Case report

A young patient with cord compression diagnosed as a solitary sacral plasmacytoma: a case report

Ishara Dissanayaka[1*], Wasantha Karunarathna[1], Prasanna Weerawansa[2]

[1] Postgraduate institute of medicine, University of Colombo.
[2] Faculty of Medicine and Allied Sciences, Rajarata University of Sri Lanka.

Abstract

Plasmacytoma is a subgroup of plasma cell neoplasms characterized by localized proliferation of neoplastic plasma cells. It is a rare disease that accounts for only less than 5% of all plasma cell tumours and can present alone or with multiple myeloma. Here, we report a clinical case of unusual presentation, in which solitary sacral plasmacytoma of a previously well young woman presented with bilateral lower limb numbness. She was found to have a sacral-originated tumour, which was later diagnosed as a solitary sacral plasmacytoma with the help of imaging and histological evaluation, along with the absence of the end organ damage.

Keywords: Solitary plasmacytoma, Multiple myeloma, Conus medullaris

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*✉**Correspondence:** iuduop@gmail.com https://orcid.org/0009-0006-8744-8087

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Introduction

Solitary plasmacytoma is a haematological neoplasm where a proliