Cervical myelopathy caused by ventrally located atlanto-axial synovial cysts: An open quest for the safest and most effective surgical management. Case series and systematic review of the literature

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Cervical myelopathy caused by ventrally located atlanto-axial synovial cysts: an open quest for the safest and most effective surgical management. Case series and systematic review of the literature.

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Abstract

Summary of Background Data: Despite a good understanding of the natural history of spinal synovial cysts (SCs), a widespread agreement regarding their optimal management is still lacking. This is particularly true for SCs occurring at the C1-C2 level, which are rare, but oftentimes lead to a rapidly evolving cervical myelopathy.

Methods: We report a series of 4 patients (M:F ratio = 1:1; mean age 63.5 years) presenting with progressive cervical myelopathy secondary to ventrally located C1-C2 SCs. All patients underwent a postero-lateral facet-sparing intradural approach with total excision of the SCs. Functional status was assessed pre- and postoperatively with Nurick scale and the modified Japanese Orthopaedic association grading. Furthermore we conducted a systematic review, following PRISMA guidelines of pertinent literature to contextualize the options for surgical management of such lesions.

Results: Complete excision of the SCs was confirmed radiologically and on histological analysis. All measures of functional status improved post-operatively, and no cyst recurrence or need for instrumented fusion were noted during follow up (range from 22 to 88 months).

Conclusion: Our experience suggests that the facet-sparing intradural approach provides excellent clinical outcomes without causing any C1–C2 instability. This is in keeping with the take home message emerging from our literature review, which confirms that treatment should aim at radical resection of SCs while minimizing the risk of postoperative instability.

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ABBREVIATIONS:

Synovial cysts (SCs)

MEPs (Motor Evoked Potentials)

SSEPs (Somatosensory Evoked Potentials)

EMG (Electromyography)

Male (M)

Female (F)

Milliliter (mL)

Nurick scale (NS)

modified Japanese Orthopaedic association (mJOA)

Computer-Tomography (CT)

Magnetic resonance Imaging (MRI)

cerebrospinal fluid (CSF)

Introduction

Synovial cysts (SCs) are well-recognized pathological entities arising at any spinal level, nonetheless those occurring at the C1-C2 level are particularly rare. Those lesions can project from the anterior or posterior aspect of the facet joint protruding internally or externally to the spinal canal. The growth of C1-C2 SCs is usually progressive, thus three modalities of clinical presentation are possible: a) these lesions may either be indolent for long periods, being asymptomatic at time of incidental diagnosis, b) they may manifest with signs and symptoms of a sizeable compression of the spinal cord, or c) they may cause an abrupt presentation as a result of an intra-cystic bleeding. A range of clinical findings may therefore be related to SCs: from unspecific cervical/nuchal pain to neurological signs of chronic or acute radicular and/or spinal cord compression[1].

SCs' optimal management is still a matter of debate. So far, several treatment modalities have been proposed depending on the spinal level affected; those strategies aim at obtaining a decompression of the neural structures (spinal cord, thecal sac and nerve roots) of the affected functional spinal unit, while preserving its stability. As such, treatment modalities include CT guided aspiration, facet-sparing excision through a postero-lateral approach, posterior laminectomy and fusion, trans-oral approach, or a combination of anterior excision and posterior instrumented fusion[1-8].

Here we report a series of 4 patients presenting with progressive cervical myelopathy secondary to C1-C2 SCs ventrally located. The clinical and radiological outcomes following a postero-lateral facet-sparing approach with total excision of the SCs, are discussed along with a systematic literature review.

Materials and Methods

We present a case series of 4 patients harbouring ventrally located C1-C2 SCs and presenting with rapidly evolving cervical myelopathy, who were prospectively collected from our tertiary centre hospital. All patient underwent radical surgical excision through a postero-lateral facet-joint sparing approach, assisted by perioperative neurophysiological monitoring (MEPs, SSEPs and free EMG). Functional status was assessed pre- and postoperatively using Nurick scale (NS) and the modified Japanese Orthopaedic association (mJOA) grading. All patients were followed-up clinically at 6 weeks, 6, 12 and 24 months, and radiologically with CCJ MRI performed at 6 and 24 months.

