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Review Article

Pathological axis fracture secondary to a solitary bone plasmacytoma: Two cases and a literature review

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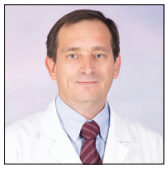

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ABSTRACT

Background: Solitay bone plasmocytoma (SBP) account for just 5–10% of all plasma cell neoplasms. They are infrequent in the cervical spine, especially involving the C0–C2 segment. In this article we conducted a literature review and present the diagnosis, management and long term course of two patients with SBP of C2 causing cervical instability.

Methods: We assessed the clinical records of two patients with SBP in C2 and cervical instability attributed to SP-B involving C2. Both patients presented with progressive, severe cervicalgia, and the "sensation" of skull instability. Magnetic resonance imaging revealed an extensive, infiltrative lesion involving C2 vertebral body and lateral masses, consistent with a plasmacytoma.

Results: Both patients underwent emergency posterior surgical stabilization with craniocervical fixation; this was accompanied by a C2 transpedicular biopsy. Postoperatively, patients exhibited no focal neurological deficits and rapidly became pain free. They additional recieved 25 sessions of local conventional radiation therapy. Both patients are doing well as respective 2 and 7-year follow-up.

Conclusion: Although rare, unstable SBP may present atypical cervical location that readily responds to surgical descompression/fusion and radiotherapy.

Keywords: Bone cyst, Cervical vertebrae, Multiple myeloma, Solitary plasmacytoma

INTRODUCTION

Solitary plasmacytoma (SP) is a rare hematological malignancy characterized by the localized proliferation of neoplastic monoclonal plasma cells, in the absence of multiple myeloma (MM), with <10% infiltration of plasma cells into the bone marrow.[2,7,8]

SBP is infrequent in the cervical spine, accounting for about 8% of cases[4] and involve C0–C2 in just 0.5% of the time.[5] Clinical presentation depends on size/severity, location, extent of bony involvement and epidural spinal extension/cord compression.[5]

We reviewed two patients with SBP at C2 who presented with severe axial neck pain and craniocervical instability treated with posterior descompression/stabilization and postoperative radiotherapy.

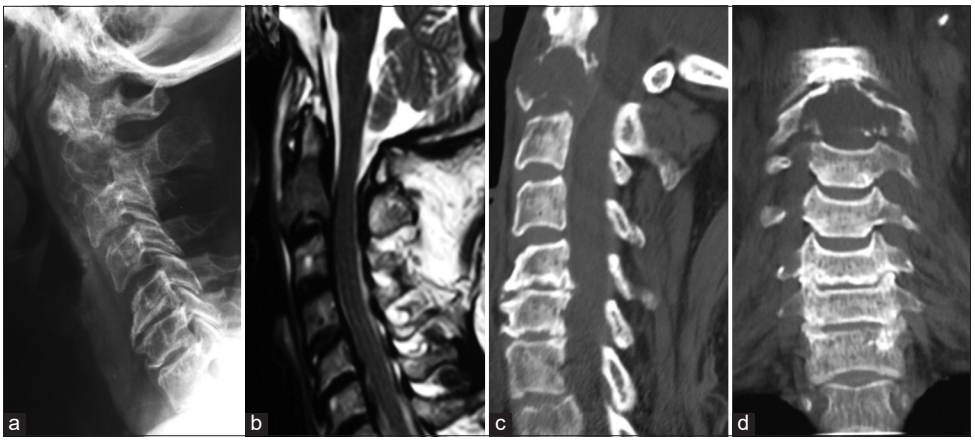

Figure 1: Case 1: Preoperative imaging. (a) Radiographs show a radiolucent C2 vertebral body, (b) T2-weighted magnetic resonance imaging (MRI) reveals hypointensity of the C2 vertebral body, (c and d) sagittal and coronal CT scan reveals instability due to severe lytic and destructive lesion in the vertebral body at C2.

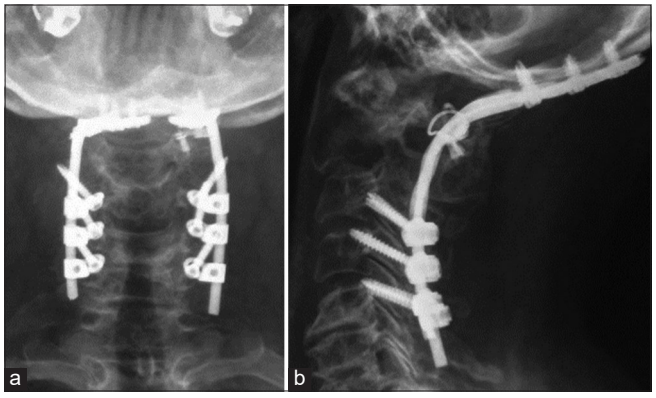

Figure 2: Case 1: Postoperative imaging. (a and b) PA and lateral radiographs – long-term postoperative follow-up at 1 year demonstrates progressive bone healing.

