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CASE REPORT

Odontoid process and clival regeneration with Chiari malformation worsening after transoral decompression: an unexpected and previously unreported cause of "accordion phenomenon"

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Abstract
Purpose Transoral odontoidectomy followed by occipito-cervical fixation is a widely used approach to relieve ventral compressions at the craniovertebral junction (CVJ). Despite the large amount of literature on this approach and its complications, no previous reports of odontoid process and clival regeneration following transoral odontoidectomy are present in the English literature.
Methods We report the case of odontoid process and clival regeneration following transoral odontoidectomy.
Results A 7-year-old boy presented with symptoms of brainstem and upper cervical spinal cord compression due to a complex malformation at the CVJ including a basilar invagination with Chiari malformation. A successful transoral microsurgical endoscopic-assisted odontoidectomy extended to the clivus was performed along with occipito cervical instrumentation and fusion. Clinical and radiological resolution of the CVJ compression was evident up to 2 years post-op, when the child had a relapse of some of the presenting symptoms and the follow-up CT and MRI scans showed a quite complete regrowth of the odontoid process, clival partial regeneration and recurrence of pre-operative Chiari malformation.
Conclusions Besides the need of an accurate complete resection of the periosteum, which apparently was incompletely performed in our case, our experience suggests the need of resection of the odontoid down to the dentocentral synchondrosis and an accurate lateral removal of the bone surrounding the anterior tubercle of the Clivus is advised when an anterior CVJ decompression is required in children presenting a still evident synchondrosis at neuroradiological investigation.

Keywords Basilar invagination · Bone regrowth · Chiari malformation · Clivus · Odontoid process · Transoral odontoidectomy

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Introduction

Transoral odontoidectomy is a well described and largely used approach directed to relieve irreducible ventral brain stem compressions at the craniovertebral junction (CVJ). A CVJ instability usually follows odontoid resections and a posterior fixation is generally required, both in the same operative session or after a settling period in a halo vest [1–3]. Recent advances in fixation materials may allow an anterior internal plate fixation [4, 5]. Basilar invagination forms a prominent component of the CVJ anomalies and Chiari malformation is the most common associate due to the small volume of the posterior cranial fossa [6].

We report the case of a 7-year-old boy with a basilar invagination and Chiari malformation who underwent transoral odontoidectomy and posterior fixation with immediate, radiologically confirmed, successful decompression but late quite complete symptomatic regrowth of the odontoid process, clival partial regeneration and recurrence of preoperative Chiari malformation. No other similar cases of

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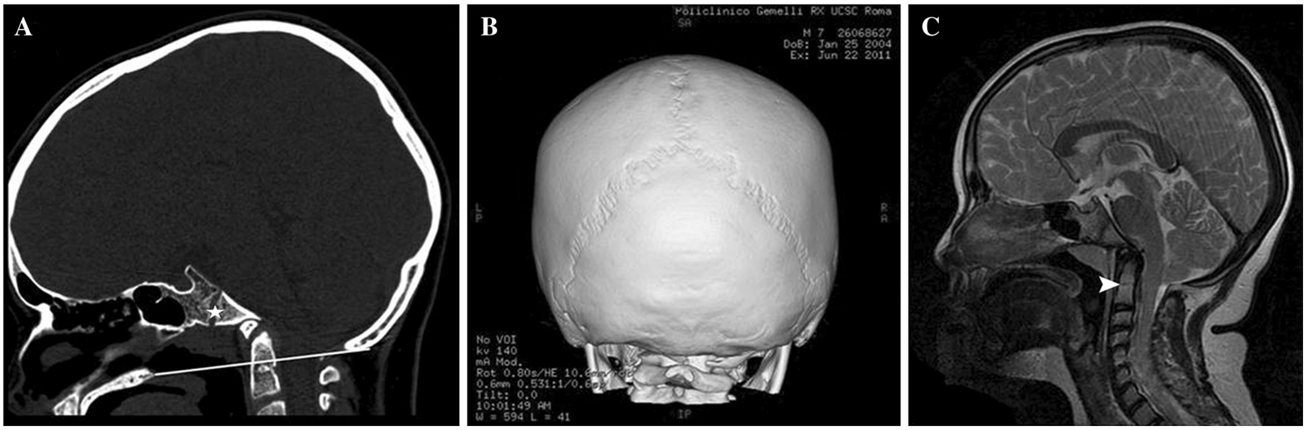

