Case report

Transnasal endoscopic removal of malformation of the odontoid process in a patient with type I Arnold-Chiari malformation: a case report

Exeresi per via transnasale endoscopica di malformazione del processo odontoideo in un paziente affetto da malformazione di Arnold-Chiari tipo 1: relazione di un caso

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SUMMARY

The endoscopic endonasal approach is emerging as a feasible alternative to the trans-oral route for the resection of the odontoid process, when the latter produces a compression of the brainstem and cervicomedullary junction. This type of approach has some advantages, such as excellent pre-vertebral exposure of the cranio-vertebral junction in patients with small oral cavities and the possibility to avoid the use of mouth retractors. A typical case of a 24-year-old male patient with a previous diagnosis of type I Arnold-Chiari Malformation, suffering from a posterior dislocation of the odontoid process causing severe anterior compression of the brainstem, is presented to stress the potential of this technique. Trans-nasal endoscopic removal of the odontoid process was performed and resolution of the ventral compression of the brainstem was achieved. This report demonstrates that in selected cases, an endoscopic endonasal approach should now be considered an excellent alternative to the traditional trans-oral approach.

KEY WORDS: Transnasal endoscopic approach • Odontoidectomy • Arnold-Chiari malformation • Brainstem compression

RIASSUNTO

L'approccio transnasale endoscopico sta emergendo come una possibile alternativa alla via transorale per la resezione del processo odontoideo, quando quest'ultimo determina una compressione del tronco encefalico e della giunzione craniovertebrale. Questo tipo di approccio presenta alcuni vantaggi, come un'eccellente esposizione prevertebrale della giunzione cervicomidollare in pazienti con piccole cavità orali, e la possibilità di evitare l'uso di retrattori buccali. Viene presentato il caso di un paziente di 24 anni affetto da malformazione di Arnold-Chiari di tipo 1, con dislocazione posteriore del processo odontoideo determinante severa compressione ventrale del tronco encefalico, al fine di sottolineare i momenti critici di questo tipo di approccio. È stata effettuata una resezione nasale endoscopica del processo odontoideo ed abbiamo ottenuto la risoluzione della compressione ventrale del tronco encefalico. Il caso clinico dimostra che, in casi selezionati, l'approccio nasale endoscopico deve essere oggi considerato un'alternativa eccellente a quello transorale tradizionale.

PAROLE CHIAVE: *Approccio transnasale endoscopico • Odontoidectomia • Malformazione di Arnold-Chiari • Compressione del tronco encefalico*

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Introduction

Type I Arnold-Chiari malformation (ACM-I) represents one of the most common pathological entities encountered by paediatric neurosurgeons and consists of a congenital malformation of the posterior cranial fossa causing herniation of the cerebellum and brainstem in the spinal canal through the foramen magnum. In a minority of cases, ACM-I is complicated by irreducible ventral compressive pathological features, such as basilar invagination or a severely retroflexed odontoid process [1]. In the presence of ventral brainstem compression, the most widely used anterior approach to the cervicomedullary junction via the atlantoaxial region is the trans-oral route [2 3], but this approach can prove difficult for a number of reasons: 1) a deep and narrow surgical field depending on the dimensions of the oral cavity of the patient; 2) the possibility of splitting of the soft/hard palate in the case of lesions located caudally to the palatal plane, causing a possible deglutition dysfunction; 3) the possibility of prolonged post-operative intubation due to massive oedema caused

by oral retraction. Nevertheless, in recent years, based on a dissection study by Alfieri et al. [4], Kassam et al. [5] have developed a new anterior trans-nasal approach for resection of the odontoid process in the case of severe brainstem and cervicomedullary junction compression in adult patients. Herein, our personal experience is reported in a case of transnasal endoscopic removal of the odontoid process in a 24-year-old male affected by ACM-I with severe basilar and brainstem compression.

Case report

A 24-year-old male patient with a previous diagnosis of ACM-I presented at our service, reporting, over the previous 3 years, severe head and neck pain localized in the occipital and vertex region associated with a reduction in motor power at the lower extremities and diplopia. One year previously, he had undergone atlo-suboccipital medial craniectomy with partial removal of the cerebellar tonsils and application of a dural plastic patch for cervicomedullary junction decompression. The patient reported a partial benefit after surgery. Pre-operative CT-scan revealed a partial fusion of the anterior portion of C1 and C2 with the cranial base and a posterior dislocation of the odontoid process causing a severe anterior compression of the brainstem; the alterations at the cervico-medullary junction were confirmed with MRI, where a partial compression of the vertebral artery could be seen and no evidence of syringobulbia and syringomyelia was present (Figs. 1A-B).

