Review Article

# Atlantoaxial instability secondary to *Bartonella henselae* osteomyelitis managed surgically by atlantoaxial instrumentation: A case report and systematic review


## ABSTRACT

Cat scratch disease (CSD), caused by *Bartonella henselae*, may atypically present with vertebral osteomyelitis. Antibiotic regimens are tailored to presentation, which is markedly variable and not well defined for any atypical disease. In cases of spinal instability, the use of antibiotics alone may not be sufficient. Atlantoaxial instability caused by osteomyelitis is a rare complication of CSD. In this report, we describe the rare case of vertebral osteomyelitis complicated by atlantoaxial instability, requiring both antibiotics and atlantoaxial fusion. We discuss our case, surgical technique, rationale, and outcome. In addition, we conducted a systematic review of the literature of vertebral osteomyelitis in pediatric secondary to *B. henselae*. A 2-year-old child presented with a 2-month history of irritability, fever, and rigid neck pain along with a recent history of feline exposure. Physical examination revealed cervical tenderness and decreased range of motion. Computed tomography (CT) showed osteolysis of the right C1 lateral mass and pars articularis; T1-weighted magnetic resonance imaging with contrast showed enhancement around the right C1 lateral mass. The titer for *B. henselae* was high. A diagnosis of cat scratch osteomyelitis with cervical instability was made, for which the patient underwent surgery with atlantoaxial fusion. Postoperative imaging demonstrated resolution of the contrast-enhanced lesion. At 6-year follow-up, the patient showed no signs of residual complications from surgical intervention with a solid fusion. Our review revealed 44 cases of pediatric CSD vertebral osteomyelitis. Conservative management with antibiotic employed in 86% while antibiotics with surgical intervention in 14% of the cases. Surgical intervention was most often in the form of incision for drainage and decompression without fusion. Average follow-up 10 months with 86% achieved complete resolution. Cervical instability caused by osteolysis is a rare complication of CSD. This can subsequently lead to vertebral instability, requiring definitive surgical intervention.


**Keywords:** Atlantoaxial instability, *Bartonella henselae*, cat scratch disease, cervical instrumentation, cervical osteomyelitis


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## INTRODUCTION

Cat scratch disease (CSD) is a zoonosis with marked clinical variability caused by the alphaproteobacteria *Bartonella henselae*, *Bartonella clarridgeiae*, or *Afipia felis* most often transmitted via direct inoculation from a feline vector.[1-3] Typical CSD is generally characterized by a self-limited low-grade fever and regional lymphadenopathy that resolves over weeks to months.[4] Less commonly, an atypical presentation of CSD represented exclusively by *B. henselae* infection may affect one or more systems. Various manifestations include Parinaud's oculoglandular


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syndrome (4.0%), appendicular arthropathy (5.5%), CSD encephalopathy (2-4%), and a mixture of axial and appendicular osteomyelitis, tendonitis, neuralgia, and neuroretinitis (all <1%).[5]

CSD osteomyelitis tends to be osteolytic and is reported to occur in 0.1%–0.3% of all cases, with preferential involvement of the axial skeleton.[5-9] Targeted prolonged antibiotic therapy leads to rapid clearance of disease, occasionally requiring adjunct surgical incision and drainage of associated abscesses with favorable prognosis.[10-14] Pediatric CSD atlantoaxial osteomyelitis is exceedingly rare, and conservative treatment may be possible in the absence of osteolytic spinal instability;[15] however, data for conservative versus surgical management are limited.

We review available literature for vertebral osteomyelitis and report an unusual case of atypical CSD osteolytic atlantoaxial disease in an immunocompetent child with resulting instability treated successfully with instrumented fusion and postoperative antibiotics.

## METHODS

We present a toddler case of atlantoaxial osteomyelitis secondary to confirmed CSD complicated with instability successfully treated with surgical atlantoaxial fusion. In addition, we performed a systematic literature review without meta-analysis in accordance with preferred reporting items for systematic reviews and meta-analysis regarding pediatric vertebral osteomyelitis secondary to *B. henselae* infection querying the National Library of Medicine PubMed and Embase databases using the combination of medical subject headings terms "*B. henselae* OR cat-scratch disease AND osteomyelitis." Abstracts were then screened using the inclusion criteria of English language reports that highlighted the treatment modalities and therapies. We excluded articles that (1) reported CSD osteomyelitis outside the vertebral column or (2) pertained to adult (age >18) cases of vertebral osteomyelitis. A search flow diagram is provided in [Figure 1].

## RESULTS

### Case presentation

An immunocompetent 2-year-old male with no past medical history who presented 2 months of progressive neck pain, irritability, and torticollis initially noticed following a minor fall from height. Per family, the child fell out of the back seat of a nonmoving truck and struck his head and neck 2 months prior. The patient was brought into a local facility 4 days later where plain cervical films were negative for acute process. The complaints as mentioned above progressed and represented 2 months later when computed tomography (CT) of the cervical spine demonstrated a right-sided, type III atlas fracture with comminuted displacement of the lateral mass [Figure 2], and the patient was transferred to our facility.