Fig. 1 Preoperative neuroimaging. **a** Sagittal CT showing platybasia and basilar invagination (McGregor's *line* is showed). **b** 3D CT showing partial assimilation of the anterior arch of the atlas with counterclockwise rotation on C2 and partial aplasia of the posterior arch. **c** Sagittal T2w MR showing the anterior compression of the brain stem and of the upper cervical spinal cord and the Chiari malformation. *Arrow* dentocentral synchondrosis. *Star* spheno-occipital synchondrosis

transoral odontoidectomy failure were previously reported in the English literature.

Case report

A 7-year-old boy presented with headache, diplopia, dizziness, episodes of fluid dysphagia, photo- and phonophobia, nuchal rigidity, weakness at the left arm and at lower limbs, paresthesias in left arm, cold dysesthesias in both upper and lower limbs, and pain at mastication. Brain and cervical spine MRI and CT scans showed a complex malformation at the CVJ: platybasia, basilar invagination, partial assimilation of the anterior arch of the atlas with counterclockwise rotation on C2, partial aplasia of the posterior arch of the atlas (bifidus) and Chiari malformation with compression of the brain stem and of the upper cervical spinal cord (Fig. 1). Brainstem auditory-evoked potentials were normal, while SEPs showed an abnormal central conduction pattern at lower limbs. Thus, a transoral microsurgical endoscopic-assisted decompression was performed according to our previous experiences; although the feeling of the surgeon was that the entire periosteum was removed along with the odontoid "more solito", postoperative MR showed some findings consistent with possible residual periosteum (Fig. 2b) [7, 8].

After 7 days in Halo Vest, the child underwent an occipito-cervical posterior fixation. The perioperative period was uneventful and the child had a rapid resolution of the preoperative symptoms. Early postoperative neuroimaging showed a satisfactory global decompression of the CVJ (Fig. 2). These clinical and radiological findings were stable at 3, 6 and 12 months follow-up (Fig. 3). Surprisingly at 2 years follow-up headache and sensory-motor deficits at lower limbs relapsed; neuroradiological investigations (CT and MR) showed a quite complete regeneration of the odontoid process along with a partial regrowth of the inferior portion of the clivus and recurrence of Chiari malformation, with a symptomatic compression of the brainstem and upper cervical spinal cord (Fig. 4). The patient was, therefore, scheduled for a redo surgery consisting of a further decompression with a more homogeneous caudal resection of the odontoid down to the dentocentral synchondrosis and an accurate lateral removal of the bone surrounding the anterior tubercle of the Clivus and complete removal of the periosteum. The parents refused a new operation. The clinical status was stabilised and unchanged up to date.

Discussion

Transoral odontoidectomy is a worldwide diffused surgical procedure and large case series of both adult and paediatric population harbouring basilar invagination have been reported in the last decades. This approach provides access to midline pathologies at the lower clivus, anterior arch of C1, odontoid process of C2, down to the C3 vertebral body in some patients [9–11]. A number of technical variations including "extended approaches", endoscopic assisted or pure endoscopic transoral and transnasal approaches have been described to extend the surgical domain of the approach up to the Clivus [8, 9, 12–17]. Classically, complications related to the transoral surgical odontoidectomy are immediate (CSF leakage, carotid or vertebral artery injuries, soft tissues oedema and cranial nerve injuries) and late (infections, wound dehiscence, CSF leak and meningitis, velopalatine incompetence, nasal speech, dysphagia and regurgitation of liquids and CVJ instability) [9, 17–20].

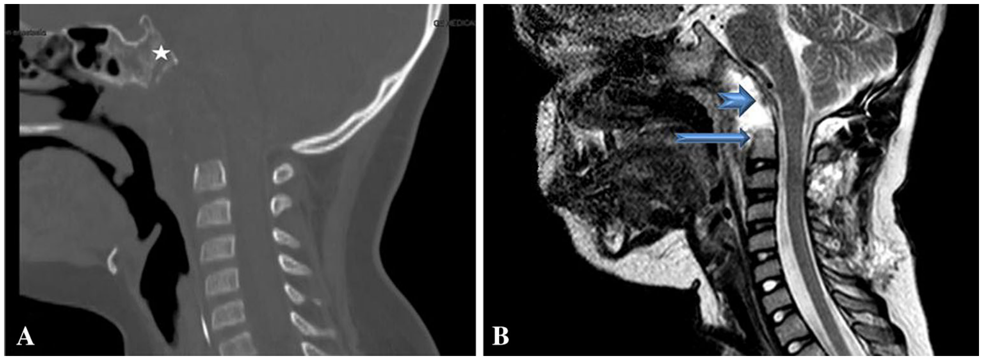

Fig. 2 Early postoperative neuroimaging. **a** Sagittal *midline* CT showing an extensive bone removal (inferior and middle third of the Clivus, anterior arch of the atlas and all the odontoid) at the anterior aspect of the CVJ with some middle third of the Clivus, the spheno-occipital synchondrosis (*star*). **b** Sagittal paramedian T2w MR showing an optimal decompression of the neural structures and the dentocentral synchondrosis left intact (*thin arrow*). Note the imaging consistent with residual periosteum recognisable on the anterior aspect of the tectorial membrane (*thick arrow*)

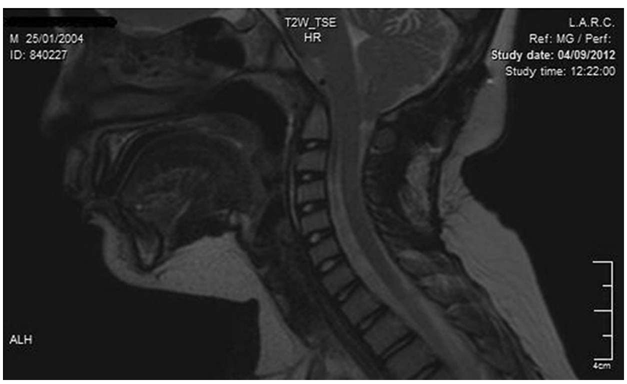

Fig. 3 1-year follow-up. Sagittal T2w MR showing a stable decompression of the anterior CVJ. Note that the resected bone edges have a more rounded aspect when compared with early postoperative imaging and Chiari malformation is lightly worsening

In 1998, Goel classified basilar invagination in two groups:

Group 1: the angle of the clivus and posterior cranial fossa volume are unaffected in presence of atlantoaxial dislocation.

Group 2: the assembly of the odontoid process the anterior arch of the atlas and the clivus is migrated superiorly in unison, resulting in reduction of posterior cranial fossa volume without atlantoaxial dislocation [6].

In 2009, we published our experience on the “always posterior strategy”; such a philosophy avoid transoral surgery as first step in view of a possible CVJ decompression by simple surgical manoeuvres during posterior fixation; nevertheless in the absence of CVJ dislocation it was not indicated in this case [21].

Since our patient was harbouring, Group 2 irreducible basilar invagination transoral decompression was performed and, by the simple transoral approach without extended variants, it was possible to remove the anterior arch of the Atlas, the odontoid and the medial aspect of the middle and inferior third of the clivus (Fig. 2a).

So far spontaneous odontoid process regrowth after transoral odontoidectomy has never been reported in the English literature as possible complication or evenience procedure related. In our case, the bone regrowth caused a renewed compression of the brain stem/upper cervical cord, surprisingly associated with the worsening of the preoperative Chiari malformation, improved soon after the operation, with a recurrence of some of the preoperative symptoms.

Bone regeneration at the foramen magnum has been previously reported after suboccipital craniectomies with or without C1 laminectomy in Chiari malformations [22–25, 32].

Complete spontaneous bone regeneration has been occasionally described in other craniofacial bones such as in mandible after partial mandibulectomy even in elderly patients [26–30].

Several conditions have been suggested to possibly influence this rare bone regeneration: young age; infections/inflammations; the development of new bone from intact periosteum or its fragments and from regenerated juvenile periosteum under the influence of a local environment which is permissive to osteogenesis due to the presence of growth factors as the VEGF; post-surgical immobilisation or even continuous functional stresses on the regenerating area [26–29, 31]. However, the mandible, as well as the majority of the skull and facial bones such as maxilla, premaxilla, zygoma, frontal parietal, vomer, palatine, and nasal bones, develops by intramembranous ossification [32], while the bony cranial base develops by endochondral ossification [33].

