Clinical Neurology and Neurosurgery 137 (2015) 1–4

Contents lists available at ScienceDirect

Clinical Neurology and Neurosurgery

journal homepage: www.elsevier.com/locate/clineuro

Case Report

Differentiated plasma cell myeloma presenting as a solitary spinal amyloidoma: A case report, possible pitfall and review to the literature

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ARTICLE INFO

Article history:
Received 17 February 2015
Received in revised form 21 May 2015
Accepted 25 May 2015
Available online 10 June 2015

Keywords:
Amyloidoma
Paraparesis
Plasmocytoma
Spinal cord compression

ABSTRACT

Objective: Solitary spinal amyloidoma is a rare entity. Amyloidomas consist of extracellular amyloid deposits with an insoluble beta-pleated proteinaceous material. Although amyloidomas are slow growing lesions, they may lead to a progressive spinal cord or nerve root compression. Moreover amyloidoma results in destruction of bone with consequence of progressive osteolysis.
Method: This study is a case presentation and review of the literature and should point out the need to explore any underlying diseases to guarantee the best therapy for the affected patient. In this case report we present a female patient with high-level paraparesis and lumbar stenosis in L2–L3 with combined spondylolisthesis (ASIA Impairemet Scale C). Paraparesis increased shortly after lumbar osteosynthesis. Contrast-enhanced MRI of the thoracic spine revealed medullary compression at the D5 level due to an epidural and paraspinal mass with concomitant bone infiltration. Operative decompression followed. Histopathological examination initially revealed amyloidoma. Finally the lesion was classified as a plasma cell myeloma.
Results: Plasma cell myeloma may rarely present as a solitary amyloidoma in the initial pathological examination with the potential to cause spinal cord compression associated to osteolytic lesions of the spine.
Conclusion: A thorough pathological work-up is mandatory in order to rule out differential diagnosis and exclude possible underlying diseases.

1. Introduction

Spinal amyloidoma is an extremely rare tumor-like lesion. It is a rare subtype of amyloidosis. Pathogenesis and etiology is unclear. Affected are older patients with a mean age of 65 years or older [1]. Amyloid was first named by Virchow in 1854. It is a fibrous protein in various tissues and organs. Normally it occurs diffuse, but when forming a solitary mass, it is called amyloidoma. It is reported in every anatomic site [2]. There can be an infiltration of single organs (like bone, skin, larynx, lymph nodes, bladder, eye, tongue and gastrointestinal system) or can occur in multiple organs, but a systemic deposition is more common. There have been descriptions of amyloid bone tumors in pelvis, sacrum, skull base, temporal bone and femur. Amyloidoma also occurs as side effect in multiple myeloma, plasmocytoma, during long-term-dialysis, lymphoma (plasmocytoid), chronic infection or inflammatory diseases. It is characterized by a focal deposition of amyloid without plasma cell dyscrasia and normal serum protein measurements [3]. Amyloidomas are characterized as a solitary nodulary mass by an extracellular deposit of amyloid an insoluble proteinaceous material with a beta-pleated sheet configuration. Differential diagnosis of solitary amyloidoma consist of metastatic processes, plasmocytoma, primary bone tumors and inflammation, so that possible underlying diseases have to be ruled out before the diagnosis of solitary amyloidoma is made.

Amyloid is an eosinophilic amorphous hyaline extracellular substance. Under the microscope Amyloid stained with Congo Red appears yellow green under polarized light, this is a unique feature differentiating it from other hyaline deposits such as collagen. Amyloidomas of the spine may present on MRI as an osteolytic lesion similar to primary or metastatic tumors and infectious or inflammatory processes [3,4].

The prognosis is related to coexisting conditions, the type of amyloidosis and possible underlying diseases. While immunocytic

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http://dx.doi.org/10.1016/j.clineuro.2015.05.027
0303-8467/

amyloidosis has the worst prognosis with a mean survival of 1 year [5], patients with reactive secondary amyloidosis-subtype show a mean survival of over 5 years.

We report a case of differentiated plasma cell myeloma mimicking a solitary thoracic spine amyloidoma in a 66-year-old female patient.

2. Case report

In October 2012 a 66-year-old female patient presented with a sudden high-level paraparesis without any sensory deficits (ASIA Impairment Scale C). Furthermore the patient reported lumbar pain with bilateral irradiation into the front thigh. Due to lumbar stenosis L2–L3 and combined spondylolisthesis, lumbar fusion L2–L5 was conducted (TLIF-PEEK-Cages). Postoperative pain decreased. Paraparesis increased shortly after the initial operation. Therefore a subsequent contrast enhanced MRI of the thoracic spine (Fig. 1a–d) was performed. It showed an extradural mass lesion compressing the spinal cord at the Th5-level and infiltrating the right pedicle, transverse process and costo-transversal joint showing a strong and homogeneous contrast enhancement. The cranio-caudal extension of the lesion reached from TH4 to the TH6 level with destruction of the posterior surface of Th5. The spinal cord was highly compressed and displaced ventrolaterally to the left side. At the level of the mass-lesion a hyperintense signal within the spinal cord was found on T2-weighted images (myelopathy). Furthermore the right radices of the Th4 and the Th5 nerve root were compressed within the neuronal foramina. The radiological diagnosis comprised metastasis and multiple myeloma. Microsurgical decompression and tumor resection was performed.

