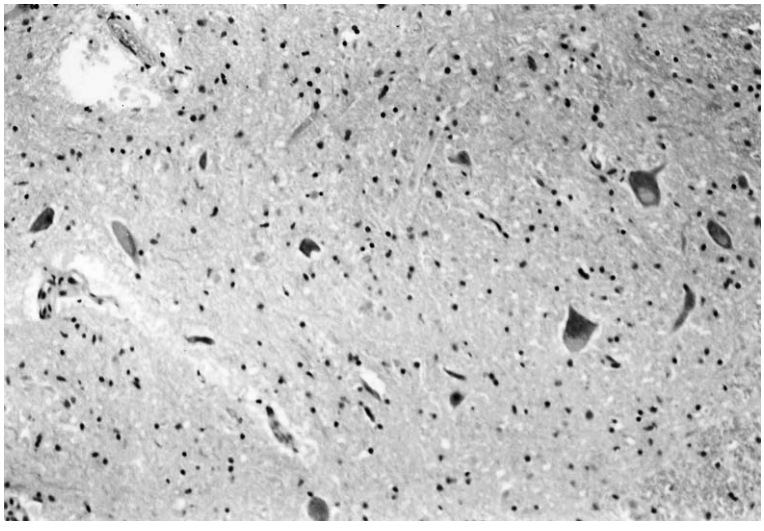

Fig. 1 H & E section of C5 spinal cord segment showing loss of anterior horn cells with only a few residual anterior horn cells (X250 original magnification).