

Case Report

Gun Seok Oh, M.D.
 Chang Il Ju, M.D.
 Seok Won Kim, M.D.
 Seung Myung Lee, M.D.
 Ho Shin, M.D.

Department of Neurosurgery
 College of Medicine, Chosun
 University, Gwangju, Korea

Complete Cord Injury after Minimal Trauma in a Patient with Forestier's Disease Accompanying Ossification of the Posterior Longitudinal Ligament

Forestier's disease is a systemic rheumatological abnormality in which exuberant ossification occurs along ligaments throughout the body, but most notably the anterior longitudinal ligament of the spine. This disease is usually asymptomatic; however dysphagia, dyspnea, and peripheral nerve entrapment have all been documented in association with the disorder. We report a rare case of catastrophic neurologic damage caused by Forestier's disease accompanying ossification of the posterior longitudinal ligament.

KEY WORDS : Forestier's disease · Ossification of the posterior longitudinal ligament.

INTRODUCTION

Forestier's disease is a noninflammatory proliferative enthesopathy that predominantly involves the spine, resulting ultimately in the establishment of characteristic bridging anterolateral osteophytes that ankylose adjacent vertebral bodies. The entity was first described in 1950 by Forestier and Rotes-Querol as senile ankylosing hyperostosis of the spine²⁾. Predominant among the pathological entities that can be confused with Forestier's disease are osteophytes accompanying degenerative disease of the cervical spine, and ankylosing spondylitis⁴⁾. A case of Forestier's disease accompanying ossification of posterior longitudinal ligament (OPLL) presenting as complete cord injury is described, which serves to emphasize that significant morbidity, although unusual, may be included in the clinical manifestations of this idiopathic condition.

CASE REPORT

A 65-year-old man weighing 92 kg, fell down during a syncopal episode in his bathroom. He had a 10 year history of diabetes mellitus and 3 year history of dysphagia and neck motion limitation. He was in quadriparetic state (grade II in upper extremities, grade 0 in lower extremities). Simple lateral radiograph and computed tomography scan revealed ossification of anterior longitudinal ligament at C2-C6 level and OPLL at C4-C7 level (Fig. 1). Magnetic resonance images revealed severe cord compression caused by OPLL and spinal stenosis (Fig. 2). Serologic tests for syphilis, rheumatoid factor, anti-nuclear antibody and serum HLA-B27 were negative and there was no sacroiliac joint ankylosis. Unfor-

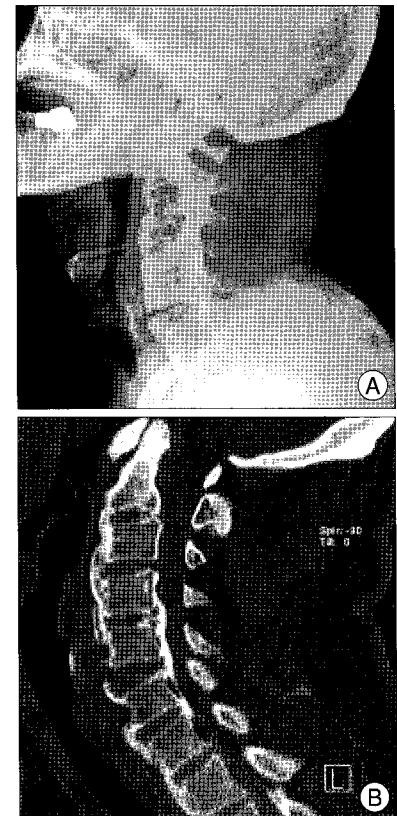


Fig. 1. A, B : Simple lateral radiograph and computed tomography show extensive ossification along the anterior and posterior vertebral bodies and widening disc space of C5-C6 level.

• Received : September 5, 2007
 • Accepted : November 12, 2007
 • Address for reprints :
 Seok Won Kim, M.D.
 Department of Neurosurgery
 College of Medicine, Chosun
 University, 588 Seoseok-dong
 Dong-gu, Gwangju 501-717, Korea
 Tel : +82-62-220-3120, 3126
 Fax : +82-62-227-4575
 E-mail : chosunns@hanmail.net



Fig. 2. T2-weighted sagittal magnetic resonance image reveals narrow spinal canal and high signal intensity at C4 level.

age groups. In the 1970s, Resnick et al.^{4,6} coined the term diffuse idiopathic skeletal hyperostosis (DISH) for Forestier's disease. They were the first to direct attention to the extraspinal ossification seen in this disease, pointed out the systemic nature of the process, and established specific radiological criteria for the diagnosis of Forestier's disease that are still used today^{4,6,10}. These criteria are as follows : 1) flowing calcified ossification along anterolateral aspect of four contiguous vertebral bodies; 2) relative preservation of intervertebral disc height in affected areas; and 3) absence of apophyseal joint ankylosis and sacroiliac joint sclerosis. Resnick et al.⁵ established these criteria to separate Forestier's disease from similar disease processes.

