A novel classification and algorithmic-based management of craniovertebral junction osteoarthrosis

ABSTRACT

Introduction: The objective of this study is to propose a novel classification and algorithmic-based management plan for craniovertebral junction osteoarthrosis (CVJOA).

Materials and Methods: A retrospective study was done based on prospective database of radiological studies and clinical history. Twenty symptomatic patients (12 females and 8 males) with a mean age of 54.8 years were identified with CVJOA. These patients underwent either nonsurgical treatment only or surgical intervention and had follow-up of at least 14 months. Classification of CVJOA is based on coronal deformity, rigidity, stability, and two modifiers. The main surgical procedures done in the surgical arm of these patients included C1–C2 fusion, C1–C2 facet distraction and fusion, and unilateral subaxial facet distraction, and posterior column osteotomy.

Results: All the twenty patients included in this study complained of either sub-occipital or upper neck pain and had radiological evidence of CVJOA. Seven patients improved with nonsurgical management and 13 underwent surgical intervention. Surgical recommendations for each type of CVJOA have been described with case examples, and algorithm for the management of CVJOA has been developed based on this study. Interobserver agreement on CVJOA classification was measured using kappa value statistics which showed moderate strength of agreement (0.467).

Conclusion: This study describes a novel classification and management of CVJOA based on algorithm and current surgical recommendations for each type of CVJOA.

Keywords: C1-C2 fusion, cervical pain, craniovertebral junction, facet distraction, osteoarthrosis

INTRODUCTION

Osteoarthrosis (OA) is a common degenerative joint disease. Involvement of the subaxial spine in this degenerative process has been reported, but craniovertebral junction involvement has not been reported frequently. OA can affect atlanto-axial facet joints (AAOA), atlanto-dental joint, and atlanto-occipital joints. Craniovertebral junction osteoarthrosis (CVJOA) can be divided into primary (idiopathic) and secondary due to inflammation, trauma, infection, and congenital deformities. Prevalence of radiographic AAOA is from 5.4% in the sixth decade to as high as 18% in the ninth decade of life. Changes of CVJOA on imaging include decreased joint space, subchondral sclerosis, and presence of osteophytes. These changes are not easily viewed on routine anteroposterior (AP) and lateral radiographs, therefore transoral radiographs are

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preferred. Computed tomography (CT) has high sensitivity and specificity in diagnosing CVJOA, and magnetic resonance imaging is also useful in the diagnosis of CVJOA and it can provide information about neurovascular structures as well.[1] Clinical manifestation of the symptoms in CVJOA can be variable. More than one-third of the CVJOA patients present with sub-occipital pain. Typical symptoms include unilateral suboccipital pain with limited range of movements in neck and pain that worsens with head movements. However, patients can present with atypical symptoms as well. Initial management of CVJOA consists of analgesics, immobilization, and physiotherapy. If patient´s symptoms do not improve after this initial management, intra-articular injections can be helpful in some cases. Surgical management with arthrodesis becomes an option if this initial noninvasive regimen (including intra-articular injections) fails to provide relief in symptoms.[2] No clear management of CVJOA is available because there are relatively few published studies on this topic. In these studies, a variety of surgical techniques were discussed without clear guidelines or indication for a particular technique. Furthermore, majority of these publications do not address the secondary causes of CVJOA including inflammation (rheumatoid arthritis), trauma, infection, and congenital deformities.[2-4] Furthermore, in some cases, CVJOA can lead to sagittal and coronal deformity, which can make management even more challenging. In this study, our objective is to share our experience of CVJOA patients' management, describe a novel classification of CVJOA, and propose an algorithmic approach to address this condition.

