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CASE REPORT

Occipital Condyles Congenital Dislocation and Condylus Tertius

An Unstable Association Revealing a New Abnormality of the Craniocervical Junction

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Study Design. A case report.

Objective. To describe a unique craniocervical junction anomaly and its implications both on mobility and stability of the skull base.

Summary of Background Data. Congenital variations in the craniocervical junction (CVJ) are rare and frequently symptomless. Mild traumas may commonly rouse symptoms which help to unveil such anomalies through radiological investigations.

Methods. A 73-year-old woman developed a monoparesis of the right arm after a mild craniofacial trauma. Neurological examination revealed hyper-reflexia in the upper limbs, confirming the strength impairment in the right one. Radiology showed a post-traumatic bulbo-medullary contusion sustained by a unique and unstable association of the first occipital condyles congenital dislocation ever reported with a rare condylus tertius. The patient underwent posterior decompression and occipitocervical screw-rod fixation and fusion. Clinico-radiological follow-up highlighted a gradual recovery of the neurologic impairment and the posterior decompression with resolution of the spinal cord contusion.

Results. Although apparently stable the hyperostosis and the irregularly shaped condylar surfaces behind the 3-points mechanism of skull base support played a critical role in determining axial instability. The imbalance due to skull-cervical spine malpositioning may consequently trigger a vicious cycle of development of osteophytes leading to spinal cord narrowing with neurologic decline. A surgical strategy providing for posterior decompression and fixation satisfied the need to solve both bulbo-medullary constriction and skull base instability.

Conclusion. Clinical evidences about CVJ anomalies are lacking and symptoms, when present, tend to be vague. Although extremely rare clinicians should be aware of CVJ variations by engaging to improve their knowledge of imaging anatomy, embryology, CVJ basic craniometry and anatomic relationships. Studies on developmental control genes may offer future perspectives of early diagnosis and targeted treatments.

Key words: craniovertebral-junction, abnormalities, instability, condylus tertius, occipital condyles, dislocation, congenital.

Level of Evidence: 4

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The anomalies of craniovertebral junction (CVJ) arouse great interest, because the numerous variants of the skeletal elements can result in neural compression, vascular compromise, alteration of cerebrospinal fluid (CSF) dynamics and even death.[1–3] Among the rarest variations is the condylus tertius, occurring as a midline projection of the clivus located at the anterior margin of the foramen magnum. Sometimes expressed as a simple rounded tubercle, in more structured cases it receives the tip of the odontoid process as an articular facet.[4] Herein we describe the case of a posttraumatic discovery of an extremely developed condylus tertius associated with the first ever reported occipital condyles congenital dislocation.

CASE REPORT

A 73-year-old woman developed a sudden monoparesis of the right superior limb soon after a craniofacial domestic trauma. No loss of consciousness was reported. Neurological examination at admission revealed a strength impairment F 3/5 to the right arm with distal paresthesias, hyper-reflexia in the upper limbs and Babinski sign bilaterally. Patient referred no functional limitations in head movement, neither neck pain nor stiffness before the trauma. She did not declare a dystocic birth in her anamnesis, she never suffered from rheumatic diseases and no head nor cervical spine traumas were reported in her history such as to lead to prefacial trauma radiological investigations. Urgent head and neck CT scan showed a thickened clivus with a sort of threefold occipital condyles attached to its lower surface. The 3 condyles were, respectively, articulated 1 with the odontoid and 2 with the posterior arch of

