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Case Report

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Inverted hypertrophy of occipital condyles associated with atlantooccipital fusion and basilar invagination: a case report

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Abstract

Atlantooccipital fusion is complete or partial congenital fusion of the atlas to the base of occiput. Basilar impression may complicate the clinical picture with the reduction of the vertical height of atlas causing odontoid or other bony structures project into foramen magnum and medulla oblongata. This results in narrowing of the foramen magnum and leads to neural compression. Authors present an unusual case in which the atlantooccipital fusion is complicated with the inverted hypertrophy of occipital condyles.

Key words: [basilar impression] [anomaly] [hypertrophy] [occipital bone] [condyle]

Introduction

Atlantooccipital fusion is one of the osseous anomalies of the craniovertebral junction. It is characterized by complete or partial fusion of the bony ring of the atlas to the base of occipital bone. The patients are commonly found to have dysplastic conditions such as achondroplasia, diastrophic dysplasia which may lead to occipitocervical instability. Twenty percent of the patients also have isolated congenital ossification abnormalities of jaw, nasal cartilage or hard plate [1–3]. Most of the patients have easily recognizable clinical picture such as torticollis, short neck, restricted neck motion, low hairline [1, 3].

In this case report, a patient with atlantooccipital fusion complicated with inverted hypertrophy of occipital condyles is presented. Inverted hypertrophy of occipital condyles reduce the transverse diameter of foramen magnum and cause brain stem and cerebellar compression. The inverted hypertrophy of occipital condyles has not been reported in the literature. The neuroradiological findings, clinical symptoms, and the surgical treatment are discussed.

Case Report

A 45-year-old woman was admitted to our hospital with complaints of occipital headache aggravated with neck movement. She had imbalance for 2 years and numbness in her left arm for 2 months which recently progressed to her left leg. On physical examination she had ataxia and decreased sensation of deep pressure, vibration and proprioception in her left side of the body. Thorough physical examination did not show any dysplastic condition.

Conventional and 3D computed tomography scans demonstrated the fusion of the posterior ring of atlas to the occiput and inverted hypertrophy of the occipital condyles, reducing the transverse diameters of foramen magnum (Figure 1). Magnetic resonance imaging (MRI) revealed the narrowed foramen magnum, ventral compression by impression of the odontoid process, posterior compression by the fused bone and signal intensity changes of the spinal cord at the level of craniocervical junction (Figure 2).

The patient underwent suboccipital craniectomy with removal of the posterior arch of the atlas along with

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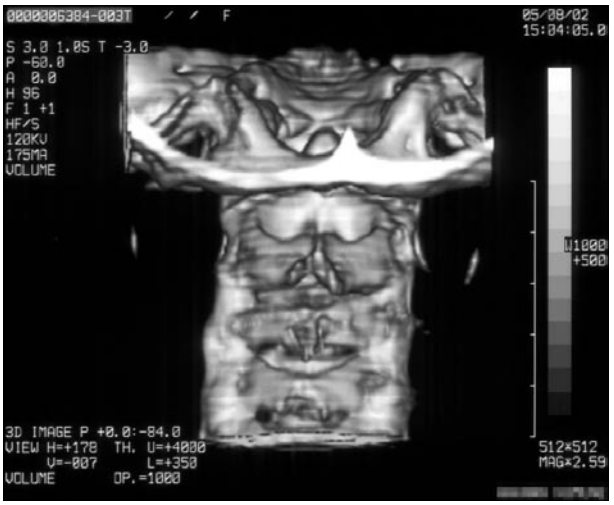

Figure 1. 3D-CT scan of the craniocervical junction showing hypertrophic bony parts are the continuity of the occipital condyles.

Figure 2. T2 weighted MR image of the cervicomedullary junction indicating the ischemia of spinal cord.

