Regression of an atlantoaxial degenerative articular cyst associated with subluxation during conservative treatment

Case report and review of the literature

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The presence of an atlantoaxial degenerative articular cyst is rare; when present this lesion extends posteriorly to the dens, causing cervicomedullary compressive myelopathy. The authors describe a symptomatic case of this lesion associated with atlantoaxial subluxation in a 76-year-old man. The patient’s neurological symptoms resolved and corresponded to a reduction in the size of the cyst. After 8 months of continued conservative treatment, in which the patient wore a Philadelphia collar, the cyst spontaneously regressed. Subsequently, a C1–2 posterior fusion was performed to treat the atlantoaxial subluxation.

KEY WORDS • atlantoaxial articular cyst • atlantoaxial subluxation • craniocervical junction • synovial cyst

AN atlantoaxial degenerative articular cyst not due to spinal trauma or a collagen disease, such as clinically and serologically proven rheumatoid arthritis, is very rare. The lesion often extends posteriorly to the dens and causes cervicomedullary compression.1–8,11–13,16 Since the first report of this cyst by Onofrio and Mih13 in 1988, to the best of our knowledge only 18 cases have been reported in the literature.1–8,11–13,16 Although surgical management leads to a good neurological outcome and no recurrence of the cyst, the natural history of atlantoaxial degenerative articular cysts is unknown. We report a symptomatic case associated with atlantoaxial subluxation, which spontaneously regressed after conservative therapy; we also discuss the pathogenesis and surgical strategy for treating such a lesion.

Case Report

History and Examination. This 76-year-old man with no significant medical history presented with weakness and numbness in all extremities as well as gait disturbance that developed during a 5-month period.

The patient was alert and his cranial nerves were intact. Motor examination revealed spastic tetraparesis; manual muscle testing demonstrated 4/5 strength in all extremities. All deep tendon reflexes were hyperreactive, and the Babinski sign was present bilaterally. Sensory examination demonstrated dysesthesia in the patient’s hands and toes. A laboratory examination of the patient’s blood revealed no remarkable abnormality indicative of inflammation, and no rheumatoid factor or tumor marker. A dynamic flexion cervical radiograph showed atlantoaxial subluxation (Fig. 1). There was no C-1 or C-2 erosion, fracture, or congenital anomaly. Cervical MR imaging demonstrated a well-circumscribed extradural cyst extending posteriorly to the dens and compressing the upper cervical cord. The mass was slightly hypointense on T1-weighted images (Fig. 2A and B) and hyperintense on T2-weighted images (Fig. 2C and D). The patient refused our recommendation that he undergo C1–2 posterior fusion and resection of the cyst, but accepted external fixation with a Philadelphia collar.

Treatment. Three months after introduction of the Philadelphia collar, the patient’s dysesthesia decreased in all extremities, and MR imaging demonstrated significant cyst reduction. At 4 months after treatment commenced, the patient’s motor and sensory disturbances had completely disappeared. Follow-up MR images obtained after 8 months of treatment demonstrated almost complete regression of the cyst (Fig. 3). On the basis of the patient’s clinical course and a review of the serial MR imaging findings, the diagnosis of an atlantoaxial degenerative articular cyst associated with atlantoaxial subluxation was considered most likely. There was no cyst recurrence after 2 years of continued conservative treatment, but the atlantoaxial subluxation remained. The patient complained
of the inconvenience of wearing a brace and opted to undergo surgery.

Operation and Postoperative Course. After 2 years of conservative treatment, we performed a C1–2 posterior fusion in which an atlantoaxial posterior fixation system (3XS; Kisco DIR, Paris, France) was placed and an iliac bone graft implanted. There was no recurrence of the cyst or atlantoaxial subluxation on radiography (Fig. 4) or MR imaging after the patient was discharged from the hospital.

Discussion

Spinal synovial and ganglion cysts that present as juxtafacet cysts commonly develop in degenerative facet joints of the cervical or lumbar spine. They are pathologically subdivided according to whether their synovial epithelial lining is connected to a joint capsule; however, differences in the histopathological appearance may be encountered in the same lesion over time. With regard to cases in which the atlantoaxial joint is affected, some authors have proposed that the more general term “atlanto-
toaxial degenerative articular cyst” be applied rather than “juxtafacet cyst” because the specific anatomical structure is not a true joint. In all reported cases the patient had no medical history of spinal trauma or a clinically or serologically proven collagen disease such as rheumatoid arthritis. Including our patient, 13 were women and six were men, ranging in age from 8 to 85 years (mean 68 years). The interval between the onset of symptoms and diagnosis ranged from 3 weeks to 2 years. Pathologically, the cyst walls consisted of fibrous or cartilaginous tissue and usually included whole or local synovial epithelium, although in some cases this was lacking. The cysts comprised amorphous and mucinous material. On MR images these pathological characteristics appeared hyperintense on T1-weighted images and hypo- or hyperintense on T2-weighted images. An epidural venous plexus occasionally appeared as a sharp thin rim of enhancement surrounding the cyst wall. Cervical radiography and computed tomography studies showed no erosion or fracture of the odontoid process. An association of chronic excessive stress or minor damage between the transverse portion of the cruciate ligament and the dens has been surmised. In our case, the pathogenesis of the atlantoaxial articular cyst was considered to be related to chronic excessive stress due to atlantoaxial subluxation, caused by degeneration related to aging.

Retrospectively, the surgical treatment in 18 of the 19 patients in the literature could be classified into two groups based on radiological findings: one group had atlantoaxial subluxation and the other did not. In patients with atlantoaxial subluxation, both posterior decompression and C1–2 posterior fusion were performed. In our case, we performed a C1–2 posterior fusion using a device and implantation of an iliac bone graft after cyst regression. In patients without atlantoaxial subluxation, however, anterior or posterior decompression combined with cyst removal via a transoral or posterolateral approach was chosen. These surgical procedures resulted in excellent neurological outcomes and there was no cyst recurrence in any patient. Interestingly, three of four patients with atlantoaxial subluxation (including ours) did not undergo resection of the cysts, which disappeared within 6 weeks and 2 months, or within 8 months (in our case). We conclude that neck immobilization with the Philadelphia collar as well as C1–2 posterior fusion resolved the chronic excessive stress between C-1 and C-2, resulting in regression of the cyst. Although surgical management may be recommended for symptomatic atlantoaxial degenerative articular cysts in selected patients with atlantoaxial subluxation the cyst may regress after external neck brace therapy. However, as in our case, a C1–2 posterior fusion after confirmed cyst regression eventually may be needed to prevent cyst recurrence.

References

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