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Spur on posterior arch of atlas with fatal spinal cord compression - a case report

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SUMMARY

The atlas vertebra has many developmental variations in its structure, mainly on its posterior arch with prevalence varying from 0.69% to 4%. It includes a spur on the anterior arch, a thickened anterior arch, posterior pointiculus and incomplete posterior arch among others. Some of the variations are associated with spinal stenosis, such as hypoplastic arch and ossification of the posterior longitudinal ligament. The anomalies are generally asymptomatic, but occasionally cause neck pain, and rarely myelopathy. In the case reported here, a spur was present in the midline on the anterior surface of the posterior arch of the atlas. This led to gradual compression of the cervical spinal cord with progressive quadriplegia, numbness and death. Myelopathy was confirmed by histopathology.

Key words: Atlas vertebra – Anterior arch – Posterior arch – Cranio-vertebral junction – Atlas spur – Spinal canal stenosis – Myelopathy

INTRODUCTION

The atlas vertebra is different from all other vertebrae. It has two lateral masses that are connected by anterior and posterior arches (Gupta et al., 2013). The body of atlas vertebra derives from the primitive fourth occipital and first cervical sclerotomes. Three or more ossification centres form the atlas. Usually, one midline centre builds the anterior arch and two ossification centres form the lateral masses. Ossification of the posterior arch starts at the lateral mass and proceeds perichodrically. Unification between the ossified atlas parts occurs anteriorly at left and right neuro-central synchondroses at five to eight years of age, and posteriorly at posterior synchondrosis at three to five years of age (Junewick et al., 2011; Guenkel et al., 2013). In addition, the posterior arch may have a supplementary ossification centre in the midline. Defective development of cartilaginous preformation or disturbance of ossification leads to developmental defect in the posterior arch (Guenkel et al., 2013; May et al., 2001).

Gupta et al. have evaluated various measurements for the atlas vertebra. They found that the maximum antero-posterior diameter of the spinal canal ranges from 2.7 cm to 3.9 cm (mean 3.04 cm). The maximum transverse diameter of the spinal canal ranges from 2.4 cm to 3.1 cm (mean 2.77 cm) (Gupta et al., 2013). Sherman et al. had measured normal the cervical spinal cord using MRI Imaging, where they found that the average antero-posterior diameter of spinal cord at C1 level is 9.3 mm, and the average transverse diameter is 11.3 mm with a standard deviation of 0.9 mm (Sherman et al., 1990).

Several variations of the atlas vertebra have been reported such as posterior pointiculus, V3 segment anomaly (VA), bifid atlas arch and incomplete posterior arch (Hong et al., 2008). Gupta et al. noted some variations in the anterior arch of the atlas, like a spur on it and thickening (Gupta et al., 2013). Due to the peculiar process of ossification, the posterior arch shows many developmental variations with prevalence varying from 0.69% to 4% (Guenkel et al., 2013; May et al., 2001). The commonest defect in the posterior arch is a cleft that

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may be either midline or on one or both sides (May et al., 2001). The anomalies are generally asymptomatic but occasionally cause neck pain. Some of the developmental anomalies are associated with spinal canal stenosis, such as hypoplastic arch, which may lead to myelopathy- degeneration of the spinal cord (May et al., 2001).

Here we present a case where a bony spur was present on the anterior surface of the posterior arch of the atlas, which led to a gradual compression of the cervical spinal cord in the individual.

CASE REPORT

A 31-year-old female from Northern India was having weakness and numbness in the left lower limb for 2-3 years, which gradually progressed to the left upper limb, then to the right lower limb and lastly to the right upper limb within a period of 6-8 months and became bedridden. Magnetic Resonance Imaging (MRI) of her cervical region revealed that the odontoid process was shifted superiorly and posteriorly with basilar invagination. The spinal canal was found narrowed at the level of C1 measuring 5 mm. Myelomalacial changes were present at C1 to C3 levels. She was referred to our hospital. Here, a Computerised Tomographic (CT) scan showed narrowing of the atlas ring. She was diagnosed to have bony canal stenosis at the cranio-vertebral junction. Laminectomy of the atlas was advised, but she was not operated upon. Her condition progressively worsened and after about four months she became unconscious at home, and was brought dead to the emergency department of our hospital.

