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Case Report

Primary cervical amyloidoma: a case report and review of the literature

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Abstract

BACKGROUND CONTEXT: Primary solitary amyloidosis or amyloidoma is a disease process characterized by the focal deposition of amyloid in the absence of a plasma cell dyscrasia with normal serum protein measurements. Solitary amyloidomas affecting the vertebrae are very uncommon but typically affect the thoracic spine. Primary cervical amyloidosis is an exceedingly rare entity with exceptionally good prognosis, but requires diligence of the treating physician to establish the diagnosis and implement the appropriate surgical intervention.

PURPOSE: This study aimed to present a rare case of primary cervical amyloidosis with long-term follow-up and review the clinical presentation, characteristic imaging findings, diagnostic pathology, differential diagnosis, treatment algorithm, and prognosis of the disease entity. This case demonstrates the progressive resorption of the amyloidoma over time after surgical stabilization. Previous reported cases of primary cervical amyloidosis will also be reviewed.

STUDY DESIGN: This study is a report and review of the literature.

METHODS: A 77-year-old woman presented with a several-week history of gradual progressive weakness in her upper and lower extremities. Computed tomography and magnetic resonance imaging demonstrated a retro-odontoid nonenhancing soft-tissue mass, with erosive bony changes and severe mass effect on the upper cervical cord. The patient was taken to the operating room for decompression and posterior spinal stabilization.

RESULTS: Intraoperative tissue specimens demonstrated amyloidosis and extensive systemic workup did not reveal any inflammatory processes, systemic amyloidosis, or plasma cell dyscrasia. Postoperatively, the patient regained full strength and ambulatory status. The patient remains asymptomatic at a 2-year follow-up. A postoperative follow-up magnetic resonance imaging demonstrated complete resorption of the residual amyloidoma.

CONCLUSIONS: Primary solitary amyloidosis is a rare form of amyloidosis that is important to differentiate given its excellent prognosis with surgical management. Treatment should include surgical decompression and spinal stabilization. This is the first case report to clinically and radiographically demonstrate the progressive resorption of a primary amyloidoma over time after surgical stabilization in the upper cervical spine. It is imperative that surgeons encountering such lesions maintain a high suspicion for this rare disease entity and advise their pathologists accordingly to establish the correct diagnosis.

Keywords: Cervical spine; Amyloidoma; Primary solitary amyloidosis; Resorption; Cervical instrumentation

Introduction

Primary solitary amyloidosis or amyloidoma is a disease process characterized by the focal deposition of amyloid in the absence of a plasma cell dyscrasia with normal serum protein measurements [1,2]. Amyloidomas can arise in the bone, skin, larynx, lymph nodes, bladder, eye, tongue,

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and gastrointestinal system [3]. Solitary amyloidomas affecting the vertebrae are exceedingly rare [1–22]. To date, only seven cases of primary solitary amyloidoma affecting the cervical spine have been reported [1,3,7,8,14,15,17].

We report a case of solitary cervical amyloidoma managed with posterior surgical decompression and stabilization with 2-year follow-up as well as review the literature on primary spinal amyloidosis. Clinicians should maintain a high degree of suspicion for this disease process when encountering spinal masses because the diagnosis requires special stains as well as specific laboratory tests to diagnose the disease and rule out systemic involvement.

Case report

A 77-year-old female with a history of osteoarthritis, gout, and hypertension presented to the emergency department with a chief complaint of worsening syncope over several weeks. Her granddaughter and husband also noted that she developed worsening altered mental status in the same period. Further questioning revealed that she had been gradually developing weakness of her bilateral upper and lower extremities over the course of several months, with acute worsening in the weeks before presentation. In the past week, she had been unable to use her walker because of hand weakness and dysfunction. Additionally, she endorsed a 50-pound weight loss in the 2 years before presentation. She denied any additional constitutional symptoms including fevers, chills, nausea, or vomiting. She denied significant neck pain. She had not undergone any spine procedures previously.