At spinal level some degree of bone regeneration has been observed after lumbar laminectomies for several degenerative conditions [34, 35].

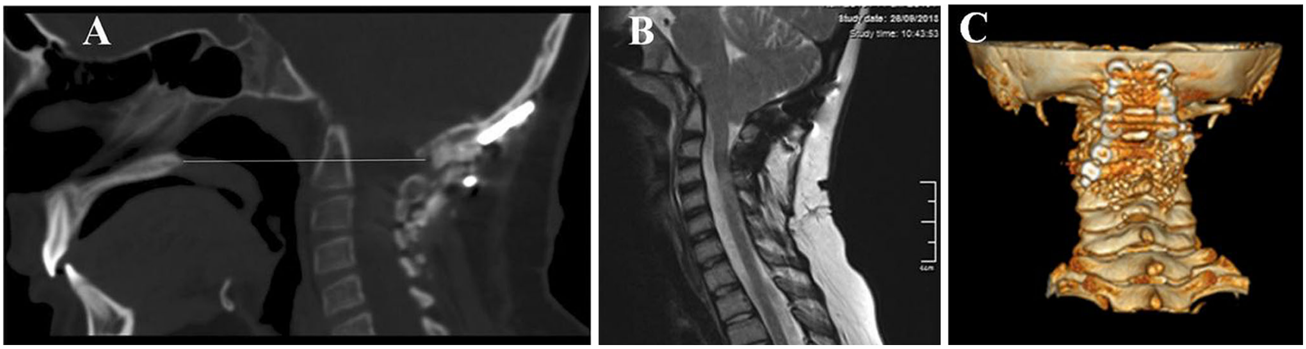

Fig. 4 2-year follow-up. **a** Sagittal CT scan showing the posterior construct with bone fusion, the spontaneous odontoid process regeneration with relapsing basilar invagination (note the McGregor's *line*) and the inferior clival regeneration. **b** Sagittal T2w MRI showing the compressive effect of CVJ on the neural structures. **c** Posterior 3D reconstruction showing a good craniocervical bone bridging

The Axis embryologically develops from four bones separated by synchondrotical articulations consisting of four ossification centres: two in the neural arches bilaterally (neurocentral synchondrosis), one in the vertebral body (dentocentral synchondrosis) and one in the odontoid process (apicodental synchondrosis) [36]. These ossification centres fuse between 3 and 7 years of age, but rarely they can remain open into adolescence [36–38]. In our case, the odontoid resection did not include the dentocentral synchondrosis, which is generally well below the superior articulating facets that constitutes the anatomical base of the odontoid process growth (Fig. 2). The ossification potential of this growing centre in a 7-year-old boy could explain the unusual odontoid regrowth associated with the Chiari worsening as the consequence of posterior cranial fossa impingement. Similarly, the development of the clivus, whose anterior tubercle develops from the hypocentrum of the fourth occipital sclerotome (proatlas), is determined by sutural growth at the level of the spheno-occipital synchondrosis and, laterally, at the level of the petro-occipital and spheno-petrosal junctions [39]. The spheno-occipital synchondrosis closes between 11 and 17 years of age, this process generally happens in girls before than in boys [40, 41]. Again, in the case here presented the spheno-occipital synchondrosis was not included in the bony resection and a quote of clival regrowth could be noticed at 2 years radiological follow-up (Fig. 4).

Moreover the inferior Clivus resection, clearly evident in the early midline CT reconstructions, was not enough laterally extended to the anterior tubercle of the Clivus, suggesting a possible lateral to medial reossification. We defined the phenomenon described as an "accordion" phenomenon. Beside the need of an accurate complete resection of the periosteum, our experience suggests to aim at a more homogeneous caudal resection of the odontoid down to the dentocentral synchondrosis and an accurate lateral removal of the bone surrounding the anterior tubercle of the Clivus when an anterior CVJ decompression is required in children presenting a still evident synchondrosis at neuroradiological investigation.

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Conflict of interest None of the authors has any potential conflict of interest.

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