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ORIGINAL ARTICLE

Craniovertebral junction lesions: our experience with the transoral surgical approach

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Abstract The aim of this study is to review our experience with the transoral surgical management of anterior craniovertebral junction (CVJ) lesions with particular attention to the decision making and to the indication for a consecutive stabilization. During 10 years (1998–2007), 52 consecutive patients presenting exclusively fixed anterior compression at the cervicomedullary junction underwent transoral surgery. Mean age was 55.85 years (range 17–75 years). Encountered lesions were: malformation (32 cases), rheumatoid arthritis (11 cases), tumor (5 cases) or trauma (4 cases). A total of 79% of patients presented with chronic/recurrent headache (cranial and/or high-cervical pain), 73% with varying degrees of quadrip aresis, and 29% with lower cranial nerve deficits. All of the patients but two, with posterior stabilization performed elsewhere, underwent synchronous anterior decompression and posterior occipitocervical fixation. Adjuncts to the transoral approach (Le Fort I with or without splitting of the palate), tailored to the local anatomy and to the extension of the lesions, were performed in seven cases. Follow-up ranged between 4 and 96 months. Of 35 patients with severe preoperative neurological deficits, 33 improved. The remaining 15 patients who presented with mild symptoms, healed throughout the follow-up. Perioperative mortality occurred in two cases and surgical morbidity in eight cases (dural laceration, cerebrospinal fluid leak with meningitis, malocclusion, oral wound dehiscence and occipital wound infection). Delayed instability occurred in one patient because of cranial settling of C2 vertebral body. A successful surgery achieving a stable decompression at the CVJ is an expertise demanding procedure. It requires accurate preoperative evaluation and, appropriate choice of decompression technique and stabilization instruments. Enlarged transoral approaches (despite higher morbidity) are a supportive means in cases of severe basilar invagination, cranial extension of the lesion or limited jaw mobility.

Keywords Craniocervical junction · Transoral odontoidectomy · Maxillotomy · Occipitocervical fusion

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Introduction

Anterior compression at the craniovertebral junction (CVJ) can occur in many types of pathology: malformation (basilar invagination, atlas assimilation, os odontoideum, atlantoaxial fixed dislocation), tumor, trauma, chronic inflammatory diseases (most frequently rheumatoid arthritis), osteomyelitis [9–13, 27]. In all these cases, transoral approaches (TOAs) consent to reach directly the site of the lesion and thus decompress the cervicomedullary junction staying away from laterally placed neurovascular structures [4, 15, 26, 27, 29]. By reporting surgical results and complications of this 52 treated patient series, the aim of this study is to revise the technical strategies, to identify the conditions requiring additional maxillotomies and to evaluate the necessity and the timing of postoperative stabilization.

Materials and methods

All patients with anterior lesions compressing the cervicomedullary tract who underwent transoral surgery during 10 years (1998–2007) were included in this study.

Radiographic imaging, previous and postoperative evaluations as well as outpatient archives were retrospectively checked. Thirty-two patients presented CVJ malformation (odontoid basilar invagination, atlas assimilation, atlantoaxial dislocation), 11 presented rheumatoid arthritis, 5 lesions were tumoral and 4 were trauma victims. The mean age at the time of treatment was 55.85 years (range 17–75 years); 28 females and 24 males were included in this study.

In one case, previous transoral decompression and occipitocervical stabilization, in one other case posterior stabilization alone and in two cases laminectomy were performed in other hospitals.

Local pain referred as cervical and/or cranial pain was present in 40 patients. Manifest quadriparesis of different grade was detected in 37 cases (motor/sensory). Cerebellovestibular signs were detected in nine patients. The lower cranial nerves were affected in 15 patients. Pathognomic physical marks such as short neck, low hairline and facial asymmetry were observed in 18 cases of CVJ malformation. Duration of symptoms at admission was 6 months to 4 years.

Atlantoaxial mobility was checked by means of dynamic X-ray (atlantodental interval, clivodental interval, spinal canal diameter) in cases of CVJ malformation. MRI and 3D-CT scan were used to detect the site of compression, the extension of the lesion (in case of tumor or rheumatoid pannus, gadolinium contrast was used), to determine the position of the vertebral artery (in order to prepare the posterior stabilization). The indications for surgery were: tumor, compressing pannus, irreducible compression (fracture, malformation), tip of the odontoid process at least 2.5 mm above the Chamberlain's line.