Her initial clinical examination on presentation to the emergency department was remarkable for 3−/5 strength in her left upper and lower extremities, and 3+/5 strength in her right upper and lower extremities. She did not have sensory deficits and had intact rectal tone. She was markedly myelopathic, with positive Hoffmans' signs bilaterally, 3+reflexes in her upper and lower extremities, and sustained clonus bilaterally. She was afebrile, with the remainder of her vital signs within normal limits. Her white blood cell count was 7.10 (4–11 k/uL), hematocrit was 40.3% (35–47%), and platelets were 211 (150–450 k/uL). Her erythrocyte sedimentation rate was 10 mm/h [0–20 mm/h] and C-reactive protein was 0.2 mg/dL (<0.8 mg/dL). Basic metabolic panel and coagulation profile were each within normal limits. Serum protein electrophoresis was 7.3 (5.8–8.1) and urine protein electrophoresis was <68 (<100) and all albumin.

Given the patient's initial presentation with syncope, a brain magnetic resonance imaging (MRI) was obtained, which did not demonstrate any intracranial pathology, but incidentally noted an upper cervical spine lesion. Initial cervical spine radiographs demonstrated erosive bony changes of the C2 vertebral body, which prompted advanced imaging. Computed tomography and MRI of the cervical spine (Figs. 1 and 2) revealed a nonenhancing soft-tissue mass centered along the posterior aspect of C2, eccentric to the left, associated with prominent erosive bony changes and severe resultant mass effect on the proximal cervical cord with associated focal cord signal change. Subaxial spinal stenosis was recognized as well. These findings were felt by musculoskeletal radiology to be consistent with a nonvascular pannus formation, or, less likely, calcium pyrophosphate deposition.

The decision was made to proceed with operative intervention for decompression of her cervical spinal cord and to obtain tissue for diagnostic pathologic examination.

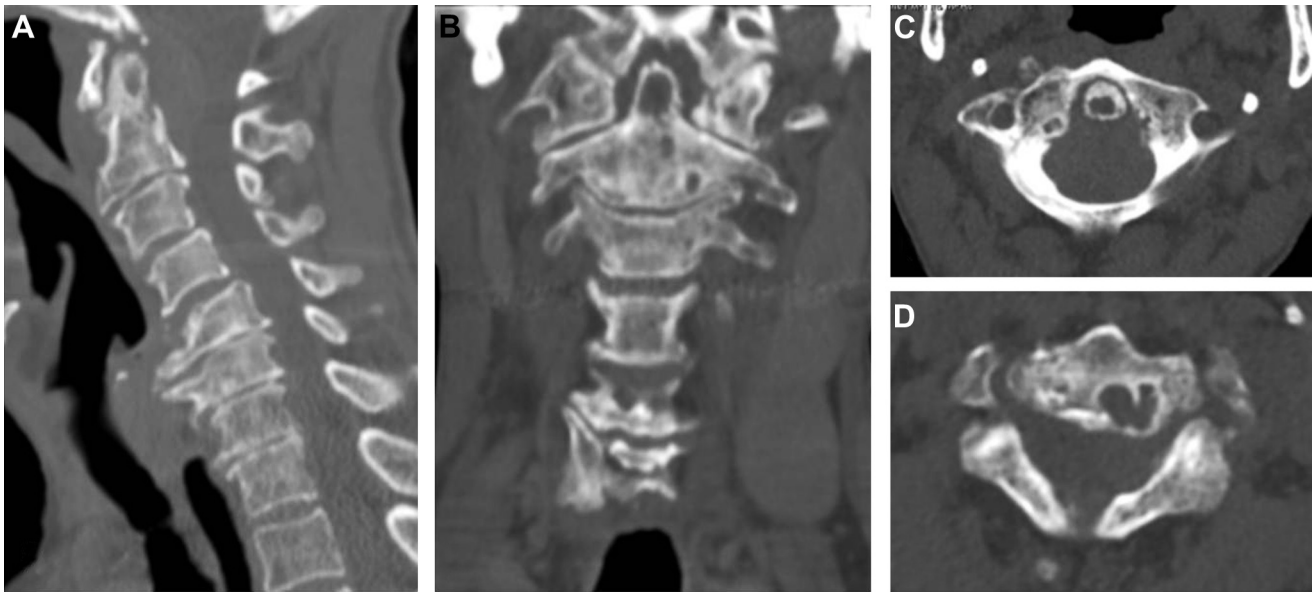

Fig. 1. Initial sagittal (A) and coronal (B) computed tomography scan obtained at presentation demonstrated a nonenhancing soft-tissue mass centered along the posterior aspect of C2, eccentric to the left, associated with prominent erosive bony changes of both the dens (C) and the body of C2 (D).