On arrival, the patient was febrile at 39.4°C and was without focal deficit intact. The cervical spinal examination was significant for paraspinal tenderness. Acute-phase reactants were mixed, with an elevated erythrocyte sedimentation rate of 30 mm/h and no elevation in C-reactive protein.

Contrasted magnetic resonance imaging (MRI) of the cervical spine additionally showed acute inflammation and an enhancing osteolytic lesion about the right C1 lateral mass with extensive involvement of the paravertebral soft tissues extending anteriorly behind the right longus colli muscle and dissecting caudally in the prevertebral space as far as C4 [Figure 3]. Vascular studies were normal. The patient was noted to have a type I atlantoaxial rotary subluxation secondary to compromise of the lateral atlantoaxial joint.

The patient was diagnosed with atlantoaxial instability as evidenced by type I rotary subluxation in the setting of a displaced unilateral pathological type III atlas fracture secondary to an undefined destructive inflammatory lesion. Management options, including instrumented stabilization, external stabilization, and treatment with antibiotics alone, were discussed extensively with the patient's family, who ultimately favored surgical intervention given the underlying instability arising from his lesion.

Intraoperative inspection demonstrated a clearly affected right C1–C2 joint. The affected joint was carefully dissected, and clear fluid came out of the joint space during dissection. The C1 lateral mass was destroyed entirely, the pieces of bone were removed, and soft tissue was debrided. There was no abscess to be incised or drained. Forward movement of C1 was observed with live fluoroscopy during drilling, which demonstrated that it was completely unstable. Due to the severe bony destruction in the right C1 lateral mass with an inadequate healthy bone for screw purchase, the patient underwent a modified Brooks procedure using sublaminar titanium wires about C1 and C2 on the right as well as to avoid placement of hardware into the infected bone. The construct was interposed with the placement of an autologous T8 rib graft. On the left side, the atlantoaxial complex was stabilized with segmental instrumented fusion with C1 lateral mass and C2 pars articularis screw placement. Postoperative X-ray confirmed appropriate alignment [Figure 4].