Standard transoral approach was used in 45 patients, Le Fort I osteotomy was required in 5 patients (platybasia, odontoid tip more than 20 mm above Chamberlain's line) and two cases required palate split. All patients but two with posterior stabilization performed elsewhere underwent posterior fixation consecutively in the same sitting.

Operative tips

The duration of consecutively performed transoral decompression and posterior stabilization was 5–8 h. We routinely employ a hot air blanket for temperature regulation and pneumatic foot compression to prevent deep venous thrombosis. Tracheostomy, used in the first 12 cases, has been abandoned (except 7 cases in which maxillotomy was performed) and fiberoptic nasotracheal intubation is preferred [17]. Intravenous prophylactic antibiotics is given in all cases. Patient is put in mild Trendelemburg position with the head stabilized in a Mayfield holder. The retraction of the soft palate is performed by means of two rubber catheters sutured to the soft palate, through the nostrils, and fixed cranially to the Mayfield holder.

During decompression, when C1 is not interested, the anterior arch is spared by using a bottom-up dens-drilling technique (by drilling the base of the dens until it is possible to clasp the tip with a rongeur and to remove the apical ligaments). In all cases, tumor, pannus or even CVJ malformation, the cruciate ligament was cut and the dura mater was exposed. Antibiotic wax is always placed in the operative cavity right before mucosa and pharyngeal muscles are sutured in single layer [5, 23, 24, 26].

Subsequently, the patient is turned prone. To achieve occipitocervical stabilization, interlaminar hooks and occipital screws were used in the first 20 cases. More recently, we use a system with a precontoured plate fixed to the occiput with bicortical screws and independent rods fixed by screws to the lateral masses of the subaxial cervical spine. Autologous bone harvested from the local spinous processes and/or from the iliac crest are used for fusion. For an operative facilitation, the stabilization system is applied in a head flexed position without blocking the connectors. In fact, these are blocked only after having re-extended the head to regain a position most proximal to a preoperative X-ray performed in a standing position.

Postoperatively all patients are transitorily transferred to an intensive care unit. Cold fluid is allowed starting from the third postoperative day. CT scans are usually performed 3–7 days after surgery and in the same period of time, patients start mobilization (Fig. 1). An MRI is always performed within 1 month from surgery. 3-, 6- and 12-month outpatient checks are performed by X-ray and neurological examinations. Additional CT scans are also performed at the last check. Successively, patients are yearly checked through outpatient visits. Decompression status (including tumor control), fusion, stability in addition to neurological status are always detected.

Results

Mortality directly correlated to surgery did not occur; however, pulmonary embolism in two cases of severe spastic quadriplegia conducted to death within 20 days (Table 1).

Complications directly related to surgery were vertebral artery perforation in one case, dural tear in one case, superficial occipital wound infection in two cases, CSF fistula in one case which leads to meningitis, malocclusion in one case of Le Fort maxillotomy, posterior pharyngeal wall dehiscence in one patient, articular screw lose due to malpositioning in one case of CVJ malformation associated with severe scoliosis.

