Amyloidoma of the spine with pathologic fracture of the odontoid process
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Section: Musculoskeletal system
Area of Interest: Haematologic Musculoskeletal spine
Imaging Technique: Conventional radiography
Imaging Technique: CT
Imaging Technique: MR
Special Focus: Metabolic disorders Case Type: Clinical Cases
Authors: Vega Chaves, Andrés; Martínez Calvo, Alberto
Patient: 64 years, male

Clinical History:
A 64-year-old male with personal antecedents of light chain multiple myeloma and AL amyloidosis with good response to Daratumumab. The patient comes to haematologic consult for revision and refers neck pain of 5 days of evolution. He had no previous trauma or neurologic symptoms.

Imaging Findings:
Cervical X-ray shows a pathologic fracture of the odontoid process. Cervical CT confirms the fracture and identifies the lytic lesion that affects the anterior arc of C1, the lateral mass of C1 and C2 and the odontoid process. Also a high-attenuation soft tissue component is visible that extends to the epidural space and especially to the prevertebral soft tissues. MRI shows a lobulated mass that affects anterior arc and lateral masses of C1, odontoid process, body and lateral masses of C2 with an important component of prevertebral soft tissue. Remarkably, it is very hypointense on T2 and STIR sequences and shows weak peripherical enhancement after the administration of Gd.

Discussion:
In an elderly patient, any pathologic fracture that affects the spine should raise suspicion for metastatic involvement, multiple myeloma or plasmacytoma. However, in this patient, affected for multiple myeloma and light-chain amyloidosis, the presence of an amyloidoma should be considered. Amyloidosis is an unusual pathology (incidence of 6-10 cases/100,000 population) caused for abnormal deposition of insoluble proteins in different parts of the organism [1]. The most uncommon manifestation is the presence of pseudotumoral lesions with aggressive growing called amyloidomas [2]. They are more frequent in patients with systemic amyloidosis and multiple myeloma. Although we refer primarily to solitary amyloidomas of the spine [3]. Amyloidomas of the spine produces symptoms depending on the location, size and compression of neurologic structures. Imaging techniques (CT and MRI) are useful for evaluation of the compression, for detecting complications such as fracture and planning surgery, either
for fixation or biopsy, which is required for definitive diagnosis [4]. On conventional radiography and CT, we can see lytic lesions with variable grades of calcification and with tendency to extend to the soft tissues. Sometimes it can be heavily calcified mimicking osteogenic sarcoma, chondrosarcoma or calcified metastases [5]. MRI findings are more suggestive for diagnosis, although not pathognomonic. MRI findings consist of low-signal on T1WI and very low signal on T2WI with a variable enhancement, usually weak [6]. 18F-FDG PET has been considered a reliable method for the detection of active multiple myeloma; however, evidence of its diagnostic power in amyloidosis remains lacking [7]. Localised amyloidosis tends to show mild-to-moderate increased FDG uptake, similar to multiple myeloma and plasmacytoma. Besides, the molecular mechanism for 18F-FDG uptake remains speculative and probably related to inflammatory changes associated [8]. In this case, considering the instability of the fracture, an occipitocervical fixation was performed. During surgery, samples of bone and soft tissue were obtained for pathological analysis. Congo red staining and green apple birefringence confirmed the presence of extensive amyloid deposition. Written informed patient consent for publication has been obtained.

Differential Diagnosis List: Cervical amyloidoma causing pathologic fracture of C2, Plasmocytoma, Metastases, Multiple myeloma, Amyloidoma

Final Diagnosis: Cervical amyloidoma causing pathologic fracture of C2

References:


Figure 1

Description: Cervical spine X-ray, sagittal view. Lytic lesion of the odontoid process with pathologic fracture (white arrow). Origin: © Department of Radiology, Complexo Hospitalario Universitario A Coruña/ Spain 2018.
Description: Cervical spine CT, bone window. Confirms transversal dens fracture and shows a lytic lesion that affects the odontoid process and the anterior arc of C1 (white arrows on A). The lesion also affects the lateral mass of C1 and the body of C2 (black arrows on B). Vertical fracture of the left superior articular process of C2 (arrowhead on B). Origin: © Department of Radiology, Complexo Hospitalario Universitario A Coruña/ Spain 2018.
Description: Cervical spine CT, bone window. Confirms transversal dens fracture and shows a lytic lesion that affects the odontoid process and the anterior arc of C1 (white arrows on A). The lesion also affects the lateral mass of C1 and the body of C2 (black arrows on B). Vertical fracture of the left superior articular process of C2 (arrowhead on B). Origin: © Department of Radiology, Complexo Hospitalario Universitario A Coruña/ Spain 2018.
Description: Cervical spine CT, soft tissues window. Extension into the epidural space of a high-density soft tissue (white arrows on A). Nodular component of the high-density soft tissue which extends to the right prevertebral space (black arrows on B). Origin: © Department of Radiology, Complexo Hospitalario Universitario A Coruña/ Spain 2018.
Description: Cervical spine CT, soft tissues window. Extension into the epidural space of a high-density soft tissue (white arrows on A). Nodular component of the high-density soft tissue which extends to the right prevertebral space (black arrows on B). 

Origin: © Department of Radiology, Complexo Hospitalario Universitario A Coruña/ Spain 2018.
**Description:** Cervical spine MR. Low intensity of the odontoid process on T1WI (white arrow on A) with a very low signal of the lesion on T2WI (white arrows on B) and STIR (white arrow on C). No enhancement after administration of Gd (arrowheads on D).

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Description: Cervical spine MR. The nodular component of the prevertebral soft tissues also shows a very low signal on T2WI (white arrow on A) with a mild enhancement after Gd (arrowheads on B).

Origin: © Department of Radiology, Complexo Hospitalario Universitario A Coruña/ Spain 2018.
Description: Cervical spine MR. The nodular component of the prevertebral soft tissues also shows a very low signal on T2WI (white arrow on A) with a mild enhancement after Gd (arrowheads on B).

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