It was decided to perform a transnasal endoscopic removal of the odontoid process; a 0° degree endoscope with an irrigated sheath was introduced into the right nasal fossa and the common landmarks (inferior, middle turbinate and nasal septum) were visualized; posteriorly the choana and rhinopharynx, cranially the anterior wall of the sphenoid sinus, caudally the soft palate and laterally the ostium of the Eustachian tubes were recognized, to correctly delimit the surgical field (Fig. 2). To guarantee a better and wide angle view, it was decided to remove the rostral portion of the nasal septum, and two flexible catheters were introduced into both nostrils and withdrawn from the oropharynx to push down the soft palate; this gave the surgeon a good angle view and also a wide surgical field to improve navigation. A U-shaped incision was performed in the mucosa of the rhinopharynx with a "Thulium" laser (LISA laser products, Katlenburg-Lindau, Germany), setting limits: superiorly at the level of the inferior edge of the sphenoid sinus and laterally at the ostium of the tubes. The mucosal flap was gently rebated in the oropharynx and thus the muscular prevertebral fascia was clearly exposed, consisting in *longus capitis* and *longus colli* muscles. These were dissected and partially sectioned to expose the atlanto-occipital membrane, the anterior arch of C1 and the body of C2. In the case reported herewith, a complete fusion of the anterior arch of the atlas with the occipital bone was present and a partial fusion of the body of C2 with the atlanto-occipital joint; after causticization and sectioning of the membrane between C1 and C2 and always taking the clival region as the superior surgical limit, we started drilling the arch of C1 to expose the odontoid process; at this point, the lateral limits of drilling were represented by the occipital condyles and the atlanto-occipital joints that should be preserved. Since the odontoid process extended over the clival limit, prohibiting complete visualization of its cranial portion, it was necessary to drill