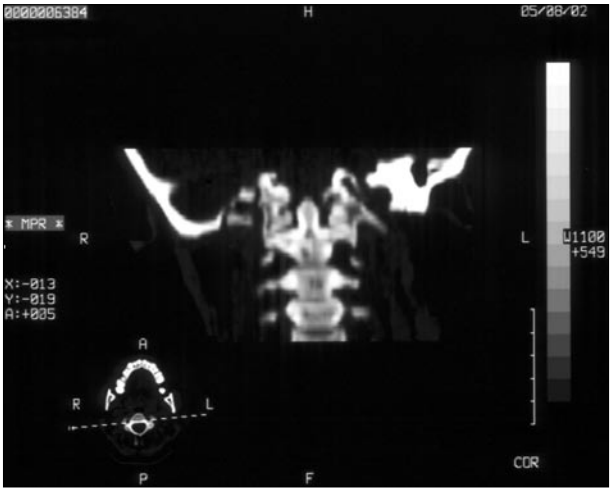

Figure 3. 3D-CT scan showing digastric line passes above the tip of the odontoid process.

releasing the posterior dural bands in the foramen magnum. The patient's postoperative course was uneventful. At 2 month follow up, she had less imbalance and headaches. Neurological examination showed improvement in ataxia and sensory deficits.

Discussion

Atlantooccipital fusion is developmental fusion of the skeleton of the atlas to the occipital bone. This is characterized by partial or complete fusion of the bony ring of the atlas to the base of the occipital bone. The associated clinical symptoms may have wide variation depending on the area of the compression on neuroanatomical structures and the secondary effects of vascular and CSF disruption. In patients with atlantooccipital fusion, the clinical findings suggest that the major neurological compression is due to the odontoid projecting into the foramen magnum. The symptoms and signs of pyramidal tract, anterior bulbar and cranial nerves involvement might be present [4,5]. Less common, if the compression occurs posteriorly by the posterior lip of the foramen magnum as seen in Figure 2, symptoms and signs are referable to involvement of the posterior column [4].

Projecting pain to the occipital region is the most frequent symptom and characterized as dull and aching, usually initiated by coughing or neck movements. Our patient had low hairline, short neck, and restricted neck movements that the patients with atlantooccipital fusion are commonly found to have same findings [4,5]. Nystagmus, ataxia and incorporation may accompany to the cerebellar tonsillar herniation. Symptoms referable to the vertebral artery compression such as dizziness, seizures, mental deterioration and syncope may occur alone or in combination with those of the spinal cord compression [4]. In our patient, beside the corporation of the posterior arch of atlas into occiput and the ischemia on cervicomedullary junction, majority of compression results from the upward extension of the hypertrophic occipital condyles (Figure 3). Although atlantooccipital fusion is a congenital condition, as in our case, many patients do not develop symptoms until the second or third decade of life [4]. This may be due to a gradually increasing degree of ligamentous laxity and instability with aging. The onset of clinical symptoms can be sudden and precipitated by relatively minor trauma, the commonest course is slowly progressive, but sudden onset or instant death has been reported [4]. It has been reported that approximately half of the patients with atlantooccipital fusion have relative basilar impression [4].

Craniocervical region CT shows bony continuity of the anterior arch of the atlas into occiput. The sagittal diameter of the foramen magnum is an important landmark in symptomatic patients. In our patient, this diameter was 25 mm that this measure is accepted abnormal when it is less than 30 mm [5]. Vega et al. also reported that range of transverse diameter of the foramen magnum was known as 28–40 mm [6]. This transverse diameter was 24 mm in our patient that was narrower than the normal range. Narrowing of the

foramen magnum results in brainstem compression. This compression is additionally complicate by inverted hypertrophied occipital condyle leading to further brainstem damage.

Craniocervical MRI is helpful to demonstrate the evidence of compression on neurological structures. On T2 weighted images of our patient, signal changing suggesting ischemia on cervicomedullary junction was seen (Figure 2).

In conclusion, the present case is an example of the atlantooccipital fusion and basilar impression complicated by presence of inverted hypertrophy of the occipital condyles. It is obviously observed that there is a continuity between the atlas and the occipital bone. It is also obvious that, the hypertrophic bony parts are the extensions of the occipital condyles. The significant measurements are the decreased sagittal and transverse diameters of the foramen magnum. In this case it was complicated with the inverted hypertrophy of occipital condyles, leaving smaller area for neural structures and causing damage to the neural elements. If the symptoms originate from cervicomedullar junction compression, then suboccipital craniectomy, excision of the posterior arch of the atlas, and removal of the dural band, if present, are suggested.

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