Forestier disease associated with a retroodontoid mass causing cervicomedullary compression

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O R I G I N A L A R T I C L E

Object. Forestier Disease (FD) is a progressive skeletal disorder affecting predominantly older men. It is also known as diffuse idiopathic skeletal hyperostosis (DISH) and is characterized by massive anterior longitudinal ligament calcification that forms a bridge on the anterior border of the thoracic and subaxial cervical spine. To the authors’ knowledge, retroodontoid masses associated with FD have not been described.

Methods. Five patients with FD and multilevel subaxial cervical fusion were treated for retroodontoid masses and cervicomedullary junction (CMJ) compression. There were four men and one woman (mean age 73 years, range 54–86 years). All patients suffered progressive neurological symptoms resulting from anterior compression of the CMJ.

Four patients underwent combined transoral resection of the ligamentous mass followed by an occipitocervical fusion procedure. One patient with circumferential CMJ compression underwent a posterior decompression and occipitocervical fusion. Histopathological examination of the mass showed hypertrophic degenerative fibrocartilage. Early postoperative neurological improvement was noted in all patients. The follow-up period ranged from 4 to 19 months. At the end of the follow-up period, four patients experienced neurological improvement. One patient died 3 weeks postsurgery of pulmonary complications.

Conclusions. The osseous elements of the occipitoatlantoaxial complex are not directly affected by FD. The ligamentous structures of the odontoid process, however, are exposed to significantly altered biomechanics resulting from fusion of the subaxial cervical spine associated with FD. Stress-induced compensatory ligamentous hypertrophic changes at the craniovertebral junction cause CMJ compression and subsequent neurological deterioration. This previously undescribed entity should be considered in patients with FD or DISH who present with progressive quadriparesis. Transoral decompression and posterior fusion are often needed in patients with large masses and severe progressive neurological deficits. Selected patients with smaller masses and milder neurological symptoms may be treated with posterior fusion alone.

KEY WORDS • Forestier disease • retroodontoid mass • cervicomedullary compression • diffuse idiopathic skeletal hyperostosis

F orestier disease, also known as DISH,22,23 and occasionally as hyperostotic spondylosis and ankylosing hyperostosis, was originally described in 1950 by Forestier and Rotes-Querol. They coined the term “ankylosing hyperostosis of the spine” to describe a spectrum of rheumatological disorders found in the elderly population, especially men in their sixth to seventh decade of life. This disorder classically involves marked calcification of the ALL and absence of other degenerative changes.

They described the condition as “usually painless, and therefore latent.” The disease process has several different presentations, however. The most common presenting symptoms of cervical FD are dysphagia and, less frequently, dyspnea, resulting from local mechanical compression of the esophagus and trachea.7,10,11 In the rare reported cases of FD associated with significant neurological deficit, the deficit is usually due to focal cervical and thoracic spinal canal stenosis.12,21,24 The extradural compression occurs secondary to focal fibrous proliferation of the ligamentum flavum, the origin of which is unclear.

Common causes of anterior CVJ compression include RA, Klippel–Feil syndrome, pseudogout, and basilar invagination.4,17,19,20,27,29 Although the pathophysiology of FD is unclear, it is not known to be associated with conditions that could cause such compression. To our knowledge, severe ligamentous hypertrophy and odontoid region pan- nus formation causing cervicomedullary compression has not been described in FD patients. In this report we discuss five patients with FD and retroodontoid masses and describe the pathophysiology of the mass formation and the treatment of this uncommon condition.

Clinical Material and Methods

Patient Population

During the 2-year period between 1995 and 1997, five
patients with FD were evaluated for CVJ compression symptoms in the Comprehensive Spine Center at the University of California, Los Angeles. There was no coexisting significant rheumatological, posttraumatic, or degenerative disease processes. A detailed retrospective evaluation of the medical records and radiological studies was performed. Characteristics in all patients fit the classic description of FD, although one patient with focal C4–5 DDD is included because no other sites of significant DDD were present. There were four men and one woman (mean age 73 years, range 54–86 years). A history of osteoarthritis of the bilateral lower extremities was noted in one patient.

**Presenting Symptoms and Signs**

All five patients suffered from significant neurological deficits when initially assessed. The majority of the patients exhibited myelopathic symptoms including hyperreflexia, spasticity, and a positive Babinski sign. To define the degree of disability, each patient was classified using the ASIA impairment scale (Table 1).