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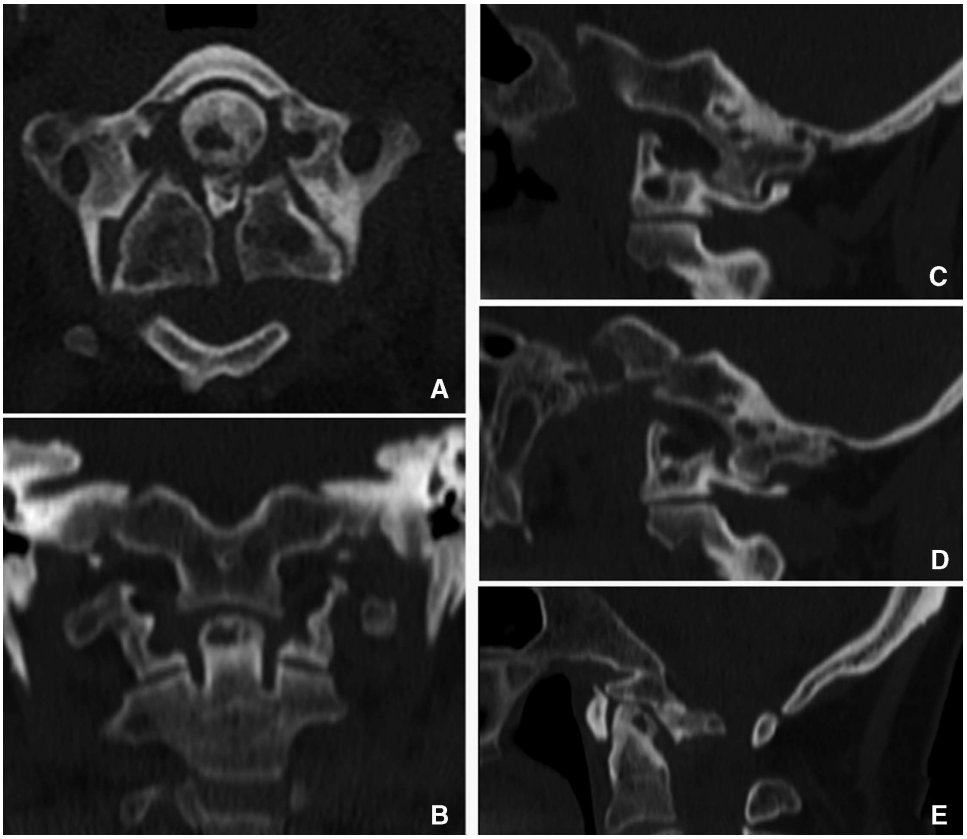

Figure 1. Preoperative CT-scan multiplanar reconstruction showing the spinal canal stenosis due to the posterior projection of the 3rd occipital condyle **(A)**, the relationship between the odontoid tip and the clival condyle on the frontal **(B)** and sagittal **(E)** plane, the connection between the left **(C)** and right **(D)** proper occipital condyles and the posterior arch of the atlas.

C1 (Figure 1A, B). The latter constituted 2 pseudo-diarthrosis with aberrant condyles sprouted where the clival hyperostosis reached the occipital bone and displaced postero-medially than the normal (Figure 1C–E). A MRI of the cervical spine outlined a severe bulbo-medullary compression mainly from the dorsal aspect, highlighting an altered T2 signal of the right hemimedulla compatible more with an acute contusion than with a chronic myelomalacia (Figure 2A). Therefore, the patient underwent C1 laminectomy to relieve the posterior compression and occipitocervical screw-rod fixation and fusion. The condylus tertius was left in place. There was no attempt at reduction intraoperatively, not being the case of an acute dislocation, but a stabilized cranio-cervical malpositioning. Postoperative CT scan documented a wide bulbo-medullary dorsal releasing. Five days after surgery a complete resolution of the paresthesias and a partial recovery of the strength in the right upper limb were noticed, so the patient was discharged. Six-months follow-up MRI demonstrated resolution of the bulbo-medullary contusion with posterior shift and relief from compression of the spinal cord (Figure 2 B). Starting from the third month follow-up the patient was assessed by the Benzel modified JOA scale revealing a performance improvement with an initial score of 14, increased to 16 at 6 months follow-up and firmly stabilized at 17 by the 9th month follow-up until now (21 months after surgery).

