Unilateral atlantal lateral mass hypertrophy associated with atlanto-occipital fusion

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Abstract

Purpose Unilateral hypertrophy of the lateral mass of the atlas is an extremely rare condition. The authors present a rare type of unilateral atlantal mass hypertrophy with atlanto-occipital fusion which is associated with an invaginated lateral mass of the atlas and the odontoid process into the foramen magnum.

Methods A 45-year-old woman presented with a 2-year history of progressive bilateral weakness in the upper and lower extremities and gait disturbance. The left lateral mass of the atlas was hypertrophied and had invaginated into the foramen magnum with the odontoid. The spinal cord was severely compressed at the level of the foramen magnum, surrounded by the lateral mass of the atlas, the odontoid process and the occipital bone.

Results First, ventral decompression was performed using a transmandibular approach. The anterior arch of the atlas, the medial side of the hypertrophied lateral mass and the odontoid process were resected. Two weeks after primary surgery, posterior occipitocervical fusion was performed. The postoperative course of the patient was uneventful. Three years after the operation, she could walk without assistance and her paresthesia improved.

Conclusions To our knowledge, such a case of unilateral atlantal mass hypertrophy associated with atlanto-occipital fusion has not been described previously. The authors discuss the pathology of this case and review the literature

on unilateral atlantal mass hypertrophy and associated anomalies of the upper cervical spine.

Keywords Atlantal lateral mass hypertrophy · Atlanto-occipital fusion · Basilar invagination · Transmandibular approach

Introduction

Various bone and soft-tissue anomalies of the craniovertebral junction (CVJ) are rare conditions [1–6]. Unilateral hypertrophy of the lateral mass of the atlas is an extremely rare CVJ abnormality [7]. To the best of author's knowledge, only six cases of unilateral atlantal lateral mass hypertrophy have been reported in the literature [7–9]. We report on a rare type of unilateral atlantal mass hypertrophy with atlanto-occipital fusion which is associated with an invaginated lateral mass of the atlas and the odontoid process into the foramen magnum.

Case report

A 45-year-old woman presented with a 2-year history of progressive bilateral weakness in the upper and lower extremities and gait disturbance. She could not walk without assistance at admission. According to her family she had showed mild gait disturbance since childhood. There was no family history of musculoskeletal disease or history of trauma. When examined on admission, the patient's gait was spastic and she showed deep tendon hyperreflexia. Her Babinski reflex was positive. There was a grade 3–4/5 weakness in both arms and legs. She complained of clumsiness in both hands and paresthesia below

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Fig. 1 **a** This 3D-CT image shows an anterior view of the cervical spine. The atlas was rotated and its left lateral mass had prolapsed into the foramen magnum. **b, c** Sagittal and coronal 3D-CT images show invagination of the left hypertrophied lateral mass of the atlas (white

arrow) and the odontoid process. The left lateral mass of the atlas fuses with the occiput (dotted arrow). **d** An axial CT myelograph at the level of the foramen magnum. Spinal cord (dotted circle), the lateral mass of the atlas (LM), odontoid process (white arrow)

the C5 level. Her neck motion was restricted, particularly in rotation. No fasciculations or cranial nerve abnormalities were present. No bladder or bowel dysfunctions were noted. She had a relatively small build, being 140 cm tall and weighing 40 kg.

Three-dimensional computed tomography (3D-CT) imaging of the patient's cervical spine showed mild scoliosis (Fig. 1a). The left anterior arch and the left lateral mass of the atlas were assimilated into the occiput. The atlas was rotated at the occipito-atlantal joint. The left lateral mass of the atlas was hypertrophied and

had invaginated into the foramen magnum with the odontoid (Fig. 1b, c). The tip of the odontoid extended 7 mm above the Chamberlain line. There was C2–3 vertebral fusion. Magnetic resonance imaging and CT myelography revealed that the spinal cord was severely compressed at the level of the foramen magnum (Fig. 2), surrounded by the hypertrophied lateral mass of the atlas, the odontoid process and the posterior lip of the foramen magnum (Fig. 1d). No apparent cervical instability was found in plain radiographs for flexion and extension positions.