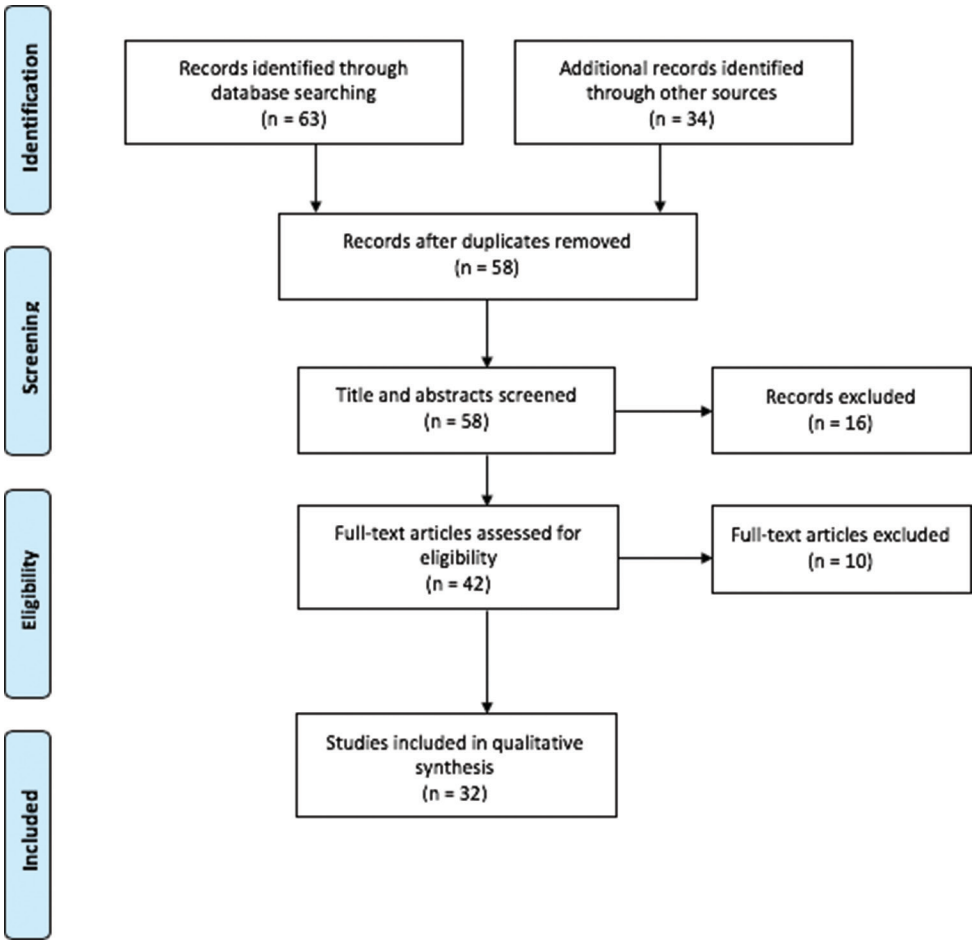


**Figure 1: Flowchart of literature review**

Histopathological examination of resected osseous and inflammatory tissue demonstrated chronic polymorphous inflammatory infiltrate and associated necrosis about the lateral atlantoaxial joint complex suspicious for ongoing bacterial infection [Figure 5]. Immunohistochemistry for neoplasia was negative. *B. henselae* titers returned positive for immunoglobulin G (IgG) (1:1280) and negative for IgM, indicative of chronic CSD. No organisms grew on intraoperative or blood cultures. Upon further history, the family confirmed a prior history of feline exposure.

Postoperatively, the rigid atlantoaxial support was supplemented with a Minerva cervicothoracic orthotic brace. The patient started broad-spectrum antibiotic therapy in the immediate perioperative period with vancomycin and piperacillin–tazobactam and was transitioned to a 6-week oral regimen of azithromycin and rifampin upon completion of cultures. The postoperative course was uncomplicated, and the patient was discharged on postoperative day 9. Six-week MRI demonstrated an interval decrease in inflammation in the prevertebral region [Figure 6] and preserved alignment of the vertebrae, solid fusion, and resolution of inflammatory changes with no associated neurological or musculoskeletal abnormalities at 6 years [Figure 7].

## Literature review

The initial literature search yielded 97 publications. Of these, 88 manuscripts were reviewed and found to meet inclusion criteria. After exclusion, a total of 32 reports[9-40] were ultimately selected for full review [Figure 1].

Included reports described 44 cases of pediatric CSD vertebral osteomyelitis [Table 1]. Males (50%) and females (50%) were affected equally, and age averaged 8 years.[9-40] Patients were most often immunocompetent (94%) and presented with symptoms of fever (91%), back or neck pain (52%), and lymphadenopathy (47%) [Table 1]. Spinal regions involved were most often thoracic (57%), followed by lumbar (45%), sacral (18%), and cervical (7%) [Table 1].

Patients were treated with antibiotic therapy alone (81%), antibiotics with adjunct surgical intervention (14%), or conservatively (5%) [Table 1]. Surgical intervention was most often in the form of incision and drainage [Table 2]. The

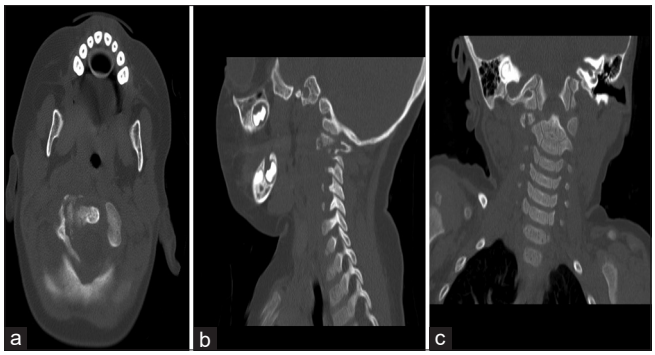


**Figure 2: Computed tomography of the cervical spine (a) axial, (b) sagittal, and (c) coronal images showing a right-sided type III atlas fracture with an overhang of C1 on C2**

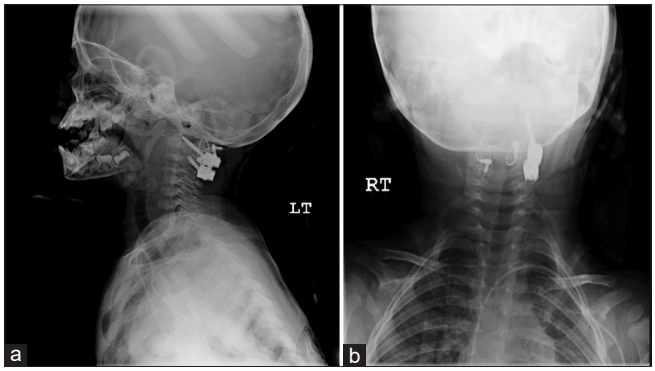


**Figure 4: Postoperative lateral (a) and anteroposterior (b) cervical spine radiograph showing atlantoaxial instrumented fusion with sublaminar wiring and autograft placement on the right and lateral mass and pars screw fixation on the left**

average follow-up time was 10 months with 86% seeing complete resolution.[9-40]

## DISCUSSION

### Diagnosis of cat scratch disease

Atypical CSD has a variable clinical presentation. In cases reviewed, regional lymphadenopathy present in 47% of cases and localized axial pain may provide a clue to spinal involvement however symptoms are generally nonspecific.[8,41] Patients with osseous involvement often present with fever and pain related to the affected bone with a median of 10 days of illness prior to diagnosis. The patient in our case would likely not have had their cervical spine initially imaged if not for an inciting trauma. When CSD is suspected, a definitive diagnosis can be achieved through conventional bacteriologic culture methods, immunoserology, or nucleic acid amplification methods.[42] In clinical laboratories, direct examination, serology, and culture have become the mainstay of presumptive diagnosis with serology, in particular rising preferred method in cases of atypical CSD involving the neuroaxis.[42] Accordingly, the patient in our case was