DISCUSSION

Congenital anomalies in the CVJ are rare and frequently symptomless. Such cases are often detected incidentally during the routine radiological clearance in traumatized patient. Through a schematic analysis the CVJ abnormalities can be classified into the following groups: assimilation of the atlas; anomalies of the atlas; manifestations of the occipital vertebra; platybasia and occipital dysplasia; degenerative manifestations and ossifications imitating variants.[5] From an embryological point of view, considering the presence of both the condylus tertius and the postero-medial dislocation of the occipital condyles, our case likely combine the characteristics of 2 subgroups in the context of the manifestations of occipital vertebra. The remarkable hyperossification may have supported the association of an incredibly structured third condyle with exuberant occipital condyles so postero-medially dislocated as to be far different from a classical post-traumatic posterior slippage but rather more comparable to a sort of postero-medial variation of processus condylicus (Table 1). Mostly occurring as a bony hunch at the frontal edge of the clivus, the third condyle may restrict or completely block rotational and flexion-extension movements of the skull.[6–8] In more developed cases a marked ossification, sustaining a 3-legged mechanism of head bearing, results in an occipital-atlanto-odontoid joint.[6,9,10] This articulated

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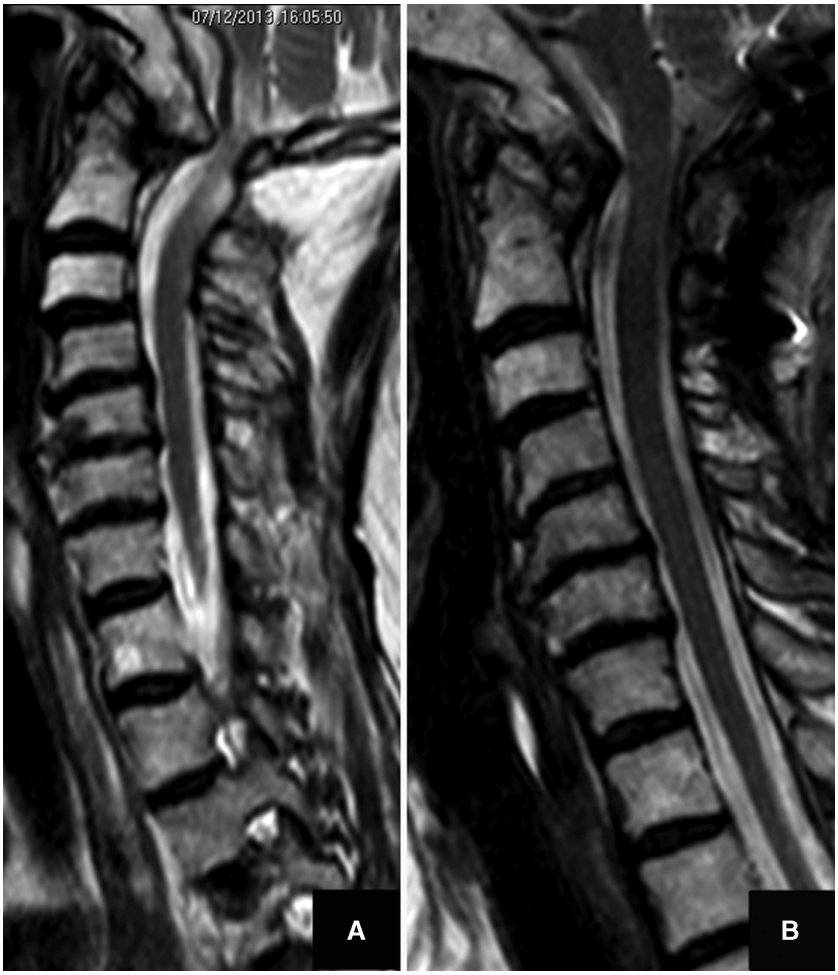

Figure 2. Preoperative **(A)** and 6 months follow-up **(B)** MRI demonstrating the posterior shift and relief from compression of the spinal cord.

