Spinal Cord Compression by Primary Amyloidoma of the Spine

Kemal Nas1, Adem Arslan2, Adnan Ceviz2, Aslan Bilici2, Ali Gür3, Mustafa Serdar Kemaloğlu3, Remzi Çevik3, and Aysegül Mali Saraç1

Department of 1Physical Therapy and Rehabilitation, 2Pathology, 3Neurosurgery, 4Radiology, Faculty of Medicine, University of Dicle, Diyarbakır, Turkey.

In this report, we presented a case of solitary spine amyloidoma, its clinical and radiological findings and management, and a review of the literature on vertebral amyloidosis.

**Key Words:** Amyloid, spine, magnetic resonance imaging, rehabilitation

**INTRODUCTION**

Amyloidosis is a systemic disease, which can affect single or multiple organs. Primary solitary amyloidosis, or amyloidoma, is a rare subset of amyloidosis in which amyloid deposition is focal and not secondary to a systemic process or plasma cell dyscrasias.

Cases of skeletal involvement in various forms of amyloidosis have been reported but these have been linked with multiple myeloma or other plasma cell dyscrasias. Primary focal localized amyloidoma of bone is extremely unusual; only fourteen cases of spinal involvement have been reported (Table 1).

**CASE REPORT**

A 60-year-old man presented with a back pain of one year duration that radiated into both legs. Three months before admission to our depart-

ment, sensory disturbances and weakness developed in both legs, and progressed resulting in moderate paraparesis.

Hypoesthesia was present below the level of the sensory segment of thoracic 9 (T9), a marked tenderness to percussion was observed over many vertebrae, and hyperreflexic knee and ankle jerks were present on admission. The erythrocyte sedimentation rate, complete blood cell count, and renal function test results were within normal limits. Levels of serum bilirubin, transaminases, and lactate dehydrogenase were also normal. Serum calcium was 9.7g/dl, total serum protein 8.5g/dl and globulin 3.8g/dl. IgG, IgM, and IgA levels were 1148 (normal values: 800-1700), 139.8 (N: 60-320) and 137.9 (N: 100-490)mg/dl, respectively.

The lower limb motor score was obtained by summing the grades of the myotomes of both lower extremities and ranged from 0 (complete paralysis) to 50 (normal power). The degree of patient impairment was graded according to the ASIA (American Spinal Injury Association) impairment scale (modified from Frankel). The mean ASIA score was 1.4 ± 0.54 preoperatively, and 3.2 ± 0.44 one week, 8.2 ± 0.83 one month, and 9.2 ± 1.09 three months after surgery.

Assessment of functional outcome was based on the Modified Bartel Index (MBI). This is an ordinal scale with total scores ranging from 0 (totally dependent) to 100 (completely independent). The mean MBI score was 4.0 ± 3.94 preoperatively and 9.5 ± 3.68 three months after the operation.

Magnetic Resonance Imaging (MRI) of the thoracic spine revealed a soft tissue mass arising...
from the marrow of the T9 vertebra. The mass had heterogeneous intermediate signal intensity on T1-weighted images and low signal intensity on T2-weighted images. After intravenous contrast medium administration, the mass showed heterogeneous minimal enhancement. The mass was found to have protruded to the left paravertebral space and showed extension to the left transverse process and the spinal canal. Loss of vertebral body height, especially anteriorly, was detected resulting in kyphotic angulation at the T9 level. Severe epidural cord compression was observed at this level by the mass. In the posterior part of the spinal canal at the T8-9 level, laminectomy defect, cystic mass and operative material were also present, remnants of previous surgery (Fig. 1). It was understood that only laminectomy had been performed during his first operation at another hospital and the pathological diagnosis was inadequate. A technetium-99m-pyrophosphate (99m Tc-PP) bone scan showed increased uptake in the same vertebra and pulmonary functions were found to be sufficient for operation.