Surgical technique:

Neurophysiological monitoring (MEPs, SSEPs and free EMG) was initiated at baseline, before flipping the patients prone on the operating table with their head slightly flexed as per the well-known Concorde position. Following a midline incision and exposure of the C1-C2 posterior elements, a posterior arch of C1 and partial C2 unilateral laminectomies were performed in order to access the

SCs. To ensure a complete exposure of the lesions a trans-dural approach was always preferred to an extradural postero-lateral approach. Having performed an arciform paramedian dural incision, and section of the dentate ligaments to avoid excessive retraction of the spinal cord, SCs were accessed through an incision of the bulging ventral dural plane. Of note, at the end of each procedure the anterior dural plane was reconstructed intradurally by sliding a self-adhesive dural patch, whereas the posterior plane was stitched with 4.0 Prolene (Ethicon Inc., USA) to provide a watertight anatomical plane reconstruction, the surgical site was then glued with a dural sealant to prevent postoperative leakage of cerebrospinal fluid (CSF).

Ethical Approval

This study was conducted according to the Ethical Principles for Medical Research Involving Human subjects stated in the 2004 and its further revision made in 2008 and 2013 of the Declaration of Helsinki. The study was approved by the research Board of the "College de Neurochirurgie" (IRB n° IRB00011687 #1: 2020/15).

Systematic review of the literature

Search strategy

A systematic review of the literature was performed for published articles reporting on atlanto-axial synovial cysts. PubMed/ MEDLINE, Scopus, and the Cochrane Database of Systematic Reviews were used to search the following keywords: "cervical AND synovial cyst," "atlantoaxial AND synovial cyst," "C1-C2 AND synovial cyst," "cervical AND juxta-facet cyst," "atlantoaxial AND juxta-facet cyst," "C1-C2 AND juxta-facet cyst", "ventral C1-C2 cyst" and "anterior C1-C2 cyst". The selected keywords were included in the title, abstract, or keywords list. The search was restricted to articles published until January 2020.

Study selection

In this study, we included articles reporting patients atlanto-axial synovial cysts. A first screening, consisted in reviewing all titles and abstract of the papers shortlisted: only articles focusing on the atlanto-axial SCs were retained. Any non-English article was excluded. The second screening was meant to select which articles fulfilled inclusion criteria and therefore deemed for full text review. To this extent case series, Case–control studies, cohort studies, retrospective studies, systematic reviews, meta-analyses or clinical trials were considered suitable for inclusion. Case reports, and technical notes were also included because of the rarity of the pathology. On the other hand, studies involving animals, as well as editorials were excluded.

The results were independently screened by four of the authors (S.C., A.G., M.G. and I.Z.); disagreements were solved by consensus among all the Authors.

Data extraction

The following data were extracted from each article, if clearly reported: year of publication, Journal of publication, type of study, number of neurosurgical centers involved, number of patients, epidemiological data (gender, average age) types of complications and average follow-up.

Results

Surgical findings

Using a postero-lateral facet-joint sparing trans-dural approach, a good dissection plane between the SCs and the spinal cord/nerve roots was identified in all cases; the working channel provided by such an approach resulted always large enough to guarantee the dexterity needed to achieve a radical excision of the SCs without compromising the C1-C2 stability. The cyst was quite soft and easily to be fragmented and aspirated in 3 cases, it was very firm in the remnant one although we were able, after a progressive volume reduction by a rongeur, to identify and separate its insertion taking it out in "en bloc" fashion.

Search results

The literature was searched up to January 2020. A total of 288 records were identified through the database and only 261 considered after duplication removal. Literature review results were included in the PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) flow diagram (see also Figure 1).

The first stage of screening of articles titles and abstracts led to the exclusion of 179 studies. From the remaining 82 studies, 41 articles were removed as they were not meeting inclusion criteria in terms of type of articles suitable for systematic review, discussed in the method section.

This screening process resulted in the selection of 41 peer-reviewed publications whose full-text were considered for further analysis and inserted in our database.