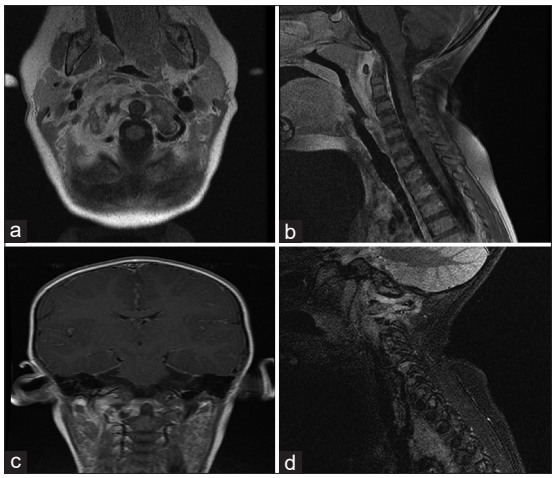


**Figure 3: Preoperative magnetic resonance T1-weighted imaging with gadolinium contrast (a) axial view, (b) sagittal view, (c) coronal view and (d) sagittal view with short TI inversion recovery showing osteolysis of the C1 vertebra with prevertebral soft tissue mass extending from the level of odontoid process to C4 as well as alignment of cervical vertebrae with an increase in inflammatory soft tissue changes**

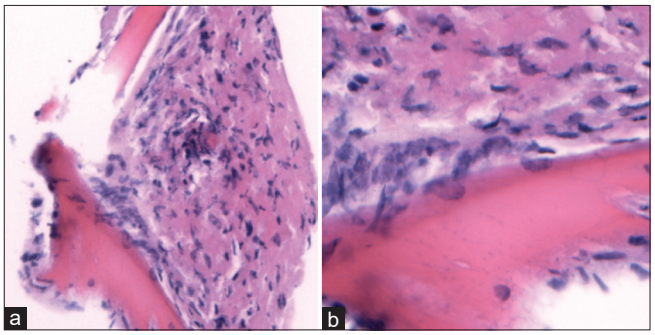


**Figure 5: Histopathologic findings on hematoxylin-eosin staining. (a) Low-power view showing reactive fibrous tissue with a polymorphous mononuclear infiltrate. (b) High-power view showing mononuclear cells with a subset possessing enlarged nuclei and low mitotic activity suggesting chronic infection**

diagnosed via serologic detection of IgG directed against *B. henselae*, suggesting chronic active infection.

In immunocompetent hosts, the histologic changes typically show granulomatous necrosis and suppurative abscess formation.[4,38] Involved osseous tissue tends to show a polymorphous inflammatory infiltrate consisting mainly of granulocytes, as was noted in our patient.[22,32]

Typical radiographic findings for vertebral osteomyelitis include MRI signal changes and enhancement of involved tissues.[15] Osteomyelitis may also be multifocal or localized,[36] and osteolytic lesions, if present, are rarely detected on plain films (0.17%).[5,15] Lesions tend to appear hypoechoic on ultrasound and hypodense with an enhancing rim on contrasted CT.[39] Spinal imaging should be used to dually evaluate diagnostic features of osteomyelitis as well as the degree of instability.[39]

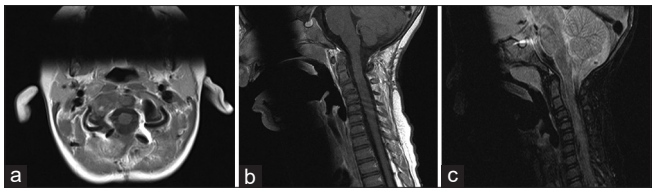


**Figure 6: Postoperative 6-week magnetic resonance contrasted T1-weighted sequences (a) axial view, (b) sagittal view, and (c) sagittal view short TI inversion recovery showing resolution of soft tissue and bony changes plus maintenance of spinal architecture**

## Nonsurgical management of pediatric *Bartonella* vertebral osteomyelitis

The majority of reviewed cases of pediatric vertebral osteomyelitis secondary to CSD were treated with antibiotics alone.[9-40] As mentioned above, the guidelines for antibiotic regimens for atypical CSD are not well defined.[36] The disease of the subaxial cervical spine and atlantoaxial complex in the pediatric population is exceedingly rare.[11,13-15,39] As such, there is further ambiguity concerning the appropriate antibiotic management of patients, such as the one presented in our case, but there is no reason to believe the regimens reported successful in other spinal segments would not be equally effective in the cranial spine. Different combinations of antibiotics including macrolides, cephalosporins, and rifampin have been reported for a mean duration of 28 days.[15]

Azithromycin is frequently part of the treatment regimen,[15] and the combination of doxycycline and rifampin is commonly chosen based on reported case data.[36] For uncomplicated CSD, a 2011 meta-analysis found no statistically significant difference in cure rates or time to achieve cure between antibiotic therapy and placebo.[43] For systemic CSD, increased severity and concern for complications may warrant treatment. However, the confusion is compounded by a limited number of reports, each with varying treatments, outcome measurements, and follow-up periods. Further investigation, preferably prospective in nature, is necessary to characterize the appropriate therapeutic approach to atypical CSD.

