Solitary Plasmacytoma of the Axial Spine and Dorsal Spine: Treatment Dilemmas and Review of Literature

Kongwad LI¹, Nair RP²*, Naylor B³ and Nagaraj A²

Abstract

Background: Solitary plasmacytoma of the spine accounts for 5% of plasmacytomas and is a rare entity [1]. More than 25-60% of these lesions are localized in the dorsal spine and cause myelopathy in 42-71% of the patients. Diagnosis and treatment protocols have been established, however the final decision of whether to stabilize and irradiate locally versus direct irradiation is controversial. Many centers advocate different treatment protocols based on their institutional experience and patient outcomes.

Clinical presentation: We present, herewith, our experience with a 63 year-old patient with upper dorsal solitary lesion, who presented with upper back pain and was diagnosed to have a D3-4 solitary plasmacytoma and another 44 year old patient who presented with spastic quadriaparesis progressing to quadriplegia over the last 6 months, who had a C2 lytic lesion with instability.

Conclusion: In the presence of solitary spinal lesions, despite the location, solitary plasmacytoma of the bone should be considered as one of the differential diagnosis. Abnormal proteinemia or proteinuria may often be absent, yet this entity is commonly encountered in clinical practice. Patients show clinical and neurological improvement with surgical decompression with/ without stabilization. Postoperatively, radiotherapy is advocated since it reduces the recurrence rates. Often confused with spinal tuberculosis, starting ATT can be detrimental to the patient since it delays the standard line of treatment.

Keywords

Cervical plasmacytoma; Dorsal plasmacytoma; Solitary plasmacytoma; Lytic lesion; Instrumentation and spinal irradiation

Case Report

Case 1

A 63 year-old gentleman presented to the outpatient department with the history of upper back pain radiating to bilateral shoulders since 2 years. He had been seen at various local hospitals and was started on analgesics to relieve his symptoms, but to no avail. The pain was radiating to the shoulder girdle [L>R], which aggravated on movement, however there were no motor deficits. On general examination he had a D3-D4 level gibbus with no spinal tenderness.

Neurological examination revealed an increase in the lower limb tone [Grade 1 Modified Ashworth]. Power seemed to be Grade 5/5 in all four limbs. Sensory [pin prick] seemed to be decreased below D3 on left and below L1 on right. Lower limb reflexes were brisk and plantars were upgoing. A clinical diagnosis of a D1 level lesion with myelomalacia [UMN signs] was made and investigations were done for localizing the lesion.

MRI Dorsal spine [done at an outside hospital] showed a D3 vertebral body destruction with epidural collection between D2-D4 level with myelomalacic changes with involvement of the posterior element of D3 (Figure 1a,b).

CT Dorsal spine showed a lytic lesion involving D2-D4 vertebrae with extension into the posterior elements of D3, with collapse of D3 body and significant paravertebral and epidural collection (Figure 2).

Patient underwent D2 to D4 laminectomy with transpedicular decompression of tumour and D2 and D5 transpedicular screw stabilization (Figure 2, far right colour plate). Postoperatively he was...
ambulated with a Taylor’s brace and discharged within a fortnight. Pathology rendered a diagnosis of Solitary plasmacytoma of the spine (Figure 7a) and he was subjected to adjuvant radiotherapy. Prior to irradiation, peripheral smear along with Bence jones proteins [in urine] were done, to rule out multiple myeloma. He underwent irradiation of the spine (C6 to D5 level). He received a total of 30Gy in 10 fractions over 2 weeks. Followup MRI showed insignificant residual lesion with no evidence of bone marrow changes and serum electrophoresis showed NO M bands (Figure 3).

Case 2

A 44 year-old gentleman, presented to the outpatient department, with the history of neck pain since 8 months. It was the first time that he was being evaluated for the same. The pain was non-radiating in nature and localized to the posterior aspect of the nape of the neck. He also complained of progressive weakness in all four limbs since the last 2 months. The progression of the weakness followed an Elsberg pattern, starting from the right lower limb. He had been bed bound since the last 3 weeks, requiring assistance for all daily activities. There was no history of cough or valsalva headache and no history of trauma.

On general examination, he was moderately built and nourished with severe, painful restriction of neck movements. Neurological examination revealed spastic quadriparesis (Modified Ashworth Grade 2A). His single breath count was grossly reduced with restricted chest expansion. He had no CVJ markers and no evidence of Horners syndrome or hearing loss. There were no obvious cranial nerve deficits including trigeminal nerve. On Motor examination, he had a flicker of movement in bilateral upper limbs (Grade 1/5) and Grade 1/5 and Grade 2/5 in the right and left lower limbs respectively. Sensation (Crude touch and pin prick) was diminished below C3-4 segments with exaggerated deep tendon reflexes in all four limbs. Superficial reflexes were absent and plantars were upgoing. A clinical diagnosis of upper cervical myelopathy secondary to cranio-vertebral junction pathology was considered and further investigations were done.

Cervical spine x ray showed a well-defined expansile lytic lesion involving the anterior and posterior elements of the C2 vertebra with associated cortical thinning (Figure 5). A corroborative computed tomographic (CT) scan of the cervical spine showed a well-defined...
lytic lesion involving the entire C2 vertebra with thinning of the cortex. There was no evidence of basilar invagination (Figure 4).