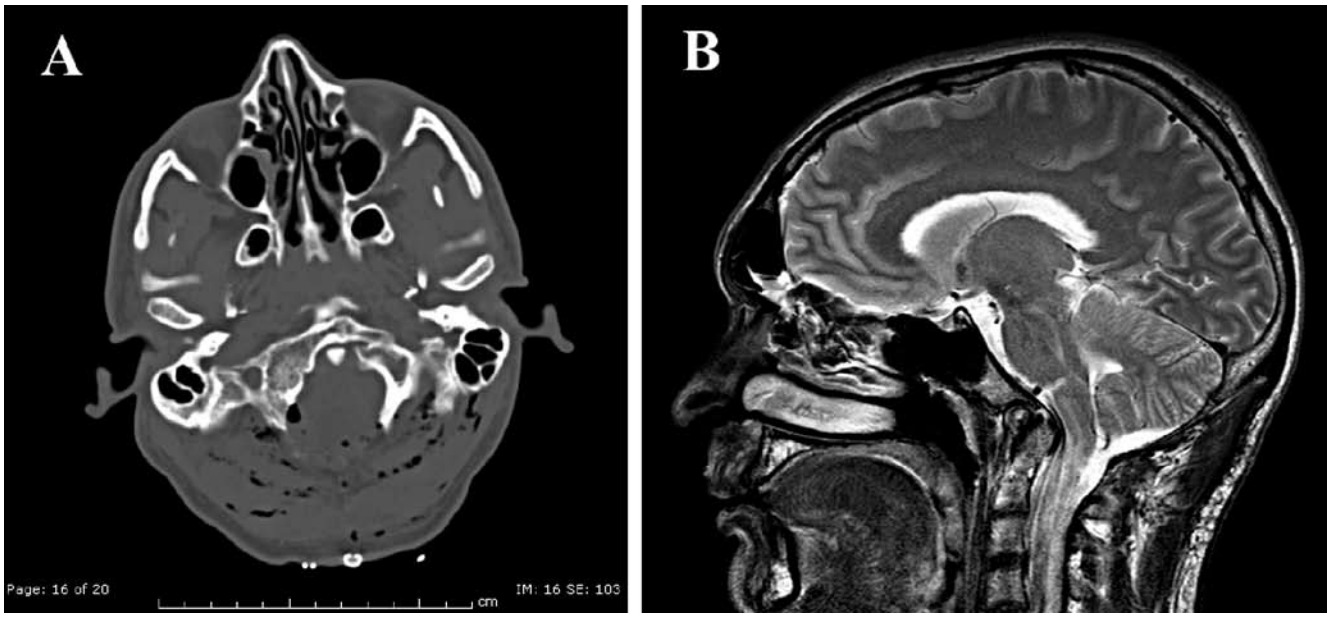

Fig. 1. A) Axial CT scan showing the relationship between C1, C2 and the vertebral canal. The odontoid process oversteps the asymmetric atlas, and both of them are partially fused with the cranial base. B) MRI T2-weighted sagittal scan demonstrating the posterior dislocation of the dens, which compresses the brainstem at the cervicomedullary junction.