Case series

The present series consists of 4 patients, 2 male and 2 female, with a mean age of 63.5 years (ranging from 58.7 to 72.3 years), all harbouring ventrally-located C1-C2 SCs, referred to our attention for a rapidly progressive cervical myelopathy. Clinical and radiological features, management strategy and outcome at follow up are summarized in Table 1. Symptoms typically included nuchalgia, swallowing difficulties, unsteadiness and tetraparesis. All patients underwent the surgical management described above: the procedure lasted on average 106 minutes (range: 75-145 minutes) and the estimated blood loss was always minimal (<300ml). No intraoperative nor postoperative complications directly related to the surgical excision of the SC were recorded. The histopathological analysis confirmed in all cases a thick wall lined with synovial cells, containing a mixture of granulation tissue, giant cells and histiocytes, hence confirming the nature of the lesion and ruling out the diagnosis of pseudo-cysts such

as ganglion or ligamentous cysts. All patient showed a good clinical and radiological outcome at last follow up as shown in Table 1.

Illustrative case

A 53-year-old woman referred to our institution with a 2-month history of worsening neck and left shoulder pain as well as numbness and tingling in both arms (predominantly on the left-side) is presented as an exemplificative case of this series. Neurological findings included a Lhermitte's sign, an unsteady gait and a mild atrophy of both hands intrinsic muscles. Preoperative MRI neuroimaging demonstrated a T1-weighted hypo-intense and T2-weighted hyper-intense cystic lesion sitting behind the dens of C2 responsible for spinal cord compression. In fact, the intramedullary T2-weighted hyper-intensity raised concerns of an ongoing spinal cord damage resulting from this compression (Figure 2). Following radical excision and histological confirmation of a diagnosis of SC the patient was rapidly mobilized to minimize the risk of thromboembolism and started an intensive rehabilitation course. The surgical wound healed nicely and no CSF leak occurred, as confirmed by the absence of clinical signs and symptoms of intracranial hypotension or formation of a postsurgical pseudomeningocele. The patient demonstrated a rapid clinical improvement by recovering to a NS 2 and mJOA 14; this outcome resulted long-lasting as confirmed on her last follow-up when patient confirmed to be pain free without any mechanical instability.

Discussion

SCs are cystic dilatations of the synovial sheaths[1]; spinal joints involvement, especially in the lumbar segment, is not infrequent and represents a well-recognized pathological entity to the community of spine surgeons. The occurrence of SCs in the occipito-cervical region is rare: C1-C2 SCs are usually connected to the articular facets of the distal vertebra, and may be responsible for mechanical or radicular pain[1].

Typically, the clinical presentation is characterized by a rapid neurological deterioration within few weeks to months, which follows a slow progression over many years [10,11]. Preoperatively, a diagnosis of SCs is suspected on CT and MRI findings. SCs are usually round-shaped cystic lesions located laterally to adjacent facet joint[12]. Dynamic imaging with flexion-extension x-rays are recommended since subclinical instability has been suggested as one of the causes for cyst formation; nonetheless signs of preoperative instability are overall rare [10,11].

Management and Literature Review: The optimal management of C1-C2 SCs is still a matter of debate; their complexity lies in the relevant biomechanical role that this articulation has on head rotation and cervical range of movement: in fact, the atlanto-axial articulation is responsible for 50% of cervical rotation (up to 50°). Several treatments have been suggested including CT-guided cyst aspiration, steroids facet injections and surgical excision with or without spinal fusion[13]. Observation with a cervical rigid collar has been advocated by some, especially in frail patients; however due to the

poor compliance, long term clinical results were largely disappointing[11]. In patients not eligible for surgery also steroid injections can be considered, but pain control is usually maintained only for few months; the time frame for cyst recurrence following CT guided aspiration is similar, and overall failure rates are high (50 to 90%) when these are used as a stand-alone treatment modalities [14,15].