Reported cases describing the use of antibiotics alone in CSD vertebral osteomyelitis focus on symptoms indicative of compression of neural elements.[39] For example, pediatric disease of C3 with mass effect in a neurologically asymptomatic case of CSD was reported to be treated successfully with external rigid orthosis and 6 weeks of azithromycin and rifampin.[39] While a spinal epidural abscess has been historically associated with high morbidity and mortality, some authors argue that advances in early detection may allow for nonsurgical antibiotic management in the absence of neurologic deficits.[36]

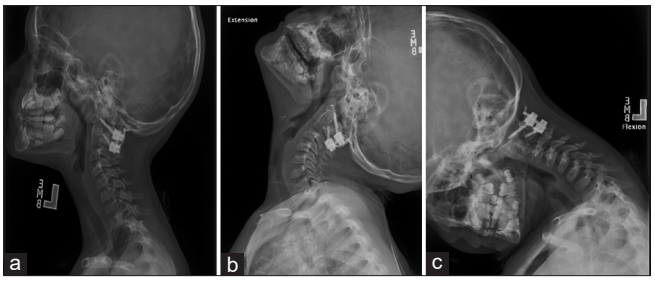


**Figure 7: Follow-up cervical x-ray neutral (a), extension (b), and flexion (c) dynamic films at 6 years, showing C1-C2 posterior spinal instrumentation and fusion. Hardware is intact. No evidence of screw failure or dynamic instability. The vertebral body heights are maintained**

Follow-up data for vertebral CSD osteomyelitis showed resolution of fever in most patients by hospital day 0, 1, or 2, and resolution of pain at an average of 9.7 days.[15] Patients improved rapidly in many cases with no anti-inflammatory use and no recurrences and resolution of lesion were noted on MRI.[15] Of the three cases found in our literature review with no treatment, two had complete resolution at 9-week and 20-month follow-ups, and one case had persistent osteolytic lesions.[16,17,24] Multiple cases presented with an associated abscess and a complete resolution with only antibiotic therapy.[36,38,39] In one interesting case of cervical osteomyelitis, an abscess at the level of C3 was noted with mass effect, and at 1-year follow-up with conservative management, the lesion had resolved and with no evidence of cervical instability on imaging.[39] Nonsurgical management with oral antibiotics appears reasonable in spinal CSD in the absence of compressive neurologic symptoms or radiographic or clinic evidence of spinal instability.

## Surgical management of *Bartonella henselae* vertebral osteomyelitis

Reported surgical interventions in pediatric CSD vertebral osteomyelitis largely consist of adjunct noninstrumented resection or incision and drainage of associated paraspinal or evacuation of compressive intraspinal abscesses.[10-14,22] We found no prior reports of instrumented fusion. For all patients who required surgical treatment, the lesions were either located in the cervical (50%) or thoracic (50%) spine, and all achieved complete resolution at an average follow-up time of 6.7 months.[10-14,22]

As with our case, patients presenting with cervical osteomyelitis requiring some degree of surgical intervention often presented with progressive pain and torticollis nonresponsive to antibiotics and were noted to have a deteriorating neurological exam.[11,13,14] As mentioned, surgical intervention is generally directed at decompression of neural elements, as necessitated by paravertebral masses, epidural abscesses and associated myelopathy, and inflammatory material extension into the foramen