Case 1

A 73-year-old female presented with 1.5 months of progressive, severe cervicalgia, and a C2 bilateral root neurological deficit.

Cervical X-rays revealed a phatologic fracture of C2 secondary to a vertebral body lytic lesion. CT scan showed the lytic lesion involving both lateral mases of C2 with pathological fracture. Magnetic resonance (MR) showed partial epidural compromise [Figure 1]. Bone scintigraphy only showed the C2 lesion. Notably, plasma protein electrophoresis was normal.

Surgery

Posterior C0-C5 fusion with craniocervical fixation under halo traction was performed accompanied by the placement of iliac crest autograft (i.e., using the CerviFix rod system Synthes®; Stratec Medical, 4436 Oberdorf, Switzerland) accompanied by the placement of iliac crest autograft. Intraoperatively, anterior displacement of C0–C1 over C2 required titanium braided wires to achieve reduction. An additional posterior transpedicular biopsy was performed of C2 [Figure 2]. Postoperatively, the patient did well, exhibiting no new neurological deficits, with X-rays, CT, and MR studies confirming stability.

Histopathology

Histopathological and immunohistochemical studies of the lesion were consistent with a plasmacytoma (i.e., the diagnosis of SP-B was confirmed). Later, the patient underwent conventional radiotherapy (RT) (25 sessions [50 Gy]) with complete remission. Seven years later, the patient remains intact, without any evidence of recurrent disease.

Case 2

A 56-year-old male presented with 1 month of progressive, severe posterior neck pain and craniocervical paresthesias.

CT scan revealed an extensive, infiltrative C2 vertebral lesion that compromised vertebral body, the lower third of odontoid process, both lateral masses, pedicles and facet processes, and part of the axis laminae. The lesion had a low signal intensity on T1 WI MR, high signal intensity on T2 and STIR WI MR and irregularly enhanced contrast images without epidural extension [Figure 3]. The positron emission tomography (PET) CT demonstrated a monostotic C2 lytic lesion with peripheral hypermetabolism associated with a pathological bone fracture. Interestingly, Bence-Jones protein, protein electrophoresis, free light chains, flow cytometric immunofixation, and screening of lymphocytes were normal.

Surgery

Surgery was performed under neuromonitoring and neuronavigation support and included a bone biopsy

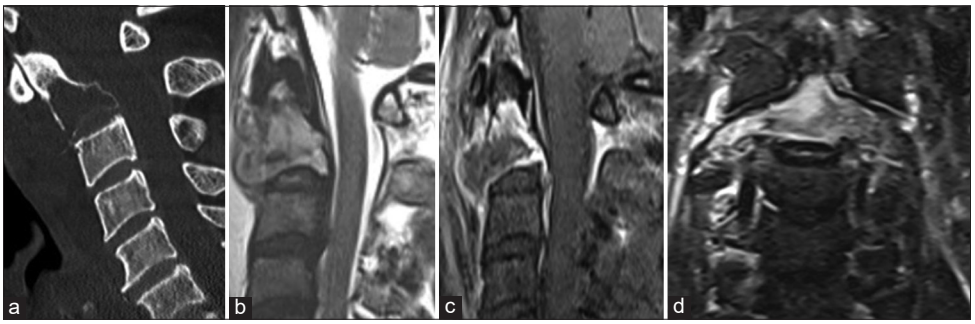

Figure 3: Case 2: Preoperative imaging. (a) CT scan shows listhesis and instability due to an infiltrative, destructive lesion in the vertebral body at C2, (b) T2-weighted MRI reveals hiperintensity of the C2 vertebral body, (c and d) MRI shows contrast enhancement in vertebral body, pedicles and the C2 laminae.

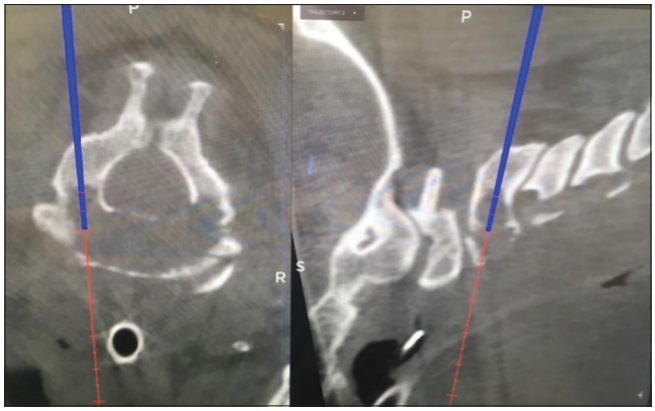

Figure 4: Case 2: C2 transpedicular bone biopsy was taken under neuronavigation support.