Magnetic resonance imaging of the cervical spine showed an expansile, lytic lesion, involving the entire C2 body [anterior and posterior elements], hypointense on T1W and hyperintense on STIR with restriction on DWI with post contrast enhancement (Figure 4). There seemed to be a soft tissue component, with possible collection, around the C2 vertebral body, tracking along the para-spinal muscles [more in the left side], extending from the superior end plate of C4 vertebra to the base of the skull. Anteriorly the collection seemed to involve the retropharyngeal space with scabbard appearance of the nasopharynx and oropharynx with maintenance of flow voids within the vessels.

Patient underwent an occipito-cervical fusion with tumour decompression (Figure 6). Post operatively, he was started on aggressive physiotherapy and he showed significant neurological improvement. His power improved to Grade 2/5 on right side and Grade 3/5 on the left.

Histopathological examination proffered a diagnosis of plasmacytoma with CD 138 positivity and he was subjected to adjuvant radiotherapy (Figure 7b).

Discussion

The optimal treatment for solitary plasmacytoma of the bone [SPB] is still controversial. Local radiation therapy without surgery is often employed [2,4], but no randomized study has confirmed its superiority over surgery alone. Often encountered in patients in the sixth decade of life, more than 80% of plasmacytoma occur in D6 - L4 region, of these, 50% are seen within D11 - L1 segments, due to biomechanical factors acting on the dorso-lumbar transition zone. It is often difficult to differentiate between potts’ spine, pyogenic and fungal vertebral osteomyelitis from primary and metastatic spinal deposits based on clinical and radiographic findings alone.

Since tuberculosis is almost ubiquitous in the Asian continent, it is often considered as the first diagnosis in young patients with an epidural collection with vertebral body destruction, in the dorsal and dorso-lumbar region, usually without significant neurological deficits. These patients are erroneously started on ATT (Anti-Tubercular treatment) without any confirmatory tests in lieu with the Middle pathway regimen as described by Tuli et al. [6]. This usually is associated with progression of symptoms and subsequent spinal instability. Taneichi et al. established a criteria [7] for vertebral collapse as a result of osteolytic metastatic lesions, they were based on:
Various diagnostic criteria have been described for diagnosing plasmacytomas, however we have adhered to the IMWG diagnostic criteria [3] (Table 1). Harrington et al. had established that patients who had more than 50% of the vertebral body destroyed required posterior stabilization along with decompression of the lesion [8]. With our experience in 2 patients, one with an upper dorsal lesion and the other with a C2 lesion, we recommend that all patients with an extradural compression, in the presence of neurological deficits, should undergo surgical decompression in the form of transpedicular decompression of the tumour with posterior stabilization. These patients [once biopsy proven] were also advised to undergo subsequent radiotherapy after 6 to 8 weeks.

In general about 75% of patients, with apparent solitary
plasmacytomas, progress to develop Multiple Myeloma. Most of these patients may have a positive monoclonal protein in blood or urine (<10 g/l) [5]. The largest series of solitary plasmacytoma, published in 2006 [4] predicted that more than one half of the patients with SPB develop multiple myeloma with a median time to development of 21 months (range 2-135 months). 5 and 10 year probability rates of developing multiple myeloma were 51% (95% CI, 43-59%) and 72% (95% CI, 62-82%), average disease free interval, after diagnosis of solitary plasmacytoma, was 81 months and the 10-year survival rate was 85%. Age more than 60 years was the only independent predictor of development of multiple myeloma [4]. Thus patients with solitary plasmacytoma can expect a benign course and a prolonged progressive free survival, unless the lesion progresses and manifests as a more moribund multiple myeloma.

**Conclusion**

We strongly recommend that patients with spinal compressive lesions should undergo decompression with or without stabilization and adjuvant radiation in lieu of diagnosis and to reduce the conversion to multiple myeloma. Radiation alone is advocated in patients who may not withstand surgery; however they have a bad prognosis and bleak survival rates. Surgical decompression offers improved progression free survival rates with a relatively benign course.

**Table 1:** International Myeloma Working Group diagnostic criteria of solitary plasmacytoma of bone, extramedullary plasmacytoma and multiple solitary plasmacytomas (+/- recurrent).

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<th>Diagnosis</th>
<th>Criteria</th>
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<tr>
<td>Solitary plasmacytoma of the bone</td>
<td>No M-protein in serum and/ or urine</td>
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<td></td>
<td>Single area of bone destruction due to clonal plasma cells</td>
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<td></td>
<td>Normal skeletal survey (and MRI of spine and pelvis if done)</td>
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<td></td>
<td>No related organ or tissue impairment</td>
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<tr>
<td>Extramedullary plasmacytoma</td>
<td>No M-protein in serum and/ or urine</td>
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<td></td>
<td>Extramedullary tumor of clonal plasma cells</td>
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<tr>
<td></td>
<td>Normal bone marrow</td>
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<tr>
<td></td>
<td>Normal skeletal survey</td>
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<tr>
<td></td>
<td>No related organ or tissue impairment</td>
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<tr>
<td>Multiple solitary plasmacytomas (+/- recurrent)</td>
<td>No M-protein in serum and/ or urine</td>
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<tr>
<td></td>
<td>More than one localized area of bone destruction or extramedullary tumor</td>
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<td>of clonal plasma cells</td>
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**References**