**Table 1: Summary of all pediatric cases of *Bartonella henselae* vertebral osteomyelitis**

| Reference | Age/sex | Location | Treatment (duration) | Outcome (duration of follow-up) |
|---|---|---|---|---|
| Shanon *et al*., 1989[16] | 11/male | L | None | CR (20 months) |
| Karpathios *et al*., 1990[17] | 8/female | T | None | Osteolytic lesions (21 months) |
| Larsen and Patrick, 1992[18] | 10/male | L | CRO+OXA | ND |
| Cohen-Abbo *et al*., 1992[19] | 9/male | T/L | CPX, ERY, GEN | CR (1 year) |
| Fretzayas *et al*., 1993[20] | 12/male | T | AMX+CFC, CTX | Persistent narrowing of T5-6 intervertebral space (2 months) |
| Koranyi, 1994[21] | 4/male | T | RIF+CFU+ERY, TMP/SMX+AMP/SUL+GEN, RIF+TMP/SMX | CR (10 weeks) |
| Hopkins *et al*., 1996[23] | 6/male | L | ND | ND |
| Gallemore and Worley., 1996[24] | 10/male | T/L | None | CR (9 weeks) |
| Robson *et al*., 1999[25] | 9/female | T | FLU+GEN, CPL, GEN, TMP/SMX+RIF | Expansion (7 weeks), CR (1 year) |
| Hulzebos *et al*., 1999[26] | 10/female | L | RIF+CIP | Spinous process destruction (3 months) |
| Liapi-Adamidou *et al*., 2000[27] | 7/Female | L | AMX, AMP+CTX+MET, CTX | CR (5 months) |
| Ruess *et al*., 2000[28] | 12/female | L | ERY | CR (15 months) |
| Fretzayas *et al*., 2001[29] | 14/male | T or L | CTX+GEN | CR (2-6 months) |
| Fretzayas *et al*., 2001[29] | 6/female | T or L | CTX | CR (2-6 months) |
| Fretzayas *et al*., 2001[29] | 9/female | T or L | CTX+GEN | CR (2-6 months) |
| Del Santo *et al*., 2002[30] | 2/female | L | CRO, CPR, AZM | CR (2 years) |
| Del Santo *et al*., 2002[30] | 2/female | L | TEI+CRO, CFC | CR (1 year) |
| Pocheville *et al*., 2002[31] | 12/male | T | ERY | CR (9 months) |
| de Kort *et al*., 2006[32] | 9/female | L/S | RIF+TMP/SMX, RIF+TMP/SMX | Pain relapse (6 months), CR (3 years) |
| Kodama *et al*., 2007[33] | 11/female | T/L | CLR, CRO+MIN, SUL/CFP, AZM, MIN | CR (18 months) |
| Rozmanic *et al*., 2007[34] | 11/male | T | CRO, AZM+RIF | ND |
| Al-Rahawan *et al*., 2012[35] | 7/male | T | CFD, AZM | Compression deformity (5 months) |
| Dusser *et al*., 2013[9] | 13/female | S | AZM | CR (10 months) |
| Dornbos *et al*., 2016[36] | 5/female | T | AZM, DOX+RIF | CR (6.5 weeks) |
| Rafferty *et al*., 2017[37] | 5/male | T | CFZ, CIP+RIF | ND |
| Kopsidas *et al*., 2018[38] | 11/male | T | CLI, DOX+GEN, DOX+RIF | CR (3 months) |
| Akbari *et al*., 2018[39] | 7/male | C | AMC, AZM+RIF | CR (1 year) |
| Harry *et al*., 2018[40] | 3/female | T | CFU, AZM | ND |
| Erdem *et al*., 2018[15] | 5/female | C/T/S | RIF+DOX | PR (6 weeks) |
| Erdem *et al*., 2018[15] | 12/female | T/L/S | RIF+DOX | PR (2 days) |
| Erdem *et al*., 2018[15] | 7/male | L | AZM | PR (1 week) |
| Erdem *et al*., 2018[15] | 5/female | T | RIF+DOX | PR (1 week) |
| Erdem *et al*., 2018[15] | 7/male | T/S | TMP/SMX, RIF+CIP | PR (18 days) |
| Erdem *et al*., 2018[15] | 3/male | L/S | TMP/SMX+RIF, CIP | PR (1 week) |
| Erdem *et al*., 2018[15] | 5/female | T | CLI, AZM | PR (8 days) |
| Erdem *et al*., 2018[15] | 5/female | L | AZM | ND |
| Erdem *et al*., 2018[15] | 10/female | C/T/L/S | RIF+DOX, PRD | Fever recurrence (2 weeks), PR (18 days) |
| Erdem *et al*., 2018[15] | 10/male | S | RIF+AZM | PR (5 days) |

Age in years, Gender, location of lesion, treatment, and outcome including follow up duration. Age given in years. Imaging refers to studies that demonstrated findings.
M indicates Male, F - Female, mo - Month(s), wk - Week(s), d - Day(s), yr - Year(s); CR - Complete resolution, C - Cervical vertebra, T - Thoracic vertebra, L - Lumbar vertebra, S - Sacral vertebra, AMX - Amoxicillin, AMC - Amoxicillin-clavulanic acid, AMP - Ampicillin, AMP/SUL - Ampicillin/Sulbactam, AZM - Azithromycin, CTX - Cefotaxime, CRO - Ceftriaxone, CLR - Clarithromycin, CLI - Clindamycin , CIP - Ciprofloxacin, CFC - Cefaclor, CFU - Cefuroxime, CFD - Cefdinir, CFZ - Cefazolin, CPL - Cephalothin, CPR - Cephradine, CPX - Cephalexin, DOX - Doxycycline; ERY - Erythromycin, FLU - Flucloxacillin, GEN - Gentamicin, MET - Metronidazole, MIN - Minocycline, OXA - Oxacillin, PRD - Prednisone, RIF - Rifampin, SUL/CFP - Sulbactam/Cefoperazone, TEI - Teicoplanin, TMP/SMX - Trimethoprim-Sulfamethoxazole, PR - Pain resolution, CR - Complete resolution, ND - No data or not documented

with radiculopathy.[10-13] Spinal instability is very rare, and destructive involvement of the lateral atlantoaxial joint was reported in just one case.[14] Consistent with worsening clinical pictures as described above, reported failures of conservative management of compressive lesions found enlargement of abscesses and worsening cord compression after short-term follow-up.[13] Similarly, Hussain and Rathore reported a case of thoracic osteomyelitis and psoas mass with neural foramen compression, and after 1 week of treatment with clindamycin and gentamicin, the psoas mass had grown with epidural extension.[12]