MATERIALS AND METHODS

After institutional review board approval (CAAE: 21225019.0.0000.5273), a retrospective study based on prospective database of radiological studies and clinical history was carried out. Twenty symptomatic patients (12 females and 8 males) were identified with CVJOA (during April 2011–March 2019). These patients were managed at the National Institute of Traumatology and Orthopedics and Spine Institute of Rio de Janeiro, Brazil. These patients ranged from 9 to 84 years of age, with the mean age of 54.8 years. Radiological studies and clinical history were used for inclusion criteria. Imaging-based findings of degenerative changes of the atlanto-axial joint (atlanto-dental and lateral mass) and atlanto-occipital joint were used to include the patients for this study. Clinical symptoms including unilateral high cervical pain, cervicogenic headache or occipital neuralgia, variable restriction of motion, and pain on head rotation or flexion were part of the inclusion criteria as well. The following variables were included in this study: age, sex, clinical history, radiological studies, applied treatment, and surgical complications. Patients who developed CVJOA idiopathically were grouped into primary CVJOA. Patients who developed CVJOA post trauma, post infection, post inflammatory conditions (e.g., rheumatoid arthritis), and post instability (e.g., congenital basilar invagination [BI]) were grouped into secondary CVJOA. These patients (primary CVJOA and secondary CVJOA) underwent either nonsurgical treatment only or surgical intervention after failed nonsurgical treatment. Nonsurgical treatment included anti-inflammatory medication, physiotherapy, collar immobilization, and intra-articular corticosteroid injection. However, if the neurological deficits were evident at the presentation, patients were evaluated for surgical management and nonsurgical trial was bypassed. All the patients included in this study had follow-up of at least 14 months. In this study, we proposed a novel classification for CVJOA and algorithmic approach for the management of CVJOA. We have described types of CVJOA based on coronal deformity, rigidity (ankylosis), stability, and two modifiers. The first modifier is based on the etiology of the OA. If the CVJOA patient has no history of any inflammatory (e.g., rheumatoid arthritis), infectious (e.g., tuberculosis), and traumatic and congenital deformity (e.g., congenital BI) condition, we added the modifier "P" which shows that the condition is primary. However, if the CVJOA patient has a history of any previously mentioned conditions, we added the modifier "S" which shows that the condition is secondary to some pathological process. The second modifier is based on neurological exam of the patient. In the absence of neurological deficits, the modifier N- was added and in the presence of neurological deficits, N+ was added with the main type of the CVJOA.

Definition of craniovertebral junction osteoarthrosis

CVJOA is defined as joint arthropathy of occipito-atlantal and atlanto-axial joints. This phenomenon can involve single or multiple joints including occipito-atlantal joint, atlanto-dental joint, and two C1–C2 lateral mass joints. CVJOA can be primary or secondary. Primary CVJOA patients have no history of any predisposing disease or event. Secondary CVJOA can be posttraumatic, postinfectious, postinflammatory condition (e.g., rheumatoid arthritis), and due to congenital deformity (e.g. congenital BI)

Types of craniovertebral junction osteoarthrosis

Type 1: Craniovertebral junction osteoarthrosis without coronal deformity is classified as Type 1

We recommend C1–C2 fusion with or without facet distraction by Goel–Harms technique for patients with CVJOA who are not responding to the conservative therapy and continue having cervicalgia/sub-occipital pain. The decision of performing facet distraction or not depends on the neurological exam of the patient. If a patient has neurological compromise, in our experience, it is better to perform facet distraction for reduction and stability of C1–C2 joint. An example of CVJOA Type 1 is shown in Figure 1.

Type 2: Craniovertebral junction osteoarthrosis with coronal deformity without fixed ankylosis

Fixed ankylosis can be between occipital and C1 joints or C1–C2 joints. If a patient has coronal deformity in the absence of fixed ankylosis, we classified it as Type 2. We recommend C1–C2 facet distraction for coronal correction for patients presenting with Type 2. This procedure not only reduces the pain but also corrects the coronal deformity which otherwise can cause significant loss of functionality due to coronal imbalance and psychological issues. An example of Type 2 is shown in Figure 2.

Type 3: Craniovertebral junction osteoarthrosis with coronal deformity and fixed ankylosis

As mentioned in the previous type, fixed ankylosis can be between occipital-C1 and C1-C2 joints and in either case, it will be classified as Type 3. In Type 3 CVJOA, unilateral subaxial facet distraction and posterior column osteotomy (PCO) is recommended. An example of Type 3 is shown in Figure 3.

Type 4: Craniovertebral junction osteoarthrosis with atlanto-axial instability is classified as Type 4 craniovertebral junction osteoarthrosis

We recommend C1–C2 fusion with or without facet distraction in this group of patients. As mentioned for Type 1, decision of whether performing C1–C2 facet distraction or not is based on preoperative neurological exam and occasionally, C1–C2 fusion only does not provide enough reduction and in those circumstances, C1–C2 facet distraction can provide better reduction in these patients. An example of Type 4 is shown in Figure 4.

Type 5: Craniovertebral junction osteoarthrosis with basilar invagination is classified as Type 5

C1–C2 facet distraction and fusion are recommended in these patients for the correction of vertical instability. A case example of Type 5 is shown in Figure 5.

Surgery procedure

Three most commonly performed surgical procedures are discussed here:

C1–C2 fusion

C1–C2 fusion was performed with C1 lateral mass and C2 pedicle screw fixation using Goel–Harms technique. Goel *et al*. first described this technique using monoaxial screws and plates[5] and Harm *et al*. modified the technique by using polyaxial screws and rods.[6] This technique provides reduction and stability to the atlantoaxial joint.