Different surgical approaches have been proposed, including the postero-lateral, lateral and trans-oral ones[16]. Through our literature review we have identified forty-two relevant articles for a total of 76 cases including those described in the present article (Table 2), of note, it emerged that no systematic review or meta-analysis of the literature has ever been conducted on this rare condition. The mean age at time of presentation was 71.24 years (range 45-85 years), whereas the most commonly performed procedure appears to be the trans-oral decompression with posterior fusion which was performed in 32 cases. Posterior laminectomy without fusion in 22 cases or with fusion in 7 cases represent the second most common management strategy; the other management options so fare reported include lateral approach with or without fusion in 4 cases, conservative management with collar, CT guided aspiration or no treatment at all was reported in 5 cases. Of note the use of trans-oral approach without fusion has been described in one patient only.

Transoral approach: The trans-oral approach is the most commonly reported approach for C1-C2 SCs. It entails an extensive drilling of the anterior arch of C1 and the dens of C2 to achieve a good exposure on the cyst[17,18]. The trans-oral approach is technically demanding and carries high intraoperative and postoperative complications rates including: dural tears, dysphagia, wound dehiscence, surgical site infections[1]. Usually this approach requires parenteral nutrition and tracheostomy, resulting in a longer hospitalisation, and more medical morbidities [1]. Moreover, due to the inherent destabilization of the occipito-cervical junction, a second surgical stage for posterior fusion is usually recommended, although one case of trans-oral approach without posterior fusion has been described [1].

Lateral approach: Overall the indications for far lateral approach (either retro-condylar or trans-condylar) are very narrow in C1-C2 SCs, and this procedure has been used only in challenging scenarios: it certainly represents an alternative to the trans-oral approach in very sizeable lesions that can hardly be accessed through a posterior route.

It should be highlighted, however, that in some, very few, specific cases this approach could require a posterior stabilization (with either Magerl, Goel or Harms technique), and therefore implies iatrogenic risks of vertebral artery and C2 ganglion damage. Although in most cases, unilateral damage to the C2 ganglion or vertebral artery are asymptomatic, other long-term complications of instrumented fixation include implant failure, screws pull out, non-fusion, etc.

Postero-Lateral approach (decompressive laminectomy) without fusion: The posterior approach is the treatment of choice for C1-C2 SCs located in the dorsolateral aspect of the spinal canal, and seems well tolerated by the patient given the very low complication rate reported. Although the exposure of ventrally located cysts might be more challenging, their cystic nature often allows for an adequate

decompression by this route [2]. In our experience this is particularly true in case of small lesions (< 1 cm in diameter). Additionally, some have advocated a subtotal resection of SCs while focusing on guaranteeing an adequate decompression of the spinal cord and nerve roots. To this regard, it is worth mentioning that the role of total resection on the recurrence rate of atlanto-axial SCs has not been established in the literature.

This approach does not cause any instability, provided that the facet joint is carefully preserved at time of SC's excision. In our experience, to achieve a complete SC excision with the facet sparing approach, the surgical team must pay a great deal of attention in optimising the exposure with a single or multilevel unilateral hemi-laminectomy, and control all sources of extradural bleeding[1]. Furthermore, it is of paramount importance to identify the origin of the SC and obtain a good dissection plane between the cyst and the spinal cord. To do so surgeons can make use of the intraoperative ultrasound[19], optimise the light source directed toward the surgical site, if necessary coupling the microsurgical view with the introduction of an endoscope (usually with a 30° lens), and rely on the neurophysiological monitoring while gently displacing the spinal cord [1].

Facet-Sparing Intradural approach: A variation of the postero-lateral approach entails the intradural access as an alternative to the purely extradural route. Madhavan K et al[58] has recently reported a 3 cases series using this approach, and proposing it for asymmetrically located extradural cysts especially in elderly patients affected by several comorbidities. The results from our case series confirm their initial findings in a much younger population, characterized by more severe myelopathic features at baseline, and with a more systematic and longer follow up. Because of these, our experience appears as a relevant contribution in demonstrating that such approach is safe, time-effective, and provides a valuable alternative to open posterior instrumented, open anterior trans-pharyngeal and endoscopic approaches, also in patients with ventrally located SCs. On the other hand this approach could be risky in case of very large SCs far laterally extending on both side due to the impossibility of retracting the spinal cord unless of performing a bilateral approach. For the latter purpose a major concern about the CCJ stability could rise. Finally, we could hypothesize that this approach might have the inconvenient, in case of anterior chronic CSF leak, of low intracranial pressure syndrome, although it was never encountered in the present small series.