**Diagnostic Investigation**

All five of these elderly patients underwent a detailed medical evaluation preoperatively with focus on nutritional status, as well as on their respiratory and cardiac status. Preoperative and postoperative swallowing evaluation with a modified barium swallow was performed. Preoperative swallowing tests demonstrated abnormal status in two cases, one of which improved after surgery (Table 2). Additionally patients underwent preoperative evoked potential testing and intraoperative monitoring. Imaging studies included plain radiography, CT scanning, and MR imaging of the affected areas. The radiological criteria defined by Resnick and Niwayama for FD was used in the assessment of this group. A retroodontoid soft-tissue mass expanding into the canal and causing significant stenosis of the CVJ was present in all patients.

**Clinical Presentation**

The most prominent complaints were gait disturbance, weakness, and paresthesias in the extremities (Table 3). Only one patient complained of difficulty in swallowing. One patient who experienced urinary frequency and ur-}

**TABLE 1**

**Summary of the ASIA impairment grading scale**

<table>
<thead>
<tr>
<th>ASIA Grade</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>complete: no motor or sensory function preserved in the S4–5 segments</td>
</tr>
<tr>
<td>B</td>
<td>incomplete: sensory but no motor function preserved below the affected level &amp; extending through the S4–5 segments</td>
</tr>
<tr>
<td>C</td>
<td>incomplete: motor function preserved below the affected level, &amp; the majority of key muscles below the affected level are &lt; grade 3/5</td>
</tr>
<tr>
<td>D</td>
<td>incomplete: motor function preserved below the affected level, &amp; the majority of key muscles below the affected level are ≥ grade 3/5</td>
</tr>
<tr>
<td>E</td>
<td>normal: motor &amp; sensory function normal</td>
</tr>
</tbody>
</table>

C-3 and C-7 (Fig. 1 upper left). The most cephalad extent was the inferior aspect of C-2 in two patients. The mean ossified ALL was approximately 1 cm in thickness and caused anterior displacement of the prevertebral soft tissue. Involvement of another spinal region (T8–11) was present in only one patient. On plain radiographs and CT scans small areas of calcification were demonstrated within a defined retroodontoid mass (Fig. 1 lower). There was no osseous erosion of the atlantoaxial complex, and the gross osseous anatomy was relatively normal. The MR imaging studies revealed a soft-tissue retroodontoid mass that was slightly hypointense to neural matter on T1-weighted sequences and areas of hypo- and hyperintensity on T2-weighted sequences (Fig. 1 upper right). The degree of CVJ stenosis was significant (mean diameter 5 mm, range 4–7 mm) (Table 4). Imaging studies obtained in all patients revealed signs of myelomalacia with signal change in the cord on T2-weighted MR images. There was fair but inhomogeneous contrast enhancement after intravenous gadolinium administration. Additionally in one patient there was radiographic evidence of basilar invagination, accentuating the degree of CVJ compression.

**Surgical Procedure**

During surgical decompression and stabilization, all patients underwent intraoperative sensory evoked potential monitoring. A transoral odontoidectomy and resection of the hypertrophic mass were performed in four patients. Three of four patients also underwent a posterior fusion procedure as an immediate second-stage procedure on the same day to stabilize the cervical spine (Fig. 2). One patient (Case 3) with associated basilar invagination required a transoral, transpalatal approach to obtain enhanced superior exposure. One patient (Case 1) underwent a posterior C1–2 fusion following a transoral decompression; this procedure was converted to an occiput–C3 fusion after dislodgment of the graft following a fall 1 month after the initial surgery. One patient (Case 4) in whom a stand-alone transoral decompression was initially performed underwent a delayed C1–2 fusion after developing a C1–2 subluxation with retrolisthesis and neurological deterioration. Another patient (Case 2) with an unusual circumferential compression and predominantly posterior column symptoms underwent a posterior decompression combined with an occiput–C4 fusion (Table 2).
TABLE 2
Summary of pre- and postoperative data in five patients with FD and a retroodontoid mass