C1–C2 facet distraction and fusion

This technique was first described by Goel *et al*.[7] where they used stainless steel spacers within the C1–C2 facet joint which provided sustained traction and fixation of C1–C2. We did a modification in Goel technique of facet distraction

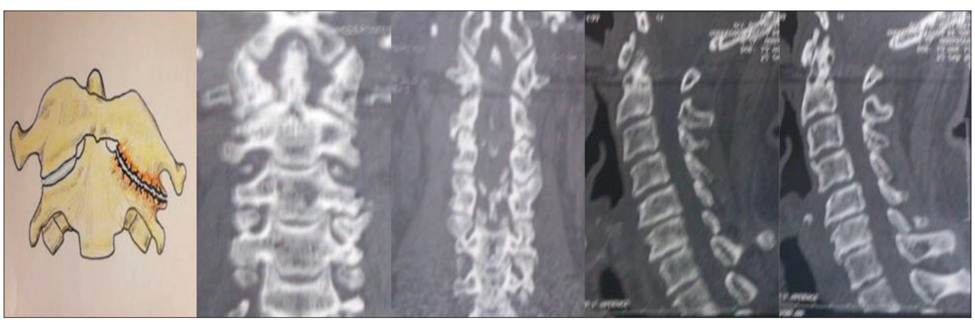

Figure 1: Craniovertebral junction osteoarthrosis Type 1, C1–C2 osteoarthrosis without deformity

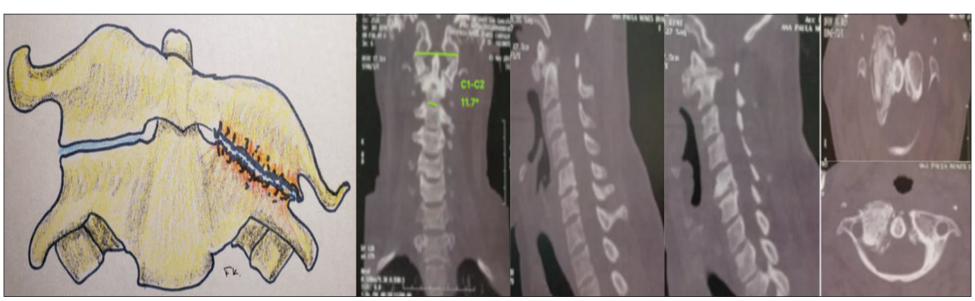

Figure 2: Craniovertebral junction osteoarthrosis Type 2, C1–C2 osteoarthrosis with coronal deformity without fixed ankylosis

and used polyetheretherketone (PEEK) cage rather than metal spacers. One significant limitation in using PEEK cage is the limited range of sizes. Therefore, if it is not possible to open and manipulate the C1–C2 facet joint enough to insert the PEEK cage, we use customized titanium mesh cage or autologous bone graft to fill the joint.[8-10]

Unilateral Subaxial facet distraction and posterior column osteotomy

Subaxial facet distraction has been described by Goel *et al*. for the treatment of single and multilevel cervical spondylotic radiculopathy and myelopathy.[11] PCO (Grade 2 osteotomy) has been described by Ames *et al*. for the deformity correction in the subaxial cervical spine.[12] In Grade 2 osteotomy, resection of both superior and inferior facets at any given spinal segment along with soft tissues and bony elements including ligamentum flavum, lamina, and spinous process is done. However, it is the first time that the combination of these two techniques in the mobile spine (C2–C3) is being described to correct the coronal deformity with fixed ankylosis (C0–C1 or C1–C2 fixed ankylosis). We performed subaxial facet distraction on the concave side of the deformity and PCO on the convex side to correct the coronal deformity in patients with fixed ankylosis.

RESULTS

Twenty patients with a mean age of 54.8 years (age range: 9–84) with CVJOA were studied. All the twenty patients complained of suboccipital or upper neck pain with variable range of motion, and all of them had radiological evidence of CVJOA. Three patients had symptomatic degenerative lesions in the subaxial cervical spine and one of them had signs and symptoms of myelopathy. Of these twenty patients, 11 had secondary CVJOA, three patients presented with fixed coronal deformity, and six had neurological deficits. Seven patients improved with nonoperative management and thirteen patients underwent surgical intervention. Of these 13 patients who underwent surgical intervention, one had a trial of intra-articular steroid injection and the symptoms did not improve and eventually, surgery was carried out. Two patients had vertebral artery injury preoperatively, one of them recovered completely, while the other developed ipsilateral visual impairment. One patient from the surgery arm developed C2 nerve dysesthesia which sustained for 8 months postoperatively, and another patient developed pseudoarthrosis and implant failure. This patient who developed pseudoarthrosis had undergone occipital-cervical fusion and posterior decompression. When we performed this surgery, we did not have experience of C1–C2 facet distraction and fusion. This patient underwent a second surgery for decompression and reconstruction (transoral). These data are shown in Table 1. Classification of CVJOA has been developed based on coronal deformity, rigidity (ankylosis), and stability. In addition, the underlying etiology and neurological exam have been addressed as modifiers in this novel classification. Table 2 shows the CVJOA classification. The main surgical interventions in this study included the following three procedures, (1) C1–C2 fusion (Goel–Harms technique), (2) C1–C2 facet distraction and fusion, and (3) unilateral subaxial spine distraction and PCO. Surgical recommendations were developed for each type of CVJOA. Table 3 shows the recommended current surgical procedure for each type of CVJOA. All patients who underwent surgical procedures had radiological evidence of solid fusion and stable fixation on 6-month and 1-year follow-up radiological exams. All patients who underwent surgical intervention had improved neurological functions and alleviation in pain symptoms postoperatively. We also developed an algorithm for the management of CVJOA which is based on our experiences [Figure 6].