Posterior fusion: Similarly to the treatment of soft pannus in patients with rheumatoid arthritis, some authors have advocated a dorsal instrumentation without decompression also for SCs. The rationale is that the treatment of the suspected underlying instability of the atlanto-axial articulation would stop the growth if not induce a shrinkage of the cyst[7,11]. Whereas stabilisation without decompression can be considered in pauci-symptomatic patients, this choice is highly debatable in case of myelopathy with evidence of intramedullary T2 hyper-intensity. On the other hand, the role of posterior instrumentation to induce fusion and reduce the risk of recurrence has been emphasized by many authors. However, a review of the current literature does not provide evidence that cases managed without instrumentation have higher rates of cyst recurrence at follow up [7].

Study limitations

The present study present few limitations as follow: although this pathology is quite rare, the presented series is too small to draw any clear conclusion, thus further studies are needed to improve/support the level of evidence of the proposed management. Also the literature review present the limitation of being focused to only English study.

Conclusion

The use of a postero-lateral facet-sparing intradural approach to C1-C2 SCs seems a valuable option to manage challenging ventrally-located SCs. Such an approach is indicated for medial and/or asymmetrically located SCs. It should be considered as an alternative to postero-lateral extradural route in patients with rapidly evolving radiculopathy and myelopathy. Also, such an approach does not require any instrumentation and therefore does not limit the cervical range of motion in the postoperative period.

Postoperative fusion does not seem to be required to avoid recurrence of C1-C2 SCs, whereas CSF leak and meningitis rate through the postero-lateral approaches, including the intradural ones, are much lower than the complications rate related to the trans-oral approach.

The postero-lateral route is certainly less invasive than the transoral one, especially in frail, elderly patients, and guarantees a shorter hospital stay with satisfactory functional outcome at late follow up.

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Figure legends

Figure 1. Concorde position adopted for the surgery

Figure 2. Sagittal T2 MRI. A hyper-intense cyst with spinal cord compression and intramedullary hyper-intensity is seen and follow-up after 4 years. The patient was pain free, with no clinical or radiographic evidence of recurrence/instability

Figure 3. steps of the surgical procedure for the removal of the synovial cysts: dural exposure (A); identification of the synovial cyst (B); removal of the cyst (C)