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Symptoms &amp; Signs</th>
<th>Preop ASIA Grade</th>
<th>Investigation</th>
<th>Op Procedure</th>
<th>Complication</th>
<th>Pathological Finding</th>
<th>Outcome</th>
<th>Postop ASIA Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>65</td>
<td>M</td>
<td>lt UE weakness: 4-/5 &amp; numbness; bilat LE weakness: 4/5</td>
<td>D</td>
<td>MRE ALL calcification, retroodontoid mass, CVJ comp</td>
<td>transoral mass resection, pos C1–2 fusion; transarticular screw &amp; Songer cable &amp; ICBG</td>
<td>dislodged graft after falling; revision: Oc–C3 fusion</td>
<td>DFIP</td>
<td>improved strength: UE: 5/5; LE: 4+/5</td>
<td>D</td>
</tr>
<tr>
<td>2</td>
<td>75</td>
<td>F</td>
<td>prog weakness lt side: 3-/5; numbness hands &amp; feet; paresthesia lt side for 6 mos</td>
<td>C</td>
<td>MRE ALL calcification, retroodontoid mass, CVJ comp, C3–4 DDD</td>
<td>pos decomp, Oc–C4 fusion; Steinmann pins &amp; Songer cables</td>
<td>none</td>
<td>no specimen</td>
<td>improved strength: UE: 4+/5; LE: 4+/5</td>
<td>D</td>
</tr>
<tr>
<td>3</td>
<td>54</td>
<td>M</td>
<td>neck pain for 5 yrs; UE weakness: 2 to 3-/5; LE weakness: 3-/5 &amp; paresthesia; dysphagia</td>
<td>C</td>
<td>MRE ALL calcification, retroodontoid mass, basilar invagination, CVJ comp; AbN swallowing test</td>
<td>transoral mass resection, Oc–C5 fusion; Steinmann pins &amp; Songer cables</td>
<td>none</td>
<td>DFIP</td>
<td>improved strength: UE: 4+ to 5/5; LE: 5-/5</td>
<td>D</td>
</tr>
<tr>
<td>4</td>
<td>84</td>
<td>M</td>
<td>prog gait instability for 8 mos; quadriaparesis: 2 to 3-/5 LE paresthesia</td>
<td>C</td>
<td>MRE ALL calcification, retroodontoid mass, CVJ comp</td>
<td>transoral mass resection, PEG for postop feeding</td>
<td>retrolisthesis of C-1 on C-2, pos C1–2 fusion (Gallie), pneumonia</td>
<td>DFIP</td>
<td>improved strength: UE &amp; LE: 3+/5; died 3 wks postop</td>
<td>D</td>
</tr>
<tr>
<td>5</td>
<td>86</td>
<td>M</td>
<td>prog quadriaparesis: UE: 3+/5 &amp; LE: 4/5; bilat hand numbness, neck pain</td>
<td>D</td>
<td>MRE ALL calcification, retroodontoid mass, CVJ comp; AbN swallowing test</td>
<td>transoral mass resection, Oc–C4 fusion; Steinmann pins &amp; Songer cables</td>
<td>pneumonia</td>
<td>DFIP</td>
<td>improved strength: UE: 4+ to 5-/5 LE: 4+/5; improved swallowing</td>
<td>D</td>
</tr>
</tbody>
</table>

* AbN = abnormal; comp = compression; decomp = decompression; DFIP = degenerative fibrocartilaginous inflammatory process; ICBG = iliac crest bone graft; LE = lower extremity; MRI = MR imaging; Oc = occiput; PEG = percutaneous endoscopic gastrostomy; pos = posterior; prog = progressive; UE = upper extremity.
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**TABLE 3**

<table>
<thead>
<tr>
<th>Symptoms/Signs</th>
<th>No. of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>gait disturbance</td>
<td>5 (100)</td>
</tr>
<tr>
<td>weakness</td>
<td>5 (100)</td>
</tr>
<tr>
<td>UE</td>
<td>5 (100)</td>
</tr>
<tr>
<td>LE</td>
<td>5 (100)</td>
</tr>
<tr>
<td>paresthesias</td>
<td>5 (100)</td>
</tr>
<tr>
<td>pain</td>
<td>4 (80)</td>
</tr>
<tr>
<td>cervical spine</td>
<td>2 (40)</td>
</tr>
<tr>
<td>UE</td>
<td>2 (40)</td>
</tr>
<tr>
<td>LE</td>
<td>3 (60)</td>
</tr>
<tr>
<td>dysphagia</td>
<td>1 (20)</td>
</tr>
</tbody>
</table>

