Resolution of retro-odontoid cyst in a patient with atlanto-occipital assimilation after occipitocervical fixation

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INTRODUCTION

Atlas occipitalization or atlanto-occipital assimilation (AOA) is one of the most common osseous congenital anomalies of the craniovertebral junction.1-4 Although most patients with atlas assimilation may be asymptomatic, some patients may experience myelopathy attributed to cervical cord impingement and occipitocervical instability. Those with progressive symptomatic segmental instability and/or increasing neurological deficits are candidates for occipitocervical reconstruction and fusion.

Here, we described a case in which AOA associated with a retro-odontoid cyst was successfully fused, resulting in spontaneous regression of the cyst.

CASE REPORT

An 80-year-old female was admitted with progressive gait disturbance, and weakness of the upper extremities of 6 months’ duration. She demonstrated a useless right hand, a spastic quadriparesis,
and diffuse hyperreflexia, accompanied by bilateral Babinski signs.

The dynamic X-rays showed AOA without atlantoaxial instability, while the sagittal MR demonstrated a retroodontoid cystic mass compressing the spinal cord [Figure 1a]. The 3D-CT documented assimilation of C1, left vertebral artery (VA) agenesis, and an anomalous right VA that entered the spinal canal/foramen magnum below the occipitalized C1 posterior arch [Figures 1b and 2].

**Operation**

An occiput–C2 fusion was performed. Screws were secured to the occipital bone while C2 pedicle screws were applied; fixation then consisted of connection of the two with bilateral rods; and the fusion construct then included placement of structural iliac bone grafts affixed with smaller screws [Figures 3 and 4].

**Postoperative course**

Within 3 postoperative months, she regained normal strength, and the follow-up MR revealed moderate regression of the retroodontoid cystic mass [Figure 3].

**DISCUSSION**

Congenital assimilation of the atlas (AOA) is caused by the failure of segmentation between the last occipital, and first cervical sclerotome during the early fetal development.\[8,9\] It is the most common anomaly of the craniocervical junction (0.8–3.6%) and occurs more commonly in males/females (5:1).\[1,3,10\] The four patterns of occipitalization follow (e.g., Zones 1, 2, and 3; the 4th is a combination of those zones) [Table 1].\[2\] Zone 2, as in our patient, involved fusion of the lateral processes (fused lateral masses).

**Symptomatic versus asymptomatic AOA**

AOA is often asymptomatic, except where occipitalization results in fixation of both atlanto-occipital joints that bear more than 50% of the flexion/extension motion.\[7\] As such, the first mobile segment between the skull and the spine becomes the C1–C2 junction; this can result in "over stretch" of transverse ligament.\[7\] Synovial cysts can also arise at the C1–C2 level reflecting joint instability.\[5,12\]

**Association with anomalous VA**

Patients with AOA may demonstrate an abnormal course of the VA involving its third segment. Our patient had such an anomaly of the right VA. Wang et al. reported four distinct types of VA anomalies associated with occipitalization of the atlas [Table 2].\[11\] For Type 2, the VA courses below the occipitalized C1 lateral mass and extends medially after it leaves the transverse foramen of the axis, thus entering the spinal canal/foramen magnum below the occipitalized C1.

| Table 1: Failure segmentation parts (Gholve et al.).\[2\] |
|--------------------------------------------------------|
| Segmentation                                           |
| Zone-1                                                 |
| Anterior arch                                          |
| Zone-2                                                 |
| Lateral arch                                           |
| Zone-3                                                 |
| Posterior arch                                         |
| Zone-4                                                 |
| combination of zones                                   |

Figure 1: (a) Sagittal T2-WI revealed retro-odontoid cystic mass compressing the spinal cord. (b) Posterior view of three-dimensional computed tomography clearly demonstrates an assimilation with agenesis of the left VA and right VA entering the spinal canal below the occipitalized C1 posterior arch [Figures 1b and 2].

Figure 2: Computed tomography demonstrates an occipito atlas assimilation.
posterior arch. As our patient had this Type 2 anomaly with a high-riding VA, C2 laminar screws were placed instead of C2 pedicle screws to avoid damaging the VA. To assess the course of the VA before this type of surgery, we recommend obtaining a preoperative cervical CTA.

**CONCLUSION**

Patients with AOA and a retro-odontoid cyst may be successfully managed with occipitocervical fixation without resection of the cyst. However, we would recommend preoperative computed tomography angiography to document whether the vertebral artery follows an anomalous course to avoid an intraoperative neurovascular injury.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent.

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**Conflicts of interest**

There are no conflicts of interest.

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