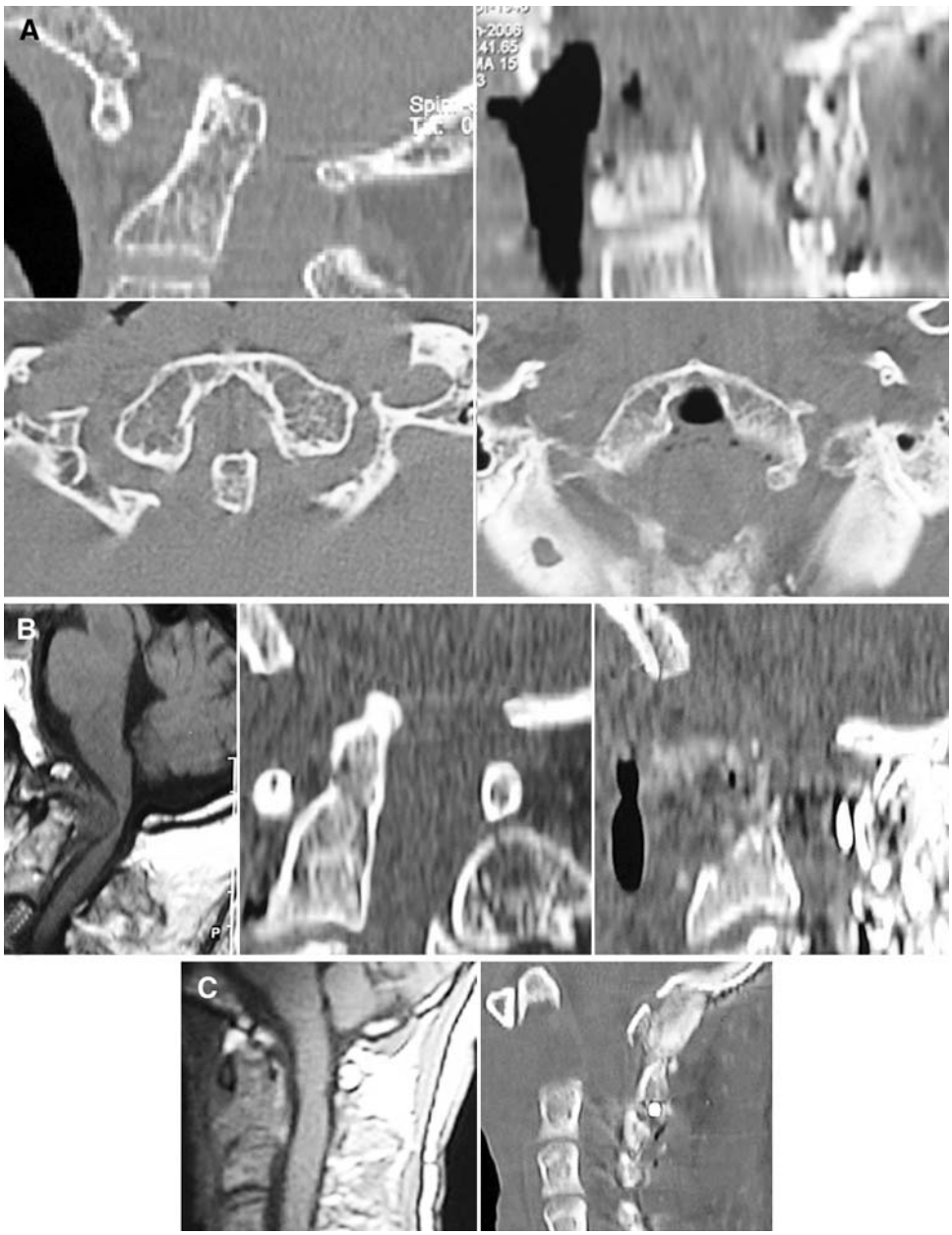

Fig. 1 Three types of cases requiring a transoral surgical decompressive approach and synchronus posterior fusion. **a** Preoperative (*left*) sagittal (*up*) and axial (*down*) reformatted CT scans illustrating a basilar invagination associated to atlantoaxial dislocation and atlas assimilation with postoperative results (*right*) in sagittal (*up*) and axial (*down*) reformatted CT scans. **b** Sagittal T1-weighted MRI (*left*) and sagittal CT scans (*middle*) presenting a case of rheumatoid arthritis with abundant pannus behind the dens compressing and deviating the cerviomedullary junction with surgical results illustrated by a postoperative sagittal CT scan. **c** A preoperative T1-weighted MRI (*left*) and postoperative sagittal CT scan (*right*) of a giant cell tumor of axis that was totally removed

Other systemic complications occurred: six deep vein thrombosis, four urinary infection, one chest infection and one pressure sore.

The mean duration of follow-up after surgery was 31 months ($\approx$2.5 years) and ranged from 4 to 96 months. On postoperative neuroradiological checks, decompression was considered satisfactory in all cases but two who needed a transoral revision. Fusion was considered proper in all surviving cases. Two patients who had postoperatively presented stable conditions in one case and initial improvement in the other, died at, respectively, 8 and 20 months from surgery due to causes unrelated with surgery. None of the patients deteriorated at follow-up. Neurological conditions in 33 of the 35 surviving patients with different level of quadriparesis improved. One case of adjunctive maxillotomy presented postoperative velopharyngeal dysfunction and needed pharyngoplasty. Postoperatively, swallowing function improved in seven patients and did not change in one.