Autopsy was performed, and during the dissection of the cervical column the cervical segment of the spinal cord was found antero-posteriorly compressed at the level of the atlas vertebra. The atlanto-axial complex was removed and the antero-posterior and transverse diameters of the spinal canal were measured with callipers. Antero-posterior and transverse diameters of the spinal cord at the level of the spur were 4.2 mm and 13.4 mm, respectively (Fig. 1). The antero-posterior diameter of the spinal canal was 5.8 mm at the level of the spur, and the transverse diameter of the spinal canal was 26 mm (Fig. 2). A bony spur was found on the anterior surface of the posterior arch of the atlas (Fig. 3). In addition, the left foramen transversarium for the vertebral artery in the lateral mass of the atlas was absent.

C1 segment of the spinal cord was dissected out of the spinal canal, fixed in 4% buffered paraformaldehyde and processed for paraffin embedding. The blocks were sectioned at 5 µm thickness, stained with haematoxylin and eosin, mounted with DPX and viewed under a motorized microscope (BX51 Olympus, Japan) attached to a CCD camera. The sections were analysed and photographed under low and high magnification. The sections revealed that in the case there was degeneration of the posterior grey column neurons and myelin degeneration in the posterior white fasciculi of the spinal cord (Figs. 3, 4).

DISCUSSION

The present case reported here had a bony spur on the anterior surface of the posterior arch of the atlas vertebra that caused gradual compression of the spinal cord and various clinical manifestations due to the ensuing myelopathy.

The normal sagittal diameter of the spinal canal

Fig. 1. Diameters of the spinal cord at the level of the spur.

varies from 16 mm to 25 mm, and that of the spinal cord from 10 mm to 12 mm at the level of atlas (May et al., 2001). The average antero-posterior diameter of the spinal cord is 9.3 mm, and the average transverse diameter is 11.3 mm, with standard deviation of 0.9 mm (Sherman et al., 1990). The spinal cord is at risk of compression when the vertebral canal diameter is less than 14 mm (May et al., 2001). In the present case, the sagittal diameter of the spinal canal at the level of the bony spur was 5.8 mm. With reduced spinal canal diameter, the spinal cord has been severely compressed and its sagittal diameter was reduced to 4.2 mm compared to an average diameter of 9.3 mm (Sherman et al., 1990). The MRI had shown a posteriorly displaced odontoid process of the axis vertebra. This would have further compressed the spinal cord at this level against the bony spur. This probably explains why the histological sections of the spinal cord showed extensive degeneration of the grey and white matter in the posterior aspect of the spinal cord. These visible changes also explain the patient's clinical condition prior to death — quadriplegia leading to being bed ridden and sensory loss. The cervical segment of the spinal cord contains sensory tracts from the entire body, including the head and neck region and motor tracts (corticospinal) for all four limbs (Standring, 2008). Compression at this level, hence, leads to quadriplegia and sensory loss in the associated areas.