The presence of ossification bridging on the anterolateral aspect of four contiguous vertebral bodies and the preservation of disc height in affected areas distinguish Forestier's disease from large anterior cervical osteophytes in the setting of degenerative disc disease. The absence of apophyseal joint ankylosis and sacroiliac joint fusion segregates Forestier's disease from ankylosing spondylitis. Forestier's disease is weakly associated with HLA B-27, suggesting that it is a distinct pathophysiological form of ankylosing spondylitis¹. Forestier's disease is not an uncommon disorder among rheumatological populations and has been reported in 12% of random autopsy series in a Veterans administration hospital population⁵. However, due to the bone projection away from the spinal cord, it is rare for a patient to have symptoms that would be elicited by a neurosurgeon. Although these patients are typically asymptomatic, there is a documentation of Forestier's disease patients presenting with spinal instability, upper gastrointestinal, respiratory, and neurological problems⁷⁻⁹.

tunately, there was aspiration of food material and his condition gradually deteriorated. He died of bronchopneumonia while hospitalized awaiting surgery.

DISCUSSION

Forestier's disease affects men more frequently than women (2:1). They are predominantly middle to older

The ossification is not always localized to the anterior longitudinal ligament, but is sometimes more extensive. It is frequently associated with OPLL and ossification of the ligamentum flavum. Most people are asymptomatic, which explains the relative paucity of data in the neurosurgical literature. Mild, painless joint or back stiffness is frequently the only complaint. This symptom is often discounted in an elderly patient population, because these patients often have other rheumatic disorders or degenerative disease of the spine. Neurological complaints are quite rare, and the most common symptoms produced by ossification of the anterior longitudinal ligament pertain to compression of the trachea and esophagus. McAfee et al.³ and Resnick et al.⁵ suggested that there was a close association between Forestier's disease and OPLL. Although conventional radiograph clearly confirms the diagnosis of DISH, computed tomography (CT) and magnetic resonance images (MRI) are more capable of detecting associated findings (eg, OPLL) and complications (eg, spinal cord compressive myelomalacia). OPLL is readily demonstrated by lateral radiograph, but because of the presence of overlying osseous structures, CT is occasionally indicated for the purpose of confirmation. Furthermore, MRI can demonstrate a narrowing of the spinal cord even though the ossification when OPLL contains fatty bone marrow, and it can detect the ligament hypertrophy that is an early change in OPLL.

CONCLUSION

Forestier's disease is a rheumatological disorder distinctly different from ankylosing spondylitis. The majority of patients are asymptomatic but it can cause severe myelopathy especially if there is a narrow spinal canal and OPLL. Awareness that OPLL in Forestier's disease can cause neurologic complications may lead to a more thorough radiologic evaluation and prevent permanent neurologic sequelae.

References

1. Ercilla MG, Brancos MA, Breyse Y, Alonso G, Vives J, Castillo R, et al : HLA antigens in Forestier's disease, ankylosing spondylitis, and polyarthrosis of the hands. *J Rheumatol (Suppl)* 3 : 89-93, 1977
2. Forestier J, Rotes-Querol J : Senile ankylosing hyperostosis of the spine. *Ann Rheum Dis* 9 : 321-330, 1950
3. McAfee PC, Regan JJ, Bohlman HH : Cervical cord compression from ossification of the posterior longitudinal ligament in non-orientals. *J Bone Joint Surg Br* 69 : 569-575, 1987
4. Resnick D, Niwayama G : Radiographic and pathologic features of spinal involvement in diffuse idiopathic skeletal hyperostosis (DISH). *Radiology* 119 : 559-568, 1976
5. Resnick D, Niwayama G : Diffuse idiopathic skeletal hyperostosis (DISH) : ankylosing hyperostosis of Forestier and Rotes-Querol, in Resnick D, Niwayama G (eds) : *diagnosis of Bone and Joint Disorders with Emphasis on Articular Abnormalities*. Philadelphia : WB Saunders, 1988, pp 1562-1602
6. Resnick D, Guerra J Jr, Robinson CA, Vint VC : Association of diffuse

- idiopathic skeletal hyperostosis (DISH) and calcification and ossification of the posterior longitudinal ligament. *AJR Am J Roentgenol* 131 : 1049-1053, 1978
7. Stechison MT, Tator CH : Cervical myelopathy in diffuse idiopathic skeletal hyperostosis. Case report. *J Neurosurg* 73 : 279-282, 1990
 8. Underberg-Davis S, Levine MS : Giant thoracic osteophyte causing esophageal food impaction. *AJR Am J Roentgenol* 15 : 319-320, 1991
 9. Warnick C, Sherman MS, Lesser RW : Aspiration pneumonia due to diffuse cervical hyperostosis. *Chest* 98 : 763-764, 1990
 10. Youn BM, Koo YD, Suk JS, Choi DY : Forestier's Disease (Ankylosing Hyperostosis of the Spine). Case Report. *J Korean Neurosurg Soc* 9 : 647-651, 1980