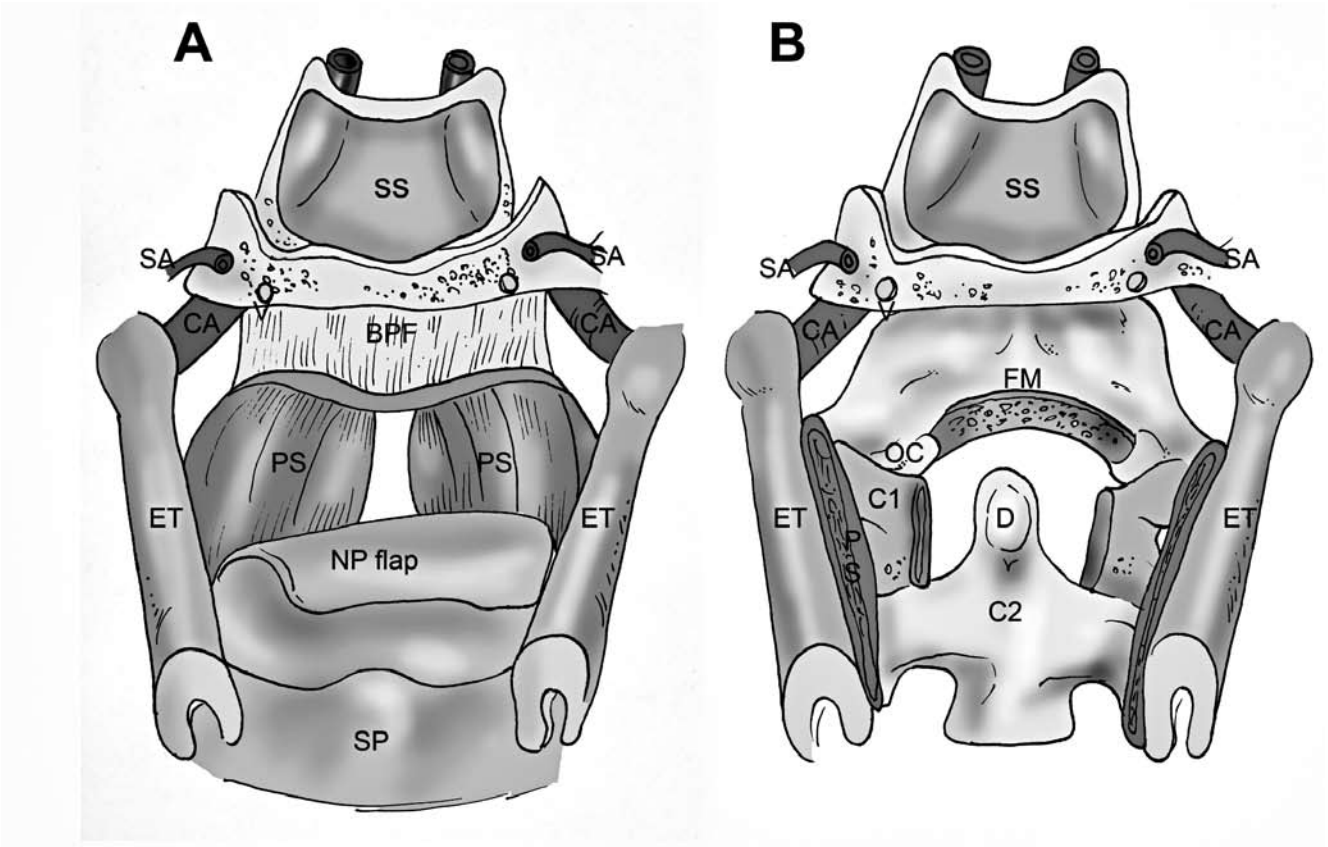

Fig. 2. Schematic drawing showing the surgical field and its exact limits before (A) and after (B) elevation of the nasopharyngeal flap and exposure of the C1 and C2 joint and cervico-medullary junction. (NP: nasopharyngeal flap; SS: sphenoid sinus; SA: sphenopalatine artery; CA: carotid artery; BPF: basipharyngeal fascia; PS: paraspinal muscles (longus capitis and colli muscles); ET: Eustachian tube; SP: soft palate; FM: foramen magnum; OC: occipital condyle; D: dens; C1: atlas; C2: epistropheus).

out part of the lower clivus; once the whole body of the odontoid was exposed, we commenced resection from the base corresponding to the body of C2. Always using a 0° degree endoscope, a thin cranial portion of the body of C2 was removed and the odontoid process was partially detached and the lateral alar ligaments sectioned. Using a 30° degree endoscope, we started drilling progressively and cranially and the odontoid was reduced to a thin bone cap, adhering to its apical ligament and piecemeal resected showing the tectorial membrane and the *dura mater*. At the end of the surgical manoeuvres, the mucosal flap was rebated in the rhinopharynx and fibrin glue and absorbable packing was placed in the rhinopharynx.

In the immediate post-operative period, the patient wore a rigid cervical collar and hospitalisation was uneventful; a post-operative CT scan revealed complete removal of the odontoid process and the anterior arch of C1, partial resection of the clival region and body of C2 and resolution of the ventral compression of the brainstem (Figs. 3A-B).

The patient was discharged on the 7th post-operative day, and after 1 month a cervical column fixation operation was planned. On the15th post-operative day, the patient reported an improvement in the diplopia and of the motor power of his lower extremities.

Discussion

In some patients, ACM-I could be associated with ventral brainstem compression [6], resulting in neurological dysfunctions such as limb weakness or diplopia. In these cases, multi-step surgery is required, including posterior cranial fossa decompression and an endoscopic approach to the ventral cranio-cervical junction.

Concerning the use of an endoscope for the anterior approach, when compared with the non-endoscopic technique, there are many surgical advantages, such as a wide panoramic view and the opportunity of placing the optical lens close to the surgical target. It also allows exploration into the narrow surgical anatomy and to obtain angled views to the anatomical corners [4].

In fact, the trans-oral approach is commonly used for resection of the odontoid process, when the latter produces a compression of the brainstem or spinal cord. This approach provides the most direct route to the ventral cranio-cervical junction, without injury to the major neurovascular struc-