During the operation laminectomy was performed posterolaterally from the left side at the T9 and T10 levels. The mass was resected totally and decompression of the spinal cord was achieved by laminectomy. Resected specimens consisted of bone fragments and reddish-pink soft tissue. Anterior intervention was not planned since the mass continued from the left lateral to the posterior of the spinal cord.

Histological examination was performed upon specimens. Hemotoxylin-eosin staining showed homogeneous pink stained masses (Fig. 2). These masses were stained pink to orange-red by Congo red, and a yellow-green birefringence under the polarizing microscope (Fig. 3). Foreign-body giant cell, lymphocyte and plasma cell infiltration were observed in the peripheries of these masses (Fig. 2), in addition some sections showed calcification. These findings are consistent with amyloidosis.

In the early postoperative period, active-assisted, and active and resistance exercises were applied. The sequencing of strengthening began with isometrics, initially with the head supported in a neutral position, positions were then varied relative to gravity and gradually progressed from neutral positioning. This was followed by isotonic exercises, with an emphasis upon the concentric strengthening of the lengthened and weakened muscles. The patient was mobilized by using a walker apparatus from the postoperative sixth day.

Six months after the operation, the patient was neurologically intact and could manage his daily activities unaided.

**DISCUSSION**

Amyloidosis is an uncommon disease, in which
various organs are infiltrated by amorphous extracellular eosinophilic material, composed of insoluble proteins, which has a high degree of beta-pleated sheet protein structure.\textsuperscript{6} Amyloid deposits may be focal or systemic and related (reactive/secondary amyloidosis) or unrelated (primary amyloidosis) to an underlying infectious or inflammatory process.\textsuperscript{2} In reactive/secondary amyloidosis, treatment of the underlying disease can commonly slow or actually reverse disease progression with 5- and 10-year survivals after diagnosis.\textsuperscript{9} Previous reports have described amyloid bone tumors in the pelvis, sacrum, skull base, temporal bone, tibia, and femur.\textsuperscript{10-13} Tumor-forming masses of amyloid in the spine have been reported in fourteen cases, ten occurred in patients with thoracic lesions\textsuperscript{14-21} and the others in patients with cervical lesions.\textsuperscript{2,24-26}

A complete evaluation of the diagnosis of systemic amyloidosis is essential for deciding upon management and estimating prognosis. Primary solitary amyloidosis is a rare subtype of
amylodosis, which has an excellent prognosis with local resection. Amyloidosis typically occurs in older patients with a mean age at presentation of 65 years. Moreover, amyloid deposits can be found throughout the body in single or multiple organs. Single soft-tissue or bone lesions have been referred to as the tumoral form of amyloid or amyloidomas. Amyloidomas of the bone are slow growing lesions, which result in progressive osteolysis.

Primary amyloid lesions of the spine are uncommon findings in cases of vertebral body fractures and collapse, osteopenia, and lytic lesions have been described. Differential diagnostic considerations include metastatic disease, plasmacytoma, primary bone tumors, or chronic inflammatory processes. The clinical presentation of amyloidoma in bone varies. Some patients experience pain, while others have pathologic fractures. All reported cases of amyloidoma involving a vertebral body became symptomatic with neurologic deficits accompanied by pain. Two-thirds of patients with thoracic lesions had paraparesis, and the remainder reported radiculopathies.

Early rehabilitation after decompressive surgery is important for patients with neurological deficits stemming from benign chronic spinal mass. In our patient, a rehabilitation program was started immediately after surgery. The main aim of this program was to maximize lower extremity muscular flexibility. Because of their attachments to pelvis, the hip flexors and extensors have a great influence on the positioning of the lumbar spine. Motor ability was graded using the ASIA scale and functional assessment based on the MBI score was gradually improved using the rehabilitation program in our patient. Thus we obtained a successful result using our exercise program.

In conclusion, localized amyloidosis without evidence of any systemic disease is a benign lesion. It may be treated surgically, and rarely recurs.

REFERENCES