Histological examination revealed plasma cell accumulation and amyloid deposits (Fig. 1e). The specimen showed a biphasic pattern of highly cellular areas (HE right) and areas free of amorphous cells (HE left) (Fig. 1e). Highly cellular areas consisted of larger cells with dense, spotted chromatin. These cells showed very low proliferation activity (Ki67) and CD138-positivity indicating plasma cells (CD138). The amorphous areas exhibited birefringence and a color shift toward apple-green under polarized light indicating amyloid deposits (Congo Red), which were partially positive for lambda light chain (lambda light chain), an amyloid

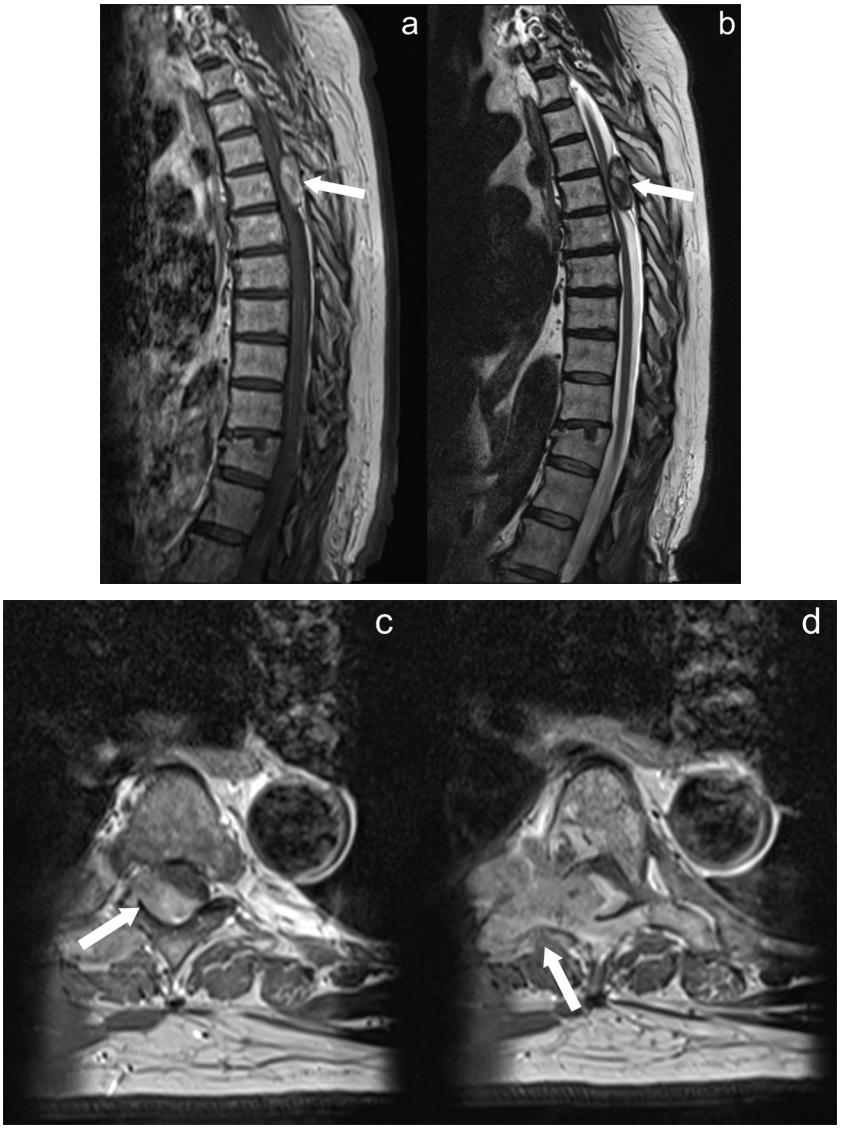

Fig. 1. (a and b) (sagittal, a: T1 + KM, b:T2): MRI showed a tumor mass with KM enhancement with expansion from middle of TH4 to cover plate of TH6 and myelopathic signal TH5. (c and d) (transversal, T1 + KM): Compression of the myelon by epidural mass, expansion paraspinal right and narrowing of neuronal foramina (transversal T1 + KM) (e) Histology showing plasma cell accumulation and amyloid deposits. The specimen showed a biphasic pattern of cell dense areas (HE right) and areas of amorphous cell free areas (HE left).