connection could paradoxically play a role in the mechanisms of axial instability,[11] probably through its irregular cartilaginous interface and the secondary effects of the ossification on the ligamentous holding force.[6] In our patient, the osseous imbrication between the tip of the dens and the ossified clivus did not result in a proper diarthrosis; the particular bony overgrowth, completely enwrapping the odontoid process, made this case to be exemplary in showing how the roughly shaped condyle was conversely rather prone to produces an incongruent joint (pseudo-joint). In this sense, it is still difficult to assess whether a third median joint may strengthen the CVJ, or hamper the movements of the head to a greater or lesser degree or even significantly.[3] Although the circumferential medullary constriction, by the third condyle anteriorly and the occipital condyle dislocation postero-laterally, we felt appropriate only relieve the prevalent indentation, thus proceeding with posterior decompression and fusion.[12] The documented CVJ abnormality may deceive clinicians, dangerously hiding its prevailing biomechanical instability behind the features of the ossification-related rigidity and the threefold condylar support.[9] Morphologically the small, irregularly shaped occipital condyles appeared displaced posteriorly and closer than usual to each other, being articulated along the medial profile of the posterior arch of the atlas. The consequent imbalance due to skull-cervical spine malpositioning, coupled with the lack of stable articular joints, may exposes patients to a high risk of axial instability and sudden neurological manifestations, even incurring a minor head injury as is the case we reported. A neurological decline may alternatively occur, regardless of the trauma, as an effect of the progressive ossification usually associated with these kind of craniocervical variants.[7] The displaced occipital condyles and the overall shift in head's balance lead to a mechanical overload of the third condyle, causing a progressive growing of osteophytes gradually shrinking around the bulb and the spinal cord.[6] Therefore, patients may develop a progressive myelopathy or, as in our case, they can demonstrate apparently good neurological conditions, as long as even a mild trauma rouses a train of symptoms revealing an acute neurological impairment together with the CVJ abnormality. A surgical strategy providing for decompression and fixation should be taken in account especially in those cases showing a propensity to develop aberrant articular facets, to prevent, respectively, spinal cord constriction and skull base instability. The rarity of these anomalies, the broad spectrum of presentations and the large number of postmortem studies give insufficient data to perceive the real impact of these CVJ variations on clinical aspects and quality of life.

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TABLE 1. Manifestations of the Occipital Vertebra

Segmentations of the Basioccipital Bone	Bony Structures at the Foramen Magnum	Bony Structures Lateral Between the Occiput and Atlas	Bony Structures Medial Between the Occiput and Atlas	Variations of the Atlas	Variations of the Axis
Transverse fissures of the basilar process of the occipital bone (Sausers fissure)	Condylus tertius (fixed or isolated)	Tuberculum paracondylicum	*Postero-medial processus condylicus/ occipital condyles postero-medial dislocation**	Ponticulus atlantis posterior	Ossiculum terminale Bergmann persistens
Canalis n. hypoglossi bipartitus	Processus basilaris (fixed or isolated)	Processus paracondylicus		Ponticulus atlantis lateralis	Orthotopic, so-called os odontoideum
	Arcus praebasiocipitalis (fixed or isolated)	Massa paracondylica		Ponticulus atlantis posterolateralis	Real os odontoideum
	Dystopic, so-called os odontoideum (fixed or isolated)	Processus epitransversus (fixed or isolated)		Facies articularis superior bipartita atlantis	
	Labia foraminis magni posteriora				
	Processus condylicus posterior				

**The new CVJ anomaly discovered in our case is shown in italics.*

CONCLUSION

For those not accustomed to such complex CVJ anomalies, our case highlights the need to become reacquainted with CVJ basic craniometry, anatomic relationship, and pathologic conditions as well as with the embryology behind the different congenital abnormalities affecting this region.[8,10] New studies focused on the role of developmental control genes and their regulatory function may offer future perspectives of early diagnosis and targeted treatments before neurological damage occurs.[2]

➢ Key Points

- ❑ Occipital condyles congenital dislocation is an unstable CVJ anomaly never reported before in literature.
- ❑ Although apparently stable the hyperostosis and the irregularly shaped condylar surfaces behind the threefold condylar support played a critical role in determining axial instability.
- ❑ Incongruent condylar joints concurred in bone overgrowth further narrowing the spinal cord.
- ❑ Surgery providing for posterior decompression and fixation was essential to solve both bulbomedullary constriction and skull base instability.

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