**TABLE 4**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Anteroposterior Diameter of CVJ (mm)</th>
<th>Levels Involved</th>
<th>Coexisting Spinal Abnormality</th>
<th>Other Areas Involved</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>5 mm</td>
<td>C3–T1</td>
<td>none</td>
<td>T8–L3</td>
</tr>
<tr>
<td>2</td>
<td>7 mm</td>
<td>C4–T2</td>
<td>C3–4 DDD</td>
<td>none</td>
</tr>
<tr>
<td>3</td>
<td>7 mm</td>
<td>C4–7</td>
<td>basilar invagination</td>
<td>none</td>
</tr>
<tr>
<td>4</td>
<td>4 mm</td>
<td>C3–6</td>
<td>none</td>
<td>none</td>
</tr>
<tr>
<td>5</td>
<td>4 mm</td>
<td>C3–6</td>
<td>none</td>
<td>none</td>
</tr>
</tbody>
</table>

**Pathological Examination**

Four patients underwent resection of the retroodontoid mass. Pathological specimens were examined both grossly and microscopically. The gross examination of the specimen showed pink to yellowish soft tissue. There were areas with a tan hemorrhagic component within the fibrous tissue. Microscopic examination showed similar pathological changes in varying degree in the material resected in the four patients. Such material would include the hypertrophic abnormal tissue found on the posterior aspect of the odontoid process, portions of the transverse/cruciate ligament, and portions of the odontoid itself. Portions of normal-appearing fibro- and hyaline cartilage were commonly found with a variable amount of normal adipose tissue and ligamentous material. In cases in which the odontoid processes were examined, unremarkable-appearing bone was demonstrated with modest endosteal fibrosis without hematopoiesis.

Significant pathological changes included fibrillation of fibro- or hyaline cartilage, loss of basophilia, and prominent zones of acellular eosinophilic degeneration (Table 2). Such zones may be very small areas surrounded by normal cellular cartilage but grow by confluence of contiguous areas to reach significant size. Multifocal zones of chondrocyte hyperplasia were observed. Such foci often bordered areas of chondroid fibrillation or acellular eosinophilic degeneration. Elsewhere, focal neovascularization was observed, occasionally accompanied by modest chronic inflammation and fibrosis. These zones were reminiscent of mature granulation tissue. Granular calcification or, rarely, dystrophic ossification was noted in the zones of acellular eosinophilic degeneration. None of the specimens had pyrophosphate dihydrate crystal deposit.

**Procedure-Related Outcome**

Neurological improvement was demonstrated in all patients immediately postoperatively, with an increase of approximately one ASIA grade in motor strength demonstrated in three patients within 2 weeks. Complications included aspiration pneumonia in two patients and graft dislodgment in the patient who underwent a C1–2 fusion. In the patient who did not undergo an immediate posterior or stabilization procedure a C1–2 subluxation developed 1 week after surgery. A posterior C1–2 fusion was performed, but the patient eventually died of pulmonary complications after experiencing initial neurological improvement. His lower-extremity strength improved from 2/5 to 3/5 within 1 week postoperatively. All patients continued to improve neurologically as of final follow up 4 to 19 months later, and there was no further progression of the neurological deficit (Table 2).

**Discussion**

**Forestier Disease**

Forestier disease or DISH is a relatively common entity as indicated by the 6 to 12% prevalence rate reported in general autopsy series. The most widely accepted radiological criteria for FD was proposed by Resnick and Niwayama in 1976. They include flowing calcification along anterolateral aspect of at least four contiguous vertebral bodies, relative preservation of intervertebral disc height in affected areas, and absence of apophyseal joint ankylosis and sacroiliac joint sclerosis. It typically affects men in their mid-sixth to seventh decade of life and most frequently affects the lower thoracic spine, causing mid- to low-back pain and stiffness. When the cervical spine is involved, dysphagia is the most common presenting complaint. Nearly 30% of patients describe swallowing difficulty and 10% require surgical intervention. It is unusual to have neurological symptoms associated with FD. In 1995 Goto, et al., described a case of FD that caused cervical myelopathy and quadriplegia. In this patient a pseudarthrosis developed between the posterior tubercle of the atlas and the spinous process of the axis and caused extradural compression.

In the group of patients in the present report FD-related progressive neurological deficits developed due to the development of a nonheumatoid retroodontoid pannus.