Fig. 1 PRISMA Flow Diagram

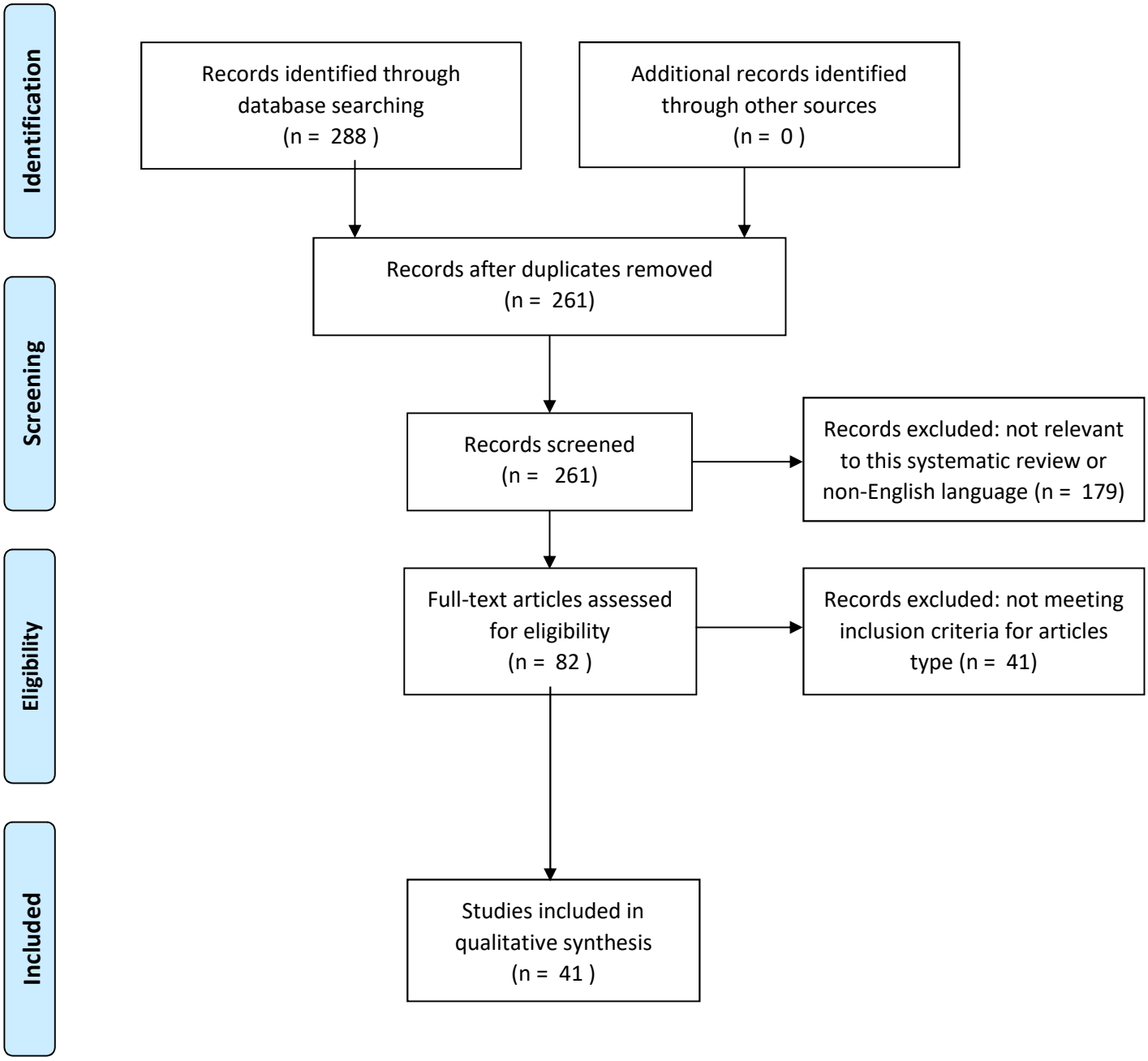

Figure 1. PRISMA flow diagram

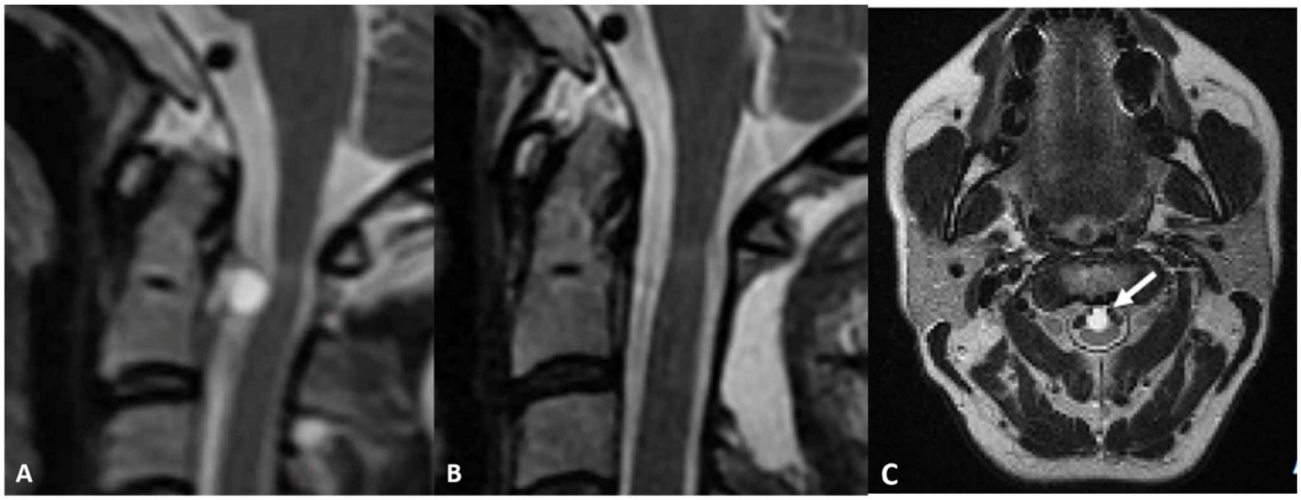

Figure 2. Sagittal T2 MRI. A hyper-intense cyst with spinal cord compression and intramedullary hyper-intensity is seen and follow-up after 4 years. The patient was pain free, with no clinical or radiographic evidence of recurrence/instability

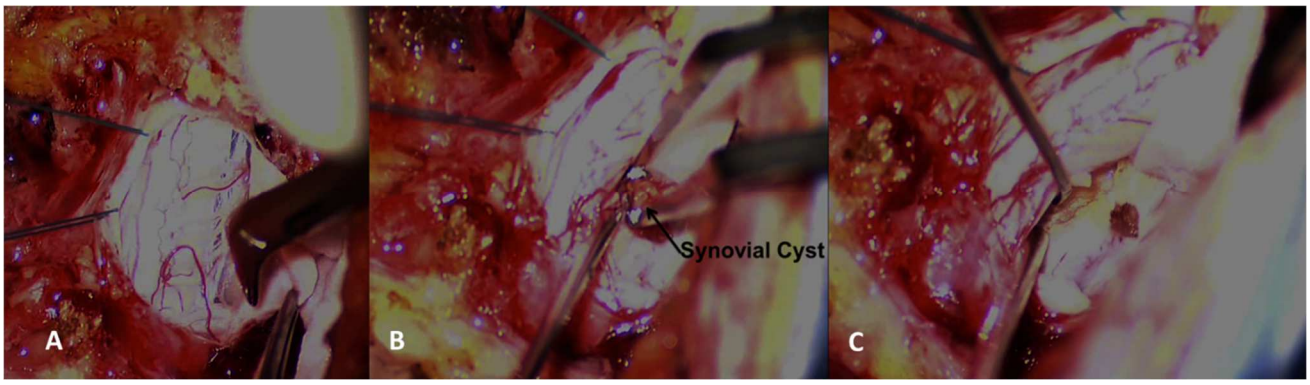

Figure 3. Steps of the surgical procedure for the removal of the synovial cysts: dural exposure (A); identification of the synovial cyst (B); removal of the cyst (C)

Table 1: Patients series demographic, pre- and postoperative functional status, FU features.

Table legend: mJOA: Japanese Orthopaedic association scale; FU: Follow-up; MTHS: months

#	age	gender	presenting symptoms	radiological findings	Preoperative Status		FU	Imaging	Postoperative Status	
					Nurick Scale	mJOA			Nurick Scale	mJOA
1	53	F	nuchalgia, bilateral hypoesthesia in upper and lower limbs, unsteady gait, tetraparesis	C1-C2 extradural cystic lesion located anteriorly in the midline posterior to the dens.	4	9	24 mths	Complete excision and not instability	2	14
2	64	F	unsteady gait	C1-C2 extradural cystic lesion located antero-laterally (right) to spinal cord, posterior to the dens	2	16	24 mths	Complete excision and not instability	1	17
3	59	M	nuchalgia, unsteady gait	C1-C2 extradural cystic lesion located antero-laterally (left) to spinal cord, posterior to the dens	2	15	24 mths	Complete excision and not instability	1	16
4	78	M	progressive unsteady gait, tetraparesis	C1-C2 cystic lesion located in the midline anteriorly to the spinal cord, posterior to the dens.	3	10	24 mths	Complete excision and not instability	2	11