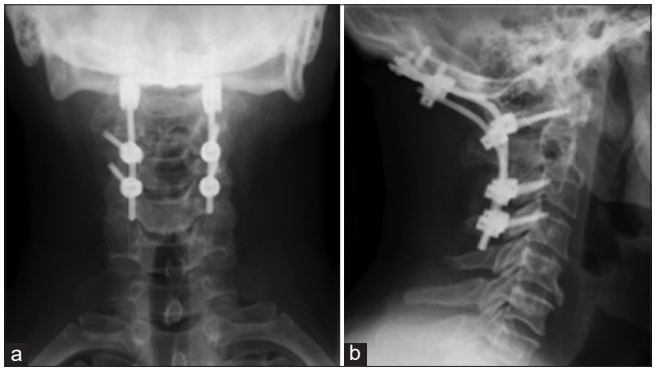

Figure 5: Case 2: Postoperative imaging. (a and b) PA and lateral radiographs – long-term postoperative follow-up at 1 year demonstrates progressive bone healing.

through the C2 pedicles [Figure 4], a C0–C4 occipitocervical fixation (Vertex rod system, Medtronic, Inc.) and fusion supplemented with bone-chip grafting [Figure 5].

Postoperative course

Within 1 day, the patient had no pain or neurological deficits. The myelogram was normal, and postoperative images documented good implant alignment and position. He was discharged on the 5th postoperative day with a Miami J collar. Histopathological and immunohistochemical studies of the lesion were consistent with the diagnosis of a SP-B.

Conventional RT (25 sessions: 45 Gy) was begun in the 6th postoperative week. Three months later, the whole-body PET CT revealed adequate local control, without other bone lesions and good alignment. However, at the current time, the clinical course appears consistent with smoldering MM, including a slight decrease in light chains.

Nevertheless, 21 months later, the cervical CT and PET showed an intact patient with good local control without recurrence of the C2 lesion.

DISCUSSION

SBP accounts for just 3% of all plasma cell neoplasms.[10] It is especially rare (0.5%) in the upper cervical region[4] where it has an especially poor prognosis.[2] The mean age of patients diagnosed with SP-B is 55 years, with males predominantly (65%) affected.[6,10]

Approximately two-thirds of SBP eventually progress to MM within 1.75–4 years of diagnosis.[6,10] There is currently no effective method to prevent SBP from progressing to MM.[5]

The current international myeloma working group criteria for SBP are (1) a solitary bone or soft tissue lesion, verified by bone biopsy with evidence of clonal plasma cells; (2) normal bone marrow, without evidence of plasma cells or, failing that, <10% involvement; (3) no evidence of bone lesions other than the primary solitary lesion on MRI and/or CT of the spine and pelvis; and (4) the absence of any target organ damage (e.g., hypercalcemia, renal failure, and anemia).[8]

Tissue biopsy and histological and immunohistochemical findings, identifying the presence of a homogeneous infiltrate of monoclonal plasma cells, make it possible to establish the initial diagnosis.[2] MRI is the gold standard imaging study for

the initial diagnosis. Both MRI and PET-CT have been shown to be of vital importance excluding progression to MM and to, thereby, be the most recommended follow-up imaging studies.[9] Posterior transpedicular biopsy is a good option for confirming the diagnosis of SBP and the posterior approach, at the same time allows the surgeon the reduction and fixation of any craniocervical instability.

Radiation therapy is first-line treatment for SBP in many patients,[3,8,11] However, in patients with spinal compression, neurological symptoms, and/or severe instability, RT is often combined with decompressive surgery and vertebral stabilization.[3,11] Ahmadi *et al*. suggested an algorithm with occipitocervical fixation as the surgical treatment option for SBP at the craniocervical junction for treating secondary instability.[1]

Despite the high rate of local control with RT and/or surgery, rates for tumor recurrence and progression to MM are high.[6] Nevertheless, in our two patients with SP-B, (e.g., despite the likely evolution of MM in one patient), tumor has not clinically become symptomatic in the postoperative periods ranging from 2 to 7 years.

CONCLUSION

SBP involving the C2 cervical vertebral body is very rare but may be successfully managed with biopsy, decompression, and fusion.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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