As mentioned, patients with suppurative compressive disease were treated by therapeutic CT-guided drainage, open surgical drainage, or open evacuation of paravertebral or epidural abscesses.[10-14,22] Two cases of cervical osteomyelitis

**Table 2: All prior cases of *Bartonella henselae* vertebral osteomyelitis that required surgical management**

| Author | Age | Sex | Clinical presentation | Immune status | Location | Imaging | Diagnostic test | Treatment (duration) | Outcome (duration of follow-up) |
|---|---|---|---|---|---|---|---|---|---|
| Bernini *et al*., 1994[22] | 5 | Female | 2 weeks: Fever, back pain, lower extremity paresthesias, and numbness | ND | T8-10, paravertebral abscess | XR, CT, BS | Biopsy, *Afipia* felis PCR, *Bartonella henselae* PCR (−) | Nafcillin (>18 days), irrigation and debridement of the paravertebral abscess via an open posterior approach, laminectomy at T9, laminotomies at T8,10, CRO (3 weeks) | CR (2 years) |
| Abdel-Haq *et al*., 2005[10] | 5 | Male | 3 weeks: Fever, headache, abdominal pain, torticollis, neck stiffness | Immunocompetent | T4-T7, epidural abscess, liver, spleen | CT, BS, MRI | Surgical specimen histopathology, Warthin-Starry (−), IgG (+), IgM (−) | CRO+VAN (ND), surgical drainage of mass on the right of the T4-T7 vertebrae suggestive of epidural abscess, CLR (2 weeks), TMP/SMX (8 weeks) | CR (8 weeks) |
| Vermeulen *et al*., 2006[11] | 9 | Female | 3 days: Fever, neck pain, torticollis | ND | C4-6, paravertebral mass | BS, MRI | Surgical specimen histopathology, IgM (+), PACR | Drainage of purulent material within thick fibrous capsule of prevertebral lesion at C5-6 via anterior cervical approach, AMC (3 weeks) | CR (3 months) |
| Hussain and Rathore, 2007[12] | 3 | Male | 1 week: Fever, back pain, stiff gait, LAD | ND | T12, paravertebral mass | CT, BS, MRI | IgG (+), IgM (−), PCR | Cefazolin (ND), CLI+GEN (8 weeks), TMP/SMX (4 weeks) | Extension of mass (3 weeks), treated with CT-guided drainage; CR (3 months) |
| Tasher *et al*., 2009[13] | 5 | Male | 2 weeks: Fever, tonsillitis, neck pain, torticollis, LAD | Immunocompetent | C3-5, epidural abscess | CT, MRI | PCR, IgM (+), IgG (−) | Cloxacillin+GEN+CRO (ND), surgical drainage of 6.3 mm epidural abscess with C3-5 laminectomy, GEN+RIF (4 weeks), RIF+AZM (6 weeks) | CR (9 weeks) |
| Mirouse *et al*., 2015[14] | 14 | Male | ND: Fever, cat scratch, neck pain, torticollis, LAD | Immunocompetent | C2 | CT, MRI | Serological tests (−) then (+) | AZM (ND) prior to admission, surgical drainage of purulent joint effusion around the right C1-C2 articulation, AMC+CIP+GEN (15 days), AMC+CIP (2.5 months) | CR (6 months) |
| Present case | 2 | Male | 2 months: Neck pain, irritability, torticollis, drooling | Immunocompetent | C1 | XR, CT, MRI | Bone fragment histopathology, IgG (+), IgM (−) | Surgical fusion of C1/C2, VAN+piperacillin-tazobactam (9 d), VAN+CRO (13 days), AZM+RIF (6 weeks) | CR (6 years) |