Fig. 2 This preoperative T2-weighted magnetic resonance imaging scan of the cervical spine shows severe compression of the spinal cord at the CVJ by the hypertrophied lateral mass of the atlas (*white arrow*)

Clinical course

First, ventral decompression was performed using a transmandibular approach under general anesthesia. Preoperative examination revealed that the patient had limited ability to open her mouth, and it was considered it would be difficult to achieve a wide operative view around the CVJ for a transoral approach without splitting the mandible. Therefore, the mandible was split in the middle using a bone saw. Then, the tongue and the caudal side of the uvula were sectioned in the midline. This approach enabled easy access to the CVJ from the clivus to the C3 vertebra. Using

Fig. 4 Plain X-ray after occipitocervical fusion. The divided mandible was fixed with a plate and screws

an operating microscope, the anterior aspect from the clivus to the base of the axis was exposed. The anterior arch of the atlas, the medial side of the hypertrophied lateral mass of the atlas and the odontoid were resected by a diamond burr. The caudal side of the clivus was also removed (Fig. 3a, b). Then, anterior decompression was achieved and pulsation of the dura was confirmed. A halo-vest was applied to prevent instability around the CVJ. Two weeks after the primary surgery, posterior occipitocervical fusion was performed. The occipital bone was fused to C4 using a plate and rod system (Fig. 4). The plate was fixed to the occipital bone by a cortical screw and fixed to C2–C4 using sublaminar polyethylene cables. Thereafter, the postoperative course of the patient was uneventful. Her bilateral arm and leg weakness improved gradually and no new neurological deficit was observed. Three years after the operation, occipitocervical bony fusion was achieved

Fig. 3 **a** 3D-CT image after anterior decompression surgery. **b** Sagittal CT image. The anterior arch of the atlas and the medial side of its left hypertrophied lateral mass and the odontoid process were removed

and the patient could walk without assistance and her paresthesia improved; however, mild spasticity remained in the lower extremities.

Discussion

Goel et al. [7] reported four cases of unilateral atlantal mass hypertrophy in 2006. They postulated that this condition is due to either a congenital abnormality or a slow-growing pathological entity. In our case, atlanto-occipital fusion which is associated with an invaginated lateral mass of the atlas and the odontoid process into the foramen magnum was present. Goel et al. [7–9] did not mention in the literature about atlanto-occipital fusion and basilar invagination. Therefore, our case is considered an extremely rare case of unilateral atlantal mass hypertrophy. The spinal cord was compressed severely by the hypertrophied lateral mass of the atlas and the odontoid process in the foramen magnum, causing severe myelopathy. Menezes et al. [10] reported similar cases of CVJ anomalies. They investigated 5,200 patients in their CVJ database and found 72 patients having symptomatic proatlax segmentation abnormalities. Proatlax segmentation abnormalities had been recorded in postmortem studies but very rarely in a clinical setting. In patients with proatlax segmentation abnormalities, various bony indentations arising from the clivus, the rim of foramen magnum and the atlas cause neural compression at CVJ lesion. They also stated that the majority of patients presented early in the first and second decades of life. However, age at onset of our patient was 45 years of age and age of patients reported by Goel et al. [7] averaged 36.5 years (ranged 21–54), which indicated slow-growing pathological entity and different pathology from proatlax segmentation abnormalities.

Atlanto-occipital fusion is defined as partial or complete congenital fusion of the occiput and the atlas [1, 4, 6]. Atlanto-occipital fusion is sometimes associated with basilar invagination, atlanto-axial instability, and vertebral fusion [1, 11–13]. Because of the decrease in the vertical height of the atlas caused by atlanto-occipital fusion, the odontoid may prolapse into the foramen magnum, resulting in “relative” basilar invagination [14]. Tubbs et al. [15] performed morphometric analysis of adult skulls with atlanto-occipital fusion and found that the area of the outlet of the foramen magnum was reduced by 15–35 %. Atlanto-occipital fusion is associated with anomalies around the CVJ that predispose to narrowing of the space available for the spinal cord or brainstem.

