Primary solitary amyloidoma of the sacrum - a case report and review of the literature.

Frank M. Klenke1, Christian Wirtz1, Yara Banz1, Lorin M. Benneker1

1) Inselspital, Bern University Hospital, Switzerland

Background:
Primary solitary amyloidoma of the axial skeleton is rare. This tumor-like lesion may have a particularly aggressive appearance characterized by local deposition of amyloid and bone destruction that can result in segmental instability. The thoracic spine is most commonly involved, followed by the cervical spine. Here we report the case of a primary, solitary amyloidoma of the sacrum, its diagnosis, treatment, and outcome.

Methods:
Case description and systematic review of the literature.

Results:
A 64-year-old female with a history of several years of back pain presented with recent exacerbation of local pain, weakness and paraesthesia of the right leg and increasing difficulties in bladder control. Imaging studies revealed an osteolytic tumor of the sacrum with extradural extension and obliteration of the spinal canal. Histopathological diagnosis of a plasmacytoid amyloidoma was made by CT-guided needle biopsy. Intralesional resection, lumbo-pelvic stabilization and postoperative radiotherapy resulted in complete resolution of neurologic symptoms. One year after surgery the patient is free of local disease. Literature review yielded less than 30 cases of primary amyloidoma of the spine. Only two previous reports of amyloidoma of the sacrum were identified. To our knowledge, this is the first case of solitary amyloidoma of the sacrum treated with surgical resection, stabilization and radiotherapy.

Conclusion:
Primary amyloidoma of the sacrum is extremely rare. Clinical presentation may be nonspecific. Imaging features are variable and can mimic malignant tumor growth. Definitive diagnosis requires histopathologic examination and immunohistochemical specification. Systemic affection must be excluded with cardiac MRI as well as colon and kidney biopsies. At the level of the spine and the sacrum resection is recommended in cases with extensive bone destruction and neurological symptoms. Intralesional resection with postoperative radiotherapy is associated with significant improvement of clinical symptoms and good local tumor control.

E-mail (main author): frank.klenke@insel.ch