**Retroodontoid Mass**

There are various conditions associated with retroodontoid degenerative soft-tissue pannus formation, but FD has not been previously included. The most commonly described cause of retroodontoid pannus formation is RA. Occasionally os odontoideum and, less frequently, pseudogout (periodontoid calcium pyrophosphate dihydrate deposition disease), nonunion of odontoid fractures, retroodontoid disc herniation, and synovial cysts have been described in association with pannus formation as well. These conditions are clearly different pathological entities that can certainly coexist in patients with FD; however, in our series of patients there was no evidence of other coexisting disease. Histopathological examination in our series showed noneoplastic, degenerative, and
inflammatory tissue. Although the process bears similarity to that in RA, there was no evidence of RA in any of our patients.

**Pathophysiology of Mass Formation**

The common finding of multilevel fusion of the subaxial cervical spine was demonstrated in all five cases. This fusion coupled with the mobility of the craniovertebral joint complex seems to play a pivotal role in the gradual development of a retroodontoid mass. The occipitoatlantoaxial joint accounts for 45° of flexion and extension and 50% of the rotation in the normal cervical spine, whereas each subaxial level in the cervical spine contributes between 10 and 20° of flexion and extension. In FD, ALL calcification–induced multilevel subaxial fusion concentrates the mechanical stresses at the atlantoaxial junction. Chronic strain on the supporting ligaments and the surrounding soft tissues, particularly involving the occipitoatlantoaxial complex, results in repeated and continual microtears of the periodontoid ligaments. These stresses, combined with the scarring associated with the ensuing healing process, cause gradual accumulation of degenerative and inflammatory material, which is accompanied by compensatory ligamentous hypertrophy. The

**Fig. 1.** *Upper Left:* Preoperative radiograph revealing marked calcification of the ALL. *Upper Right:* Preoperative MR image revealing marked cervicomedullary compression. *Lower:* Preoperative axial CT scan demonstrating a retroodontoid soft-tissue mass (arrows).
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theory of ligamentous hypertrophy secondary to increased mechanical stress is not a unique concept and has been well documented to occur in the ligamentum flavum of the lumbar region.\(^{8,15}\)

The histopathological findings are likely explained by a series of events beginning with fibrous matrix and eosinophilic degeneration. Subsequently, focal dystrophic calcification and inflammatory neovascularization occur as a result of chronic focal stress. This process is analogous to granulation tissue formation that occurs during scarring.

Perioperative Management

Surgical management is often the only viable option to halt the progressive neurological deficit that develops in these frequently frail and debilitated patients. Grob, et al.,\(^{13}\) have shown that posterior fusion alone allows gradual regression of anterior cervicomedullary pannus size in patients with RA. Similarly, a stand-alone posterior fusion in FD patients would most likely yield the same results, leading to pannus involution over a period of several months. This would be feasible in patients with mild or static neurological symptoms, avoiding the potential complications associated with a transoral approach. In patients with severe and rapidly progressive motor symptoms due to spinal cord compression, a transoral approach may be used to achieve an immediate decompression. In four of our five patients severe anterior cervicomedullary compression and MR imaging signal changes were demonstrated, with rapidly progressive myelopathy and quadriparesis. A transoral approach was indicated in these patients to address the severity of the anterior compression directly. Although transoral resection of the mass may directly address the primary problem, the resulting instability requires either a C1–2 fusion or an occipitocervical fusion.\(^{7}\) A C1–2 fusion is most likely inadequate because of the potential for failed fusion, particularly in light of the severely altered biomechanics related to the underlying FD. A more definitive occipitocervical fusion appears to be the optimal procedure (A Crockard: Personal communication, April 1997).\(^{12,22}\)

The disadvantages of a longer operative time and marked reduction of neck motion is often justified to prevent a serious neurological complica-

Conclusions

Forestier disease most commonly presents in the elderly population, causing dysphagia, and FD-associated neurological deficits are rare. The development of a soft-tissue retroodontoid hypertrophic pannus in association with progressive neurological deficit is caused by altered biomechanics at the atlantoaxial junction. The typical degenerative hypertrophy of the soft tissues in the retroodontoid region in FD patients is distinctly different from other well-known causes. There should be a heightened suspicion for CVJ lesions of this nature in patients with FD who present with cervical myelopathy.

References


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