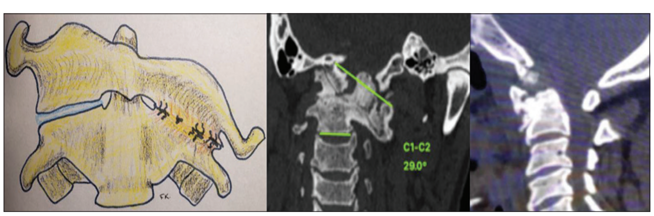

Figure 3: Craniovertebral junction osteoarthrosis Type 3, C1–C2 osteoarthrosis with coronal deformity and fixed ankylosis

Interobserver agreement

Interobserver agreement for the CVJOA classification was measured using Kappa value statistics in the SPSS software, version 27, IBM company, New York, USA. The data were collected from two consensus meetings (1 week apart) where five cases (one case of each type) were presented


Figure 4: Craniovertebral junction osteoarthrosis Type 4, C1–C2 osteoarthrosis with atlanto-axial instability

Table 1: Demographic and clinical data of the patients included in the study

Patient number	Age	Sex	Joints involved	Primary or secondary CVJOA	Neuro exam	Coronal deformity on radiological studies	CVJOA type	Symptomatic subaxial spine OA	Conservative management/ intra-articular steroid injection	Surgical management	Complications
1	80	Female	C0–C1, C1–C2 LM (UL)	Primary	N−	No	1PN−	No	Conservative	None	N/A
2	54	Male	C1–C2 AD	Primary	N−	No	1PN−	No	Conservative	None	N/A
3	49	Male	C1–C2 LM (UL), AD	Secondary (trauma)	N−	No	1SN−	No	Conservative	None	N/A
4	29	Male	C1–C2 LM (UL), C0–C1	Primary	N−	No	1PN−	No	Conservative	None	N/A
5	63	Female	C1–C2 AD	Primary	N−	No	1PN−	No	Conservative	None	N/A
6	84	female	C1–C2 LM (UL)	Primary	N−	No	1PN−	No	Intra-articular steroid injection	None	None
7	9	Female	C1–C2 LM (UL), AD	Secondary (trauma)	N−	Yes	3SN−	No	Conservative	Trans-oral release and C1–C2 fusion	None
8	61	Female	C1–C2 LM (BL)	Secondary (RA)	N+	No	5SN+	Yes	No	C1-C2 facet distraction with PEEK cage and fusion + subaxial decompression and fusion	None
9	67	Male	C1–C2 LM (UL) with fixed ankylosis, AD	Secondary (trauma)	N−	Yes	3SN−	No	Conservative	Unilateral subaxial facet distraction + PCO	Vertebral artery injury with postoperative neurological recovery
10	56	Male	C1–C2 LM (UL)	Primary	N−	No	1PN−	No	Intra-articular steroid injection	C1–C2 fusion and facet distraction with PEEK cage	C2 nerve dysesthesia for 8 months
11	51	Female	C1–C2 LM (UL)	Primary	N−	Yes	2PN−	No	Conservative	C1–C2 facet distraction with bone graft and fusion	None
12	30	Male	C1–C2 LM (BL)	Secondary (RA)	N+	No	4SN+	No	No	C1–C2 fusion	None
13	75	Female	C0–C1 (UL)	Primary	N−	No	1PN−	No	Conservative		N/A
14	42	Female	C1–C2 LM (BL)	Secondary (os odontoideum)	N+	No	4SN+	No	No	Occipito-cervical fusion and posterior decompression	Pseudoarthrosis implant failure, recurrence of neurological deficits. 2nd intervention, trans-oral decompression and reconstruction
15	61	Female	C1–C2 LM (BL)	Secondary (RA)	N−	No	5SN−	No	Conservative	C1–C2 facet distraction with PEEK cage and fusion	None
16	39	Male	C1–C2 AD	Primary	N+	No	1PN+	Yes	Conservative	Posterior C1–C2–C5 fusion + ACDF	None

Contd...

Table 1: Contd...