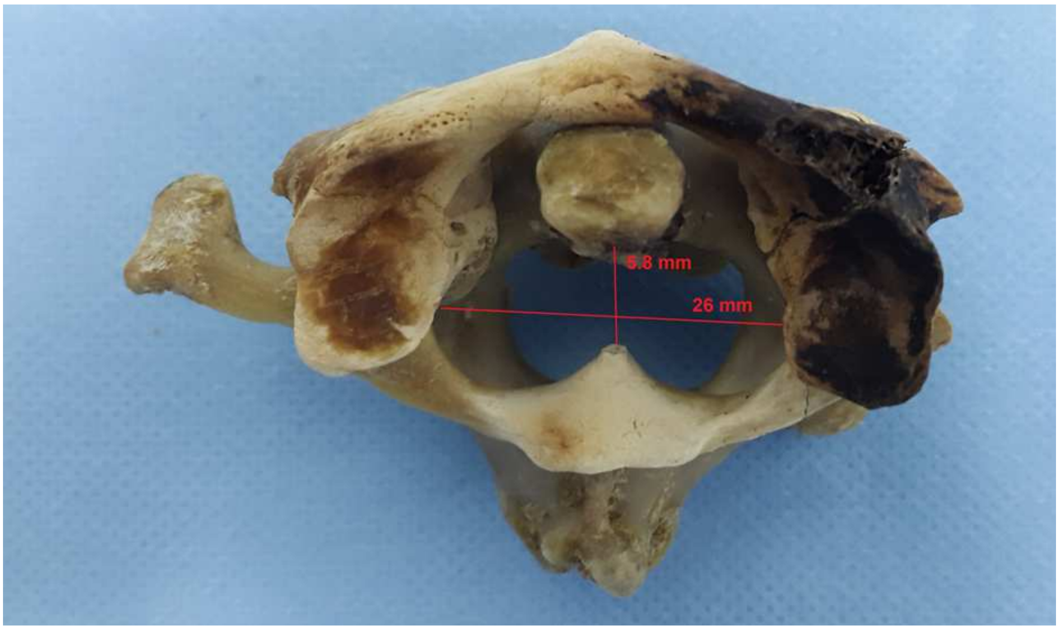

Fig. 2. Diameters of the spinal canal at the level of the spur.

Fig. 3. Atlas of the patient, showing the spur (arrow).

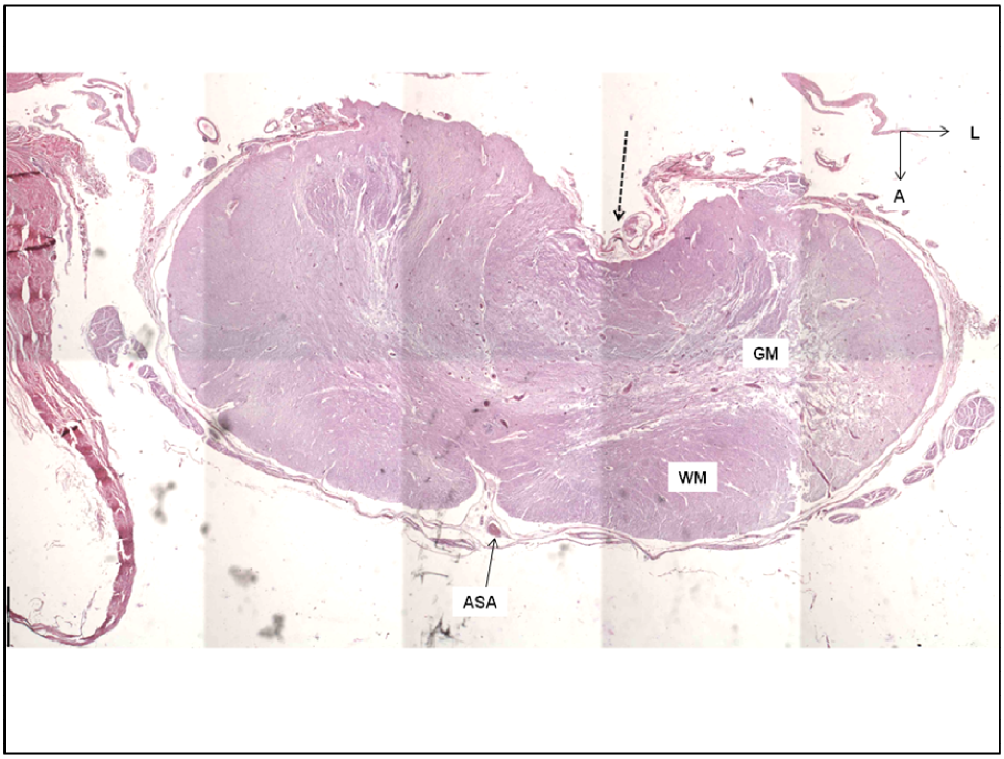

Fig. 4. Stitched photomicrograph of an H&E stained section of the spinal cord showing grey matter (GM) and white matter (WM). The anterior spinal artery is seen in the anterior median fissure and the dotted arrow indicates the area of compression on the posterior aspect of the spinal cord caused by the body spur on the posterior arch of the atlas. Scale bar: 1000 µm (left lower corner). Directions: A= Anterior; L= Left.