Discussion

Transoral decompression

Posterior approaches at the CVJ have been successful for posterior fusion and thus play a role only in cases of reducible compression [9–11, 20–22, 27, 30]. Nevertheless,

Table 1 A short review of the presented series including presentation, surgery and follow-up

	Age, sex	Lesion	Previous surgery	Approach	Complications	Revision	Mean preop symptoms	Neurological results	Follow-up (months)
1	62, M	Malf	No	TO	–	No	Headache, quadriparesis	↑	90
2	65, F	RA	No	TO	–	No	Headache, low nerves	=	96
3	17, M	TUM	No	TO	Urinary infection	No	Headache	=	72
4	67, F	Malf	No	TO	Dural laceration, urinary infection	No	Headache, quadriparesis	↑	86
5	56, F	Malf	Posterior decompression	TO	–	No	Headache, quadriparesis, low nerves	=	89
6	63, F	Malf	No	TO	Pulmonary embolism	No	Headache, quadriparesis	–	Death 7 days
7	61,M	Malf	No	TO	–	No	Headache, quadriparesis	↑	88
8	53, F	Malf	TO + Posterior fusion	TO	–	No	quadriparesis	↑	85
9	56, M	Malf	No	TO	Oral wound dehiscence	Oral wound revision	quadriparesis, low nerves	=	72
10	54, M	Malf, KF	No	TO	–	No	Headache, quadriparesis	↑	70
11	45, F	TUM	No	TO	DVT	No	Headache, quadriparesis	↑	73
12	50, M	Malf	No	TO	Pseudoarthrosis	TO revision for cranial settling	Headache, quadriparesis	↑	66
13	26, M	TR	No	TO	–	No	–	=	12
14	69, F	Malf	No	ODM	Velopharyngeal dysfunction, DVT	No	Headache, quadriparesis	↑	52
15	58, M	Malf	No	TO	–	No	Headache, quadriparesis	↑	48
16	37, M	Malf	No	TO	–	No	Headache, quadriparesis, low nerves	↑	45
17	65, F	TUM	No	TO	Pulmonary embolism	No	Headache	–	Death 15 days
18	59, F	Malf	No	TO	DVT	No	Quadriparesis	↑	42
19	32, M	Malf	No	TO	–	No	Headache	=	37
20	72, F	RA	No	LFO	Malocclusion	No	Headache, quadriparesis, low nerves	↑	44
21	70, M	TR	No	TO	CSF fistula, meningitis	No	Headache	=	12
22	66, F	RA	No	TO	–	No	Headache	=	26
23	75, M	Malf	No	ODM	Pressure sore	No	Headache, quadriparesis, low nerves	↑	24
24	50, M	TUM	No	TO	DVT	No	Headache, quadriparesis	↑	36
25	64, F	Malf, KF	No	TO	–	No	Headache, quadriparesis	=	18

Table 1 continued

	Age, sex	Lesion	Previous surgery	Approach	Complications	Revision	Mean preop symptoms	Neurological results	Follow-up (months)
26	66, F	Malf	No	TO	–	No	Headache, quadriparesis, low nerves	↑	17
27	47, F	Malf, KF	No	TO	–	TO revision	Quadriparesis, low nerves	↑	14
28	62, F	RA	No	LFO	–	No	Headache	=	15
29	50, F	Malf	No	TO	–	No	Headache, quadriparesis, low nerves	↑	13
30	74, F	Malf	No	TO	DVT	No	Headache, quadriparesis	↑	Death 20 months
31	67, F	RA	No	TO	–	No	quadriparesis	↑	13
32	42, F	Malf	No	TO	–	No	Headache	=	12
33	67, M	TR	No	TO	Pressure sore	No	Headache	=	11
34	64, F	Malf	Posterior decompression	TO	Urinary infection	No	Headache, quadriparesis, low nerves	↑	6
35	18, M	RA	No	TO	–	No	Headache	=	7
36	57, M	TR	No	TO	–	No	quadriparesis	↑	5
37	56, F	Malf	No	TO	–	No	Headache, quadriparesis	↑	6
38	57, F	RA	No	TO	–	No	Headache	=	Death 8 months
39	66, M	Malf	No	LFO	Screw lose	No	Headache, quadriparesis, low nerves	↑	7
40	75, F	RA	No	TO	DVT	No	Quadriparesis, low nerves	↑	6
41	23, M	Malf, DS	No	TO	–	No	Headache	=	6
42	68, F	RA	No	TO	Vertebral artery perforation	No	Headache, low nerves	↑	4
43	62, F	Malf	No	TO	–	To revision	Headache, quadriparesis	↑	5
44	67, M	RA	No	LFO	–	No	Quadriparesis, low nerves	=	4
45	44, M	TUM	No	TO	Urinary infection	No	Quadriparesis	↑	6
46	66, M	Malf	No	TO	–	No	Headache, quadriparesis	↑	5
47	46, F	Malf, KF	No	LFO	–	No	quadriparesis	↑	6
48	18, M	Malf, DS	No	TO	–	No	Headache,	=	6
49	52, F	RA	No	TO	–	No	Headache, quadriparesis	↑	5
50	71, M	Malf	Posterior fusion	TO	–	No	Headache, low nerves	↑	5
51	68, M	Malf	No	TO	–	No	Headache, quadriparesis	↑	4
52	59, F	Malf	No	TO	–	No	Headache, quadriparesis	↑	4