Patient number	Age	Sex	Joints involved	Primary or secondary CVJOA	Neuro exam	Coronal deformity on radiological studies	CVJOA type	Symptomatic subaxial spine OA	Conservative management/intra-articular steroid injection	Surgical management	Complications
17	61	Female	C0–C1–C2 Clivus to C2, LM (BL)	Secondary (tuberculosis)	N−	Yes	3SN−	No	Conservative	Unilateral subaxial facet distraction + PCO	None
18	57	Female	C1–C2 LM (UL)	Secondary (congenital BI)	N+	No	5SN+	No	No	C1–C2 facet distraction with PEEK cage and fusion	Vertebral artery injury during C2 screw insertion. Ipsilateral visual impairment
19	65	Female	C1–C2 LM (UL)	Secondary (RA)	N−	Yes	2SN−	No	No	C1–C2 fusion with autologous bone graft + Gallie instrumentation	None
20	64	Male	C0–C1 LM (UL)	Secondary (congenital BI)	N+	No	5SN+	Yes	No	C1–C2 facet distraction with PEEK cage and fusion + extended subaxial spine fusion	None

CVJOA - Craniovertebral junction osteoarthrosis, ACDF - Anterior cervical discectomy and fusion, PCO - Posterior column osteotomy, RA - Rheumatoid arthritis, BI - Basilar invagination, N/A - Not available, LM - Lateral mass, UL - Unilateral, BL - Bilateral, AD - Atlantodental

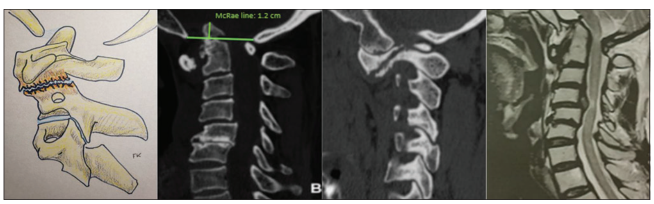

Figure 5: Craniovertebral junction osteoarthrosis Type 5, C1–C2 osteoarthrosis with basilar invagination

after explaining the classification. In this meeting, four spine surgeons (three orthopedic surgeons and one neurosurgeon) took part. The mean kappa value showed moderate strength of agreement (0.467) for the main types of CVJOA and almost perfect agreement on both modifiers (1.00). However, in the second meeting, the interobserver agreement for the main type of CVJOA showed significant strength of agreement (0.623). We believe that observers' familiarity with the classification, better understanding, and knowledge of radiological parameters in the second meeting improved the statistics. These meetings were conducted online using Zoom application because of restrictions during the COVID-19 pandemic.

DISCUSSION

As a cause of debilitating chronic pain in adults, CVJOA affects the quality of life and occasionally leads to depression as well. As life expectancy improves, it can be anticipated that CVJOA will become more prevalent and will have more significant impact on active population. As limited studies are available on this topic, primary care providers are not quite familiar with CVJOA and routine cervical radiographs also do not provide the details of craniovertebral junction clearly. Therefore, patients suffering from this disorder remain in pain for long duration before receiving definite diagnosis and proper management.[3,4,13]

In this study, we proposed a classification of CVJOA and recommended surgical interventions for each type of CVJOA. If there are no neurological deficits on physical exam and there is no compression and instability evident on radiological exams, we recommend nonoperative management including intra-articular corticosteroid injection [Figure 7] and if this management does not provide significant symptomatic relief, surgery can be indicated. However, if a patient has already signs and symptoms of neurological deficits or there is radiological evidence of compression or instability on initial presentation, surgery can be performed without a trial of nonsurgical treatment.

Degenerative changes in the joints cause instability, therefore it is important to manage these individuals by providing stability, that is, fusion. Previous studies have shown that C1–C2 arthrodesis can achieve good fusion without causing

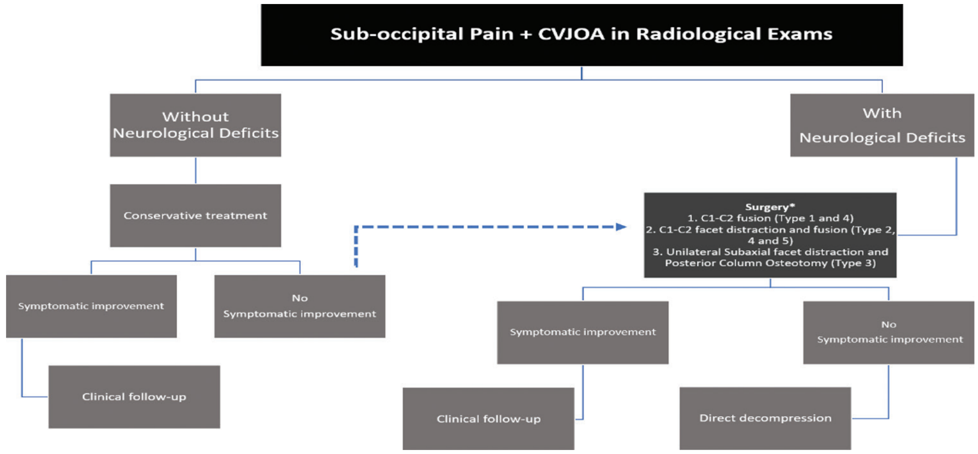

Figure 6: Algorithm showing management plan for craniovertebral junction osteoarthrosis. *:Appropriate surgical procedure is chosen on patient's craniovertebral junction osteoarthrosis type and neurological exam