authors and year	number of patients	age (years)/ mean age (range)	sex	Surgery	cyst resection	outcome
Onofrio and Mih, 1988[20]	1	73	M	DL and suboccipital craniectomy	total	fair
Miller et al. 1989[9]	1	67	F	DL C1/C2	total	fair
Choe et al. 1992[10]	1	61	F	TO + PF	total	good
Goffin et al. 1992[11]	1	65	M	DL C1/C3	total	good
Quaghebeur and Jeffree 1992[12]	1	82	F	DL	sub-total	good
Weymann et al, 1993[13]	1	-	-	DL	total	-
Birch et al, 1996[10]	5	72 (60-85)	-	TO + PF (2); DL (2); conservative treatment (1)	total (3) sub-total (1) none (1)	good
Kaufmann et al. 1996[14]	1	52	M	TO	total	good
Vergne et al. 1996[15]	1	64	F	DL C1	total	good
Fransen et al. 1997[16]	1	75	F	DL C1/C2	total	good
Akiyama et al. 1999[17]	1	51	F	DL + C1 PF	total	good
Aksoy et al. 2000[18]	1	61	M	DL C1/C2 + AF	none	-
Chang et al. 2000[19]	1	45	M	C1/C2 + PF	none	good
Cai et al, 2001[20]	2			TO + C1-C2 PF (2)	sub-total	-
Zorzon et al. 2001[21]	2	79(74-84)	F	DL C1/C2	total	good
Eustachio et al. 2003[22]	1	75	M	DL C1	subtotal	good
Morio et al. 2003[23]	1	71	F	C1/C2 PF	none	good
Okamoto et al. 2004[24]	1	72	M	DL C1/C2 + PF	subtotal	good
Sagiuchi et al, 2006[25]	1			C1-C2 PF	none	-
Cecchi et al, 2008[26]	1	80	-	Hard collar (Philadelphia)	none	good
Velan et al, 2008[27]	1			Percutaneous cyst aspiration (repeated twice)	sub-total	-
Elhammady et al. 2009[28]	1	67	F	DL C1	total	fair
Kirk and Pik, 2009[29]	1			TO + PF	total	
Marbacher et al. 2009[30]	1	60	M	TO + C0-C3 fusion	total	good
Aizawa et al, 2010[31]	1			hemilaminectomy, foramen magnum craniectomy	sub-total	
Harries et al, 2010[32]	1			far-lateral excision	sub-total	

Weng et al, 2010[33]	1			DL + craniocervical PF	none	
Henaux et al, 2011[34]	1			conservative (no treatment)	none	
Lyons and Birch, 2011[35]	11	76 (54-84)	-	TO + PF (11)	total	good
Van Gompel et al, 2011[36]	10	75,2 (54-81)	-	TO + PF (9); DL (1)	total	-
Lee et al, 2013[37]	3	68	-	TO + PF (3)	total	good
Parks et al, 2013[38]	1	-	-	FL + PF (transarticular screws)	-	-
Puffer et al, 2013[39]	3	78 (76-81)	-	PF without DL (2), TO + PF (1)	sub-total (1); none (2)	good
Sameshima et al. 2013[40]	1	-	-	hemilaminectomy, transcondylar resection	total	-
Sheen et al. 2013[41]	1	72	F	aspiration via TO approach + PF (cervico-occipital)	sub-total (aspiration)	good
Colasanti et al. 2014[2]	1	75	M	navigation-guided endoscope assisted posterior approach via a C1 hemilaminectomy	sub-total	good
Ikegami et al. 2015[42]	1	52	F	lateral atlantoaxial joint puncture	none	good
Ohnishi et al, 2015[43]	1			anterolateral approach with C1 post arch and C1 lateral mass resection	total	
Hartmann et al. 2016[44]	1	74	M	C1 hemilaminectomy and minimal suboccipital craniectomy	total	
Theodotou et al. 2016[45]	1	45	M	C1 partial laminectomy	total	good
Madhavan et al. 2018[46]	3	75 (70-81)	-	Transdural approach	Total	Good
present case series	**4**	63.5 (53-78)		**posterior decompression**	**total**	**good**

Table 2: literature review of case reports on the management of synovial cysts of the atlanto-axial segment. DL: decompressive laminectomy; AF: Anterior Fusion; PF: Posterior Fusion; TO: transoral decompression