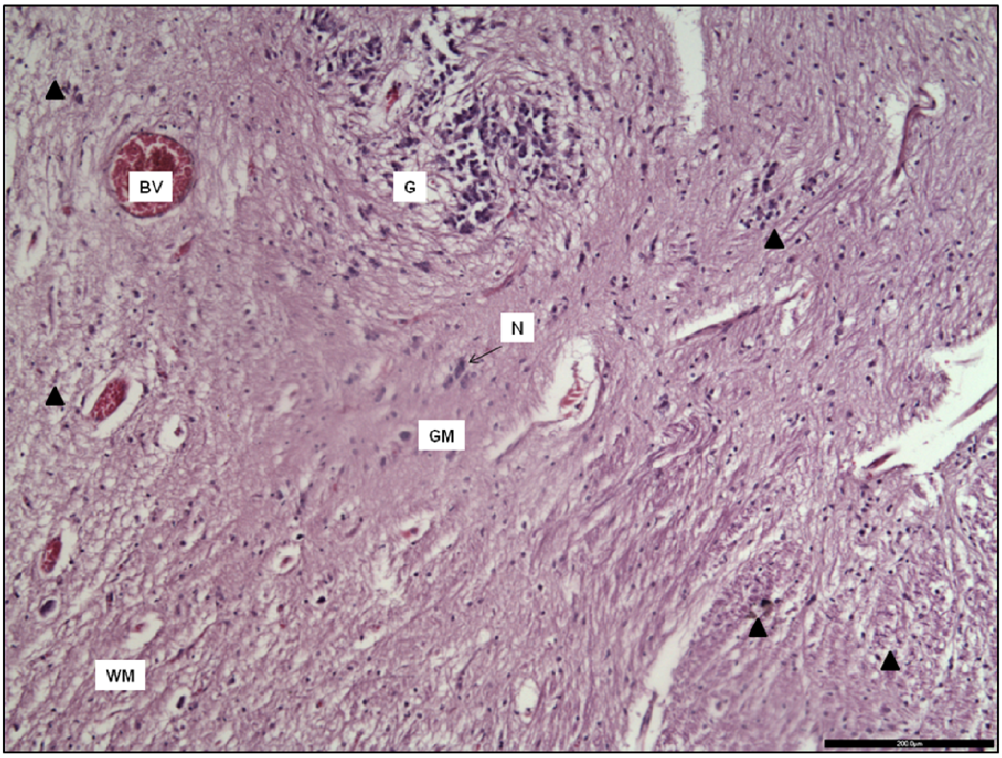

Fig. 5. Photomicrograph of an H&E stained section of the spinal cord showing gliosis (G) in the grey matter (GM). Neurons (N) are also apparent. The white matter (WM) also shows a large number of nerve fibre bundles showing degenerative changes. Blood vessels (BV) were also seen. Scale bar: 200 µm.

Harsh et al. had described 20 cases of cervical spinal stenosis where the posterior longitudinal ligament was calcified. These cases had a mean residual spinal canal diameter of 9.42 mm. All the cases had presented with steadily progressive cervical myeloradiculopathy (Harsh et al., 1987). The present case also had myelomalacia that was detected by MRI and histologically. This may have been due to the severe compression, since the available diameter of the spinal canal was only 5.8 mm at the level of the spur. There was splaying of the cord due to the constant pressure, the transverse diameter being increased to 13.4 mm compared to normal transverse diameter of 11.3 mm (Sherman et al., 1990).

The atlas vertebra has many variations in its structure, due to its peculiar ossification process as described. Common variations are posterior pointiculus, V3 segmental anomaly and bifid arch (Hong et al., 2008), and partial bilateral agenesis of the posterior arch with cleft or defect (May et al., 2001; Castaño-Duque et al., 1997). Some of the anomalies are associated with spinal stenosis, such as hypoplastic arch which decreases antero-posterior diameter of the spinal canal (May et al., 2001) and ossification of posterior longitudinal ligament (Harsh et al., 1987). The posterior arch particularly has more structural variations. This is often due to the peculiar ossification process, which starts from the lateral mass and proceeds to the posterior arch. In addition, it may have an independent centre of ossification in the midline (Guenkel et al., 2013; May et al., 2001). In this case, the posterior centre of ossification probably continued to grow as a traction epiphysis, and led to gradual compression of the spinal cord that manifest in the various neurological presentations and ultimately lead to the death of the patient.

Such findings may be difficult to pick up on routine and specialised imaging techniques, as happened in this case. Had the patient agreed to a posterior arch decompression, her condition may have improved. But as a word of caution this bony arch may have appeared as a surprising observation to the operating surgeon, and may have caused inadvertent damage to the already compressed spinal cord during surgery, especially if the bony spur had fragmented and displaced anteriorly.

The authors hope that the reporting of this case, which has not been reported before, would add to the knowledge regarding such debilitating anomalies of the atlas vertebra.

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