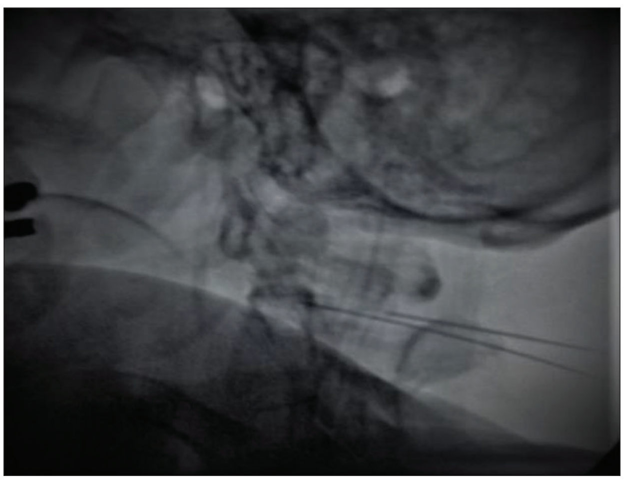

Figure 7: Intra-articular steroid injection in craniovertebral junction osteoarthrosis

Table 2: The craniovertebral junction osteoarthrosis classification

CVJOA classification
Type 1: CVJOA without coronal deformity
Type 2: CVJOA with coronal deformity and without fixed ankylosis*
Type 3: CVJOA with coronal deformity and with fixed ankylosis*
Type 4: CVJOA with AAI
Type 5: CVJOA with BI

*Fixed ankylosis can be between occipital-C1 or C1–C2 joints.
CVJOA - Craniovertebral junction osteoarthrosis, AAI - Atlantoaxial instability, BI - Basilar invagination

hyperlordosis of the C1–C2 segment and subaxial cervical kyphosis.[5,6] The prevalence of primary CVJOA is more common, and studies can be seen addressing this primary condition, but secondary CVJOA due to trauma, infection, inflammatory conditions (e.g., rheumatoid arthritis), and congenital deformities (e.g., congenital BI) have not been reported to a significant extent, and we have presented these secondary CVJOA cases and options for managing these cases as well.[13-15]

One of the important concepts to understand here is that OA is a degenerative progression in the joint after inflammatory changes. It is not merely an inflammation which can be referred as "osteoarthritis." OA can include erosion of the joint surfaces, decreased joint spaces, formation of subchondral cysts, subchondral sclerosis, osteophytes formation, pseudotumor, and ankylosis. These degenerative changes can be primary or secondary. If CVJOA occur without any predisposing factors, we can use the term "primary osteoarthrosis." When such changes occur due to secondary factors such as inflammatory conditions (e.g., rheumatoid arthritis), posttrauma, postinfection, and congenital deformities, it can be termed as "secondary osteoarthrosis." Although management of primary and secondary CVJOA is the same, surgical management only changes based on CVJOA type, neurological exam, coronal deformity, and instability in this study. This additional modifier (P or S) gives health-care professionals better understanding and the underlying culprit (if secondary CVJOA) is not ignored while addressing CVJOA.

Presence or absence of neurological deficits plays an important role when planning for surgical intervention. In CVJOA patients who also have atlanto-axial instability or BI, C1–C2 fusion will be sufficient to provide symptomatic relief and stability in the absence of neurological deficits. However, patients who have developed neurological deficits will require C1–C2 facet distraction and fusion as C1–C2 facet distraction provides indirect decompression. Therefore, the modifier "N" has an important role in not only defining the status of the patient rather but also influencing the surgical intervention in specific situations.

Occasionally, patients with CVJOA present with retro-odontoid pseudotumor which not only causes pain but can also lead to neurological deficits. In such patients, our preferred surgical intervention is C1–C2 facet distraction and fusion and the reasons for this procedure are to decompress and provide stability with fusion. However, occipital-C1–C2 fusion can also be done in patients with CVJOA and pseudotumor to provide solid fusion and stability of craniovertebral junction. We performed occipito-cervical fusion in one patient [patient number 14 in Table 1] when we did not have experience of C1–C2 facet distraction. This patient developed pseudoarthrosis and implant failure, and trans-oral decompression and reconstruction was done as second procedure in this patient. Now, our preferred surgical intervention is C1–C2 fusion with or without facet distraction for these patients as this technique provides better biomechanical stability.[16,17] Decision of whether facet distraction should be performed is based on the presence of neurological deficits in these patients and the quality of reduction achieved with C1–C2 fusion preoperatively.