52 patients with craniovertebral junction lesions surgically treated by a transoral approach

Malf craniovertebral malformation (atlantoaxial dislocation, basilar invagination, Atlas assimilation), *KF* Klippel-Feil anomaly, DS *down syndrome, DVT* deep venous thrombosis, *LFO* Le Fort I osteotomy, *ODM* open-door maxillotomy; *preop* preoperative, *postop* postoperative, *TO* transoral, ↑ improved, = stable, *M* male, *F* female

progressive understanding of the craniocervical dynamics and innovative techniques to determine the site of encroachment, lead to conclusion that the transoral approach, with addition of the proper type of stabilization, is the gold standard in the treatment of pathologies causing irreducible anterior compression of the cervicomedullary segment [6, 7, 20–22]. Also lateral approaches are practicable but they require an extensive dissection, risk of injury of the vertebral artery, of the jugular bulb, and of the hypoglossal nerve and we prefer to reserve them to massive tumor masses involving lateral structures [1, 27, 30, 31].

Over time, our surgical treatment philosophy evolved and we adopted several technical variations for transoral operations. In fact, we have started using a prophylactic broad-spectrum antibiotic therapy to all patients instead of basing our antibiotic coverage on preoperative pharyngeal flora, as advocated by some authors. Preoperative tracheostomy has been abandoned except for cases of transoral extended maxillotomy approach [17, 32]. Le Fort I maxillotomy, permitting sufficient rostral exposition of the clivus and the tip of invaginated dens as in severe basilar invagination or in voluminous tumor masses, has surrogated soft palate incision [2, 4, 12, 27, 28]. By this way, complications related to soft palate wound dehiscence are avoided [4, 13]. Careful neuroimaging preoperative study of the pathologic anatomy of this area is crucial to select the appropriate approach and to avoid catastrophic complications [13]. Other approaches that can assist the transoral approach include the open-door maxillotomy approach, the transoral–translabiomandibular approach, the transoral bilateral sagittal split mandibular osteotomy approach and the mandibular swing-transcervical approach [6, 7, 17, 32].

In patients with limited jaw mobility we favor use of the open-door maxillotomy approach [3, 7, 14, 17]. The open-door maxillotomy approach should be restricted to the needs of the individual patients because of its inherent morbidity including potential dental malocclusion, velopharyngeal dysfunction and palatal fistulas [3, 17–19].

Instability after transoral approaches

No diagnostic means are available to correlate the transoral odontoidectomy extension with the CVJ instability [8]. Nonetheless, most authors advocate posterior fusion following transoral decompression [8]. Significantly, we reported one postoperative death from acute dislocation of the axis on the day before a planned posterior fixation in a previous report of 25 CVJ malformations treated by TOA [10]. In order to eliminate the risk of acute postoperative instability and to early mobilize the patients we have moved from performing a planned posterior fixation as a second procedure to a one-stage transoral decompression and occipitocervical fixation. Based on biomechanical studies, the integrity of C1-ring and lateral masses minimizes the risk of cranial settling [8, 23, 24]. In this series, one patient experienced symptomatic cranial settling of C2 vertebral body after one-stage transoral decompression despite occipitocervical fixation and required a second anterior decompression. In our subsequent patients, in order to conserve the ring of C1 anteriorly, we used the bottom-up C1 sparing drilling technique. More recently, we have been using precontoured plate rod systems with screws in the occipital bone, C2 pedicles and lateral masses of the cervical spine which, accordingly to recent biomechanical studies, has resulted in major stability and resistance to cranial settling [16, 25].

Conclusions

One-stage transoral decompression and posterior occipitocervical fixation are feasible and effective in patients with CVJ anterior fixed compression, including malformation, tumor, trauma, inflammatory diseases. The transoral approaches must be tailored to a particular patient's pathological findings with a thorough understanding of the malformed anatomy of the CVJ. Severe basilar invagination, cranially extended lesions or cases of limited mandibular excursion may require a transmaxillay approach. Successful transoral decompression of complex CVJ malformations remains a major neurosurgical challenge and requires an adequate learning curve.

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

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