Age, Gender/Sex, clinical presentation, immune status, location of lesion, significant imaging, diagnostic test, treatment, and Outcomes including duration of follow-up.
Age given in years. Imaging refers to studies that demonstrated findings. M indicates Male, F - Female, mo - Month(s), wk - Week(s), d - Day(s), yr - Year(s), LAD - Lymphadenopathy, C - Cervical vertebra, T - Thoracic vertebra, XR - Radiography, CT - Computerized tomography, BS - Bone scintigraphy, MRI - Magnetic resonance imaging, PCR - Polymerase chain reaction, AMC - Amoxicillin-clavulanic acid, AZM - Azithromycin, CRO - Ceftriaxone, CLR - Clarithromycin, CLI - Clindamycin, CIP - Ciprofloxacin, GEN - Gentamicin, RIF - Rifampin, TMP/SMX - Trimethoprim-Sulfamethoxazole, VAN - Vancomycin, CR - Complete resolution, ND - No data or not documented

received drainage of purulent material alone from an anterior or posterior approach, one with a paravertebral lesion with foraminal extension at C5–C6 and another with an effusion near the C1-C2 articulation.[11,14] A case of cervical osteomyelitis with an epidural abscess causing cord compression received a laminectomy as well as drainage of the abscess.[13] Of the three cases with thoracic osteomyelitis, two underwent surgical resection of an abscess and the third a complete laminectomy secondary to osteolytic involvement.[10,22] The last case of thoracic vertebral osteomyelitis was initially treated conservatively, but after enlargement of a paravertebral mass with 3 weeks of antibiotic therapy, it was drained by CT guidance.[12] These patients completely recovered after their procedures without radiographic evidence of progressive or severe instability, so instrumented fusion was not indicated. Such cases demonstrated symptomatic improvement shortly after surgical treatment and resolution of neurologic symptoms within 6 weeks.[11] Furthermore, abscesses have completely resolved radiographically with no sequelae within 6 weeks

after surgery.[13] Unlike these cases, our pediatric patient did not present with an abscess requiring drainage but instead presented with atlantoaxial instability and rotary subluxation requiring fixation and fusion.

Other management options of CSD-related cervical instability in pediatric patients include external orthosis, fixation, or traction as an adjunct or alternative to surgical intervention.[11,14] Vermeulen *et al*. reported a case with the usage of a rigid collar and amoxicillin–clavulanate treatment following drainage of the abscess, while Mirouse *et al*. used traction and immobilization via halo orthosis for 3 months with concurrent amoxicillin–clavulanate, ciprofloxacin, and gentamicin administration following posterior drainage of the abscess.[11,14] CSD osteomyelitis of the atlantoaxial complex with underlying instability provides a challenging clinical scenario necessitating surgical stabilization of instability in addition to prolonged antibiotic administration.[1-3] Unfortunately, data on C1–C2 instability in CSD osteomyelitis are limited, and the use of external fixation in atlantoaxial instability may result in failure of treatment with a possible risk of worsening spinal deformity and neurological deficit, and it may be inferior to internal fixation in some cases.[44]

### Present case

Our present case demonstrates CSD osteomyelitis affecting the atlantoaxial joint and causing severe instability as shown by fracture type with type I atlantoaxial rotary subluxation and severe motion observed intraoperatively. Infection of the atlantoaxial joint is a very rare entity,[45] and CSD osteomyelitis is also very rare with largely undefined treatment strategies. While our review demonstrates that many cases of CSD osteomyelitis may be very mild and treated conservatively and in some cases with progressed and worsening neurologic symptoms, it may necessitate surgical decompression by laminectomy or incision and drainage. Similarly, in our case, symptoms progressed with cervical deformity and mechanical pain, indicating that surgical intervention is most likely necessary for cervical stabilization and fixation; thus, all options had to be explored. Moreover, the rarity of a septic atlantoaxial joint represents a challenge to treatment. Still, surgery has been shown to be safe if necessary to protect the spinal cord from unstable elements.[45] Placement of screws in an infected area is controversial beside poor bone purchase; therefore, lateral mass screw placement was avoided in our case, and we used modified Brooks instead on the infected side. Due to the progression of our patient's symptoms, the severity of atlantoaxial instability, and the unpredictability of CSD osteomyelitis, we believe that early surgical intervention with fusion was the most appropriate treatment option. Furthermore, the use of a rigid collar or halo orthosis may have been inappropriate in the setting of a progressive infection with underlying instability in a potentially noncompliant child of only 2 years, and there are reports of failure of external fixation in unstable atlantoaxial fractures.[44] Due to the rarity of this disease in the cervical spine, there is not enough evidence to predict natural fusion of joint space and nonprogression of the disease with conservative treatment alone. After discussion of management options with the patient and family, instrumented atlantoaxial fusion was successfully employed via a hybrid Brooks and Harms procedure on the right and the left sides, respectively. The favorable postoperative course of the patient on long-term follow-up with continued construct stability and normal developmental progression provides evidence that atlantoaxial fusion is a viable treatment option in appropriately selected pediatric patients with atlantoaxial CSD.

## CONCLUSION

Atypical CSD with chronic vertebral osteomyelitis secondary to *B. henselae* infection can often be managed with prolonged antibiotic regimens and adjunct noninstrumented surgical evacuation of a compressive inflammatory lesion with a favorable prognosis in the pediatric population. We report a case of instability at the atlantoaxial complex that was treated successfully with atlantoaxial instrumented fusion without significant morbidity on long-term follow-up.

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### Conflicts of interest

There are no conflicts of interest.

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