Another important scenario which requires attention is the involvement of the subaxial spine in CVJOA patients.

Table 3: The recommended current surgical procedures for each type of craniovertebral junction osteoarthrosis

Recommended current surgical procedures for CVJOA	
CVJOA types	**Recommended procedures**
1	C1–C2 fusion with or without facet distraction*
2	C1–C2 facet distraction and fusion
3	Unilateral subaxial facet distraction + PCO
4	C1–C2 fusion with or without facet distraction*
5	C1–C2 facet distraction and fusion

*In both situations we opt for C1-C2 fusion and facet distraction if there are neurological deficits and in the absence of neurological deficits, we mostly do C1-C2 fusion only. CVJOA - Craniovertebral junction osteoarthrosis

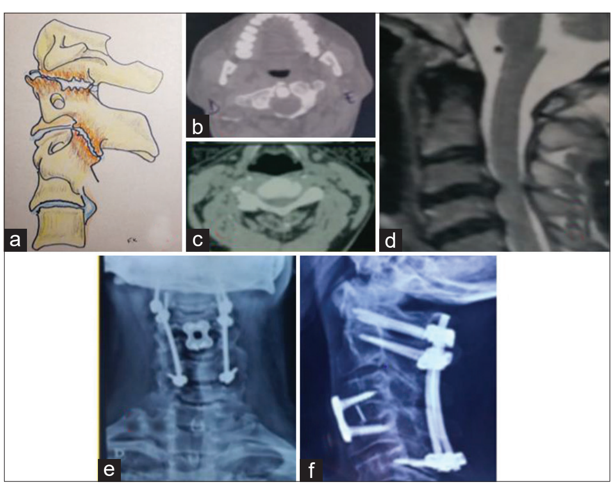

Figure 8: (a-d) Schematic diagram of subaxial spine osteoarthrosis and preoperative radiological studies in a patient with craniovertebral junction osteoarthrosis type 1PN+. (e and f) Postoperative radiographs with C1–C2–C5 fusion (posterior) and anterior cervical discectomy and fusion

Previous studies have shown that patients with CVJOA can present with signs and symptoms of myelopathy,[13,18] hence we recommend addressing myelopathy by staged subaxial segment decompression if it is present because only C1–C2 fusion can provide significant pain relief in this condition, but still these patients will continue to have disability due to myelopathy. Case example: Figure 8a-d shows the schematic diagram of CVJOA with subaxial spine OA and preoperative radiological studies of a patient with CVJOA type 1PN+. Figure 8e and f also shows postoperative radiographs showing C1–C2–C5 (posterior) fusion and anterior cervical discectomy and fusion (ACDF).

If subaxial spine also has degenerative changes and these changes are not causing neurological deficits, but significant pain is present, we recommend extended fusion to the subaxial spine. Furthermore, the other common procedure in the management of CVJOA is the C1–C2 facet distraction and fusion. We made a small modification in Goel technique and we started using PEEK cage instead of metal spacers. These PEEK cages provide good distraction, less subsidence, increased fusion rate, and better visualization on radiological exams.[8,9] Occasionally, it is not possible to place PEEK due to the lack of availability in smaller sizes, in those circumstances, we either use customized titanium mesh (around 2.5 mm in height) or autologous bone graft to fill the facet joint. A previous study[19] has shown that a cage of 2.5 mm in height used in subaxial spine stabilization with good biomechanical outcomes, and we chose the same height while customizing titanium mesh for C1–C2 facet distraction. Case example: Figure 9a-c shows preoperative radiological studies of a patient who presented with signs and symptoms of CVJOA type 5SN+. Figure 9d-f also shows postoperative radiological studies in this patient. This patient underwent C1–C2 facet distraction with PEEK cage and fusion.

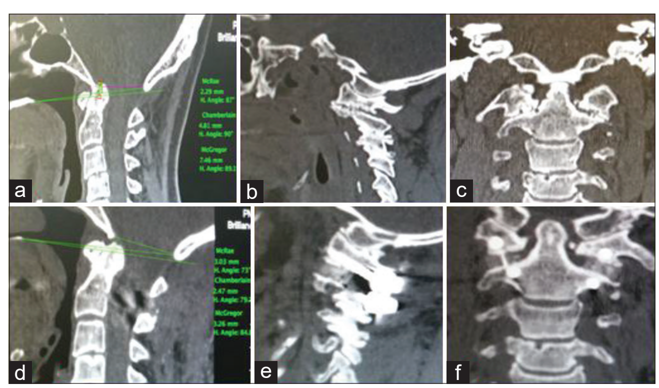

Figure 9: Preoperative computed tomography images (a-c) and postoperative computed tomography images showing C1–C2 facet distraction with polyetheretherketone cage (d-f) of a patient with craniovertebral junction osteoarthrosis type 5SN+

Similarly, if the patient presents with CVJOA Type 3, another option is anterior release of the ankylosed bone with trans-oral approach. Although we do not perform this surgery frequently, in some special circumstances, anterior decompression is indicated especially when release of ankylosis is not possible via posterior approach only. In our data, a 9-year-old female patient [patient number 7 in Table 1] developed CVJOA secondary to traumatic rotatory dislocation of C1–C2 with fixed ankylosis. This patient underwent trans-oral release of the ankylosis and C1–C2 fusion. However, now our preferred approach is unilateral subaxial facet distraction and PCO for patients who present with CVJOA Type 3.