Gholive et al. [11] proposed a morphological classification of atlanto-occipital fusion according to the location of the fused area: zone 1, a fused anterior arch; zone 2, fused lateral masses; and zone 3, a fused posterior arch. They

found an association of basilar invagination, canal encroachment, and C2–3 fusion with atlanto-occipital fusion of zone 1 and/or 2. In their study, spinal canal encroachment occurred in 37 % of patients, and half of these patients had the clinical symptoms of myelopathy. In our patient, the atlas was fused with the occiput at zones 1 and 2, and both basilar invagination and C2–3 fusion were found. Goel et al. [7] also reported associated fusion of the C2–3 segment in a patient of unilateral atlantal lateral mass hypertrophy. Tubbs et al. [15] stated that fusion of the atlas based primarily along the anterior rim of the foramen magnum causes greater obstruction of its outlet. Gholive et al. [11] also reported that the highest prevalence of spinal canal encroachment (63 %) occurred in patients with fusion in zone 2. The pathology of our case can be explained as follows. There was congenital atlanto-occipital fusion with relative basilar invagination. The lateral mass of the atlas, which had prolapsed into the foramen magnum, had become hypertrophied with age and this caused canal encroachment into the foramen magnum. To our knowledge, this type of CVJ anomaly has not been described previously.

Surgical treatment of anomalies of the CVJ is challenging because of its location and complicated bony structure [1, 4, 5]. Goel et al. [7–9] stated that most of the patients of atlantal lateral mass hypertrophy could be treated by only posterior decompression, usually without fusion. However, atlanto-occipital fusion associated with basilar invagination was present in our case. Patients with atlanto-occipital fusion associated with basilar invagination occasionally undergo a trial of axial cervical traction to reduce the prolapsed odontoid process [1, 5, 16]. Among patients with a reduced odontoid process, basilar invagination can be treated with posterior decompression and occipito-atlantal stabilization [5, 16]. However, ventral decompression may be necessary for basilar invagination with ventrolateral neural compression that is not reducible, and is usually followed by posterior occipitocervical stabilization [5, 16, 17]. In our case, the patient was a 45-year-old woman with restricted range of motion of the neck. Because the CVJ was considered very tight, cervical traction could have been dangerous and was not applied. First, anterior decompressive surgery was performed because the hypertrophied lateral mass of the atlas and odontoid process was the main cause of the cervical myelopathy, which severely compressed the spinal cord from the ventral side. During the ventral decompression surgery, the odontoid and lateral mass of the atlas were removed, which might have destabilized the upper cervical spine. This indicates that occipitocervical fusion is necessary to prevent subsequent instability. If the patient is small with limited capacity to open the mouth, as in our case, the surgeon may have difficulty in exposing a wide view around the upper

cervical lesion by the transoropharyngeal approach. For such cases, several extensions of this approach have been described to increase the field of exposure, including splitting of the hard palate, splitting of the mandible, median tongue splitting and splitting of the mandible and maxilla [1, 5, 16].

In some cases of CVJ anomalies, orientation during surgery may be difficult with limited exposure of the operative field. Detailed preoperative assessment of the CVJ is essential for accurate surgery. CT imaging with coronal, sagittal and 3D reconstructions provides visualization of the complicated osseous anatomy.

We report on a rare type of unilateral atlantal mass hypertrophy with atlanto-occipital fusion which is associated with an invaginated lateral mass of the atlas and the odontoid process into the foramen magnum. To our knowledge, such a case of unilateral atlantal mass hypertrophy has not been described previously. The patient was successfully treated by anterior decompression followed by posterior occipitocervical fusion. Wide exposure around the CVJ and accurate decompression could be performed using a transmandibular approach.

Conflict of interest None.

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