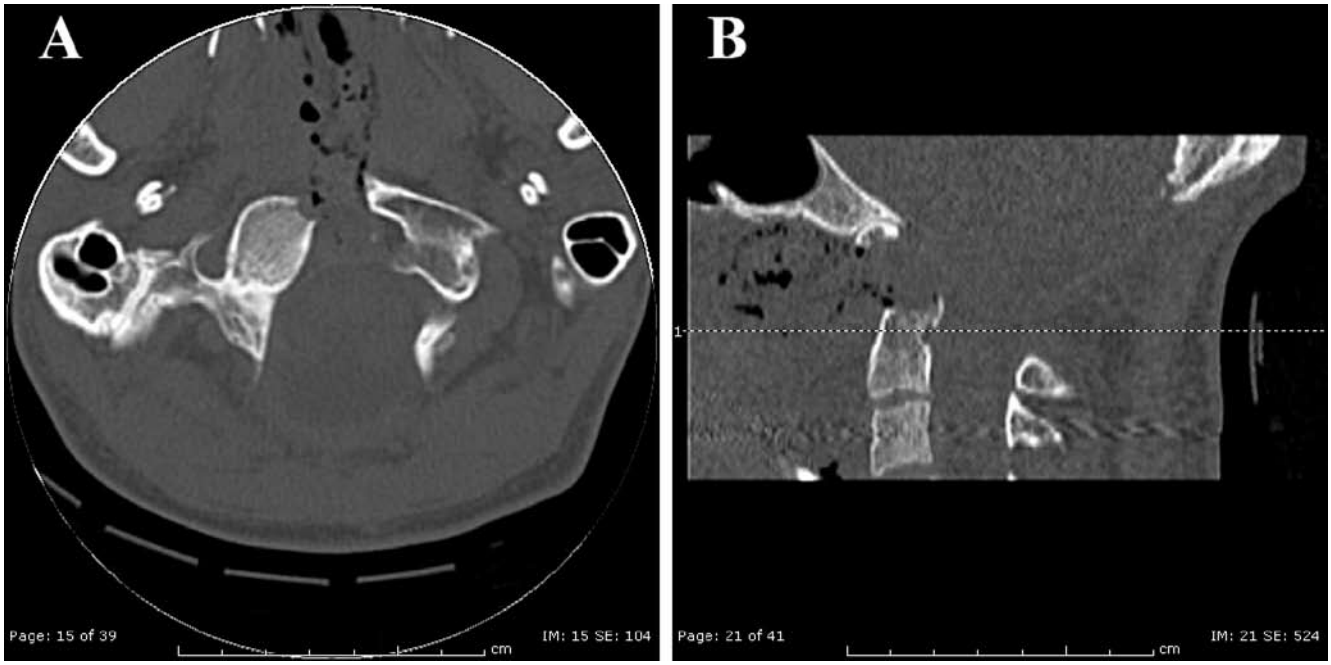

Fig. 3. A) Axial CT scan showing resection of the anterior arch of C1 and, at the back, removal of the odontoid process. B) Sagittal CT scan demonstrating the results of the trans-nasal endoscopic resection of the odontoid process. The brainstem at the level of the cervicomedullary junction is decompressed.

tures [4 7 8]. However, this kind of surgical technique presents some disadvantages: 1) difficulty in dural and pharyngeal closure with an increased risk of CSF leakage and contamination of the wound by saliva with a heavy bacterial flora, respectively [5 8]; 2) splitting of the soft and hard palate (that can lead to dysphonia), when an additional rostral and clivus exposure is needed, and occasionally, transmandibular or transmaxillary extension [7-9]; 3) velopharyngeal insufficiency; 4) tongue swelling and ischaemic necrosis, as a result of prolonged compression; 5) risk of damaging teeth with retractors; 6) the possible addition of tracheostomy, to avoid post-operative problems derived from airway swelling and upper airway obstruction, and gastrostomy or nasal feeding, that prolong hospitalisation time [8 10 11].

The introduction of the endoscopic endonasal approach to the ventral cranio-cervical junction results from years of experience and improvements with this technique in neurosurgical practice [12 13]. The endoscopic trans-nasal approach provides excellent prevertebral exposure of the craniovertebral junction in patients with small oral cavities, such as those with craniofacial abnormalities, and unlimited surgical access to the rostral midline cranio-cervical junction; however, it permits less access below C2 than the trans-oral approach and is limited by the patient's nasal anatomy [4 6].

Other advantages of the endo-nasal route are: the surgical corridor is now above the hard palate, and avoids splitting this structure; the use of mouth retractors is no longer necessary, which eliminates oral trauma with all the related complications. Furthermore, the defect created by this approach should not be exposed to the same degree of bacterial contamination from the oral cavity and oropharynx [5].

Nevertheless, odontoidectomy using the endoscopic endonasal approach is limited by the level of the C1-C2 junction, that cannot be below the level of the hard palate. One or both nostrils can be used to introduce the endoscope and the other instruments, such as the drill, and even though a middle turbinectomy or removal of the posterior portion of the nasal septum is required to improve surgical manoeuvrability, this does not produce any kind of problem for the patient [8].

Although further experience will be needed to improve the endoscopic trans-nasal approach to the ventral craniocervical junction, this technique should be considered an important alternative to the trans-oral route in selected cases, because of its advantages during surgery and in follow-up.

Conclusions

In the case reported here, we removed the odontoid process using a transnasal endoscopic route, in a patient with ACM-I, complicated by a brainstem compression. This approach provides an excellent surgical route, with many advantages when compared to the trans-oral approach, potentially avoiding serious intra-operative and post-operative complications. Even though surgeons have to gain more experience with endoscopic endo-nasal odontoidectomy, the minimally invasive access and the fewer complications associated with this technique, make it a valid alternative, in selected cases, for resection of the odontoid process in patients with severe brainstem compression.

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