Of the three procedures for the surgical management of CVJOA, to our knowledge, it is the first time that unilateral subaxial facet distraction and PCO combination is being reported for the management of CVJOA with coronal deformity and fixed ankylosis. In this procedure, subaxial facet (mostly C2–C3 segments) distraction on the concavity for the elongation and PCO is performed on the convexity. This combination not only provides significant pain relief in patients with Type 3 CVJOA but also reduces the deformity. Case example: Figure 10a-d shows preoperative photograph and radiological studies of a patient with CVJOA type 3SN-. This patient had coronal deformity and fixed ankylosis between clivus-C1–C2. Figure 10e-h also shows postoperative photograph and radiological exams of this patient. This patient underwent unilateral subaxial (C2–C3) facet distraction and PCO. It is important to be careful during this procedure as overdistraction can cause cervical kyphosis and there is a risk of vertebral artery injury as well. We had one case [patient number 9 in Table 1] who suffered partial vertebral artery injury during this procedure, but he recovered completely over time.

Furthermore, if the appropriate surgical procedure fails to provide improvement in neurological deficits or if patient develops new neurological deficits, one must analyze the compression and if there is compression, direct decompression should be carried out. If the compression is posterior, direct decompression can be done posteriorly, that is, laminectomy and if the compression is anterior, decompression should be done anteriorly.

Another important point to mention here is the understanding of ankylosis on radiological exams while classifying CVJOA. Fixed ankylosis is not merely a presence of osteophyte; in ankylosis, one can see complete bone bridges and synostosis between the adjacent bones. Therefore, it is important for spine surgeons to be comfortable with these parameters so that they will not confuse fixed ankylosis with other nonfixed degenerative changes. Dealing craniovertebral junction (CVJ) is challenging, and the learning curve is steep, but fair understanding of CVJ anatomy and the pathological processes helps in approaching and managing these disorders. Regardless of the approach or technique the surgeon is performing, it is crucial to be capable of managing all the possible complications and surgical techniques of CVJ.

CONCLUSION

This novel classification of CVJOA describes and classifies CVJOA with practical approach. Clear recommendations for management and current surgical procedure for each type of CVJOA along with management plan based on algorithmic approach will guide spine surgeons significantly to deal with this challenging pathology effectively. This study will also help to open new approach toward further research in this area.

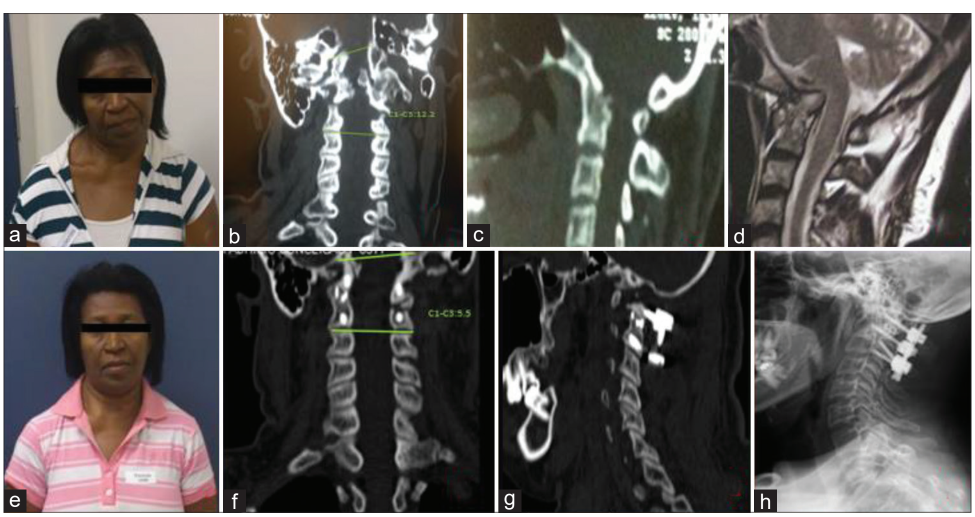

Figure 10: Preoperative photograph of a patient and computed tomography scan showing fixed ankylosis between clivus-C1–C2 (a-d). Postoperative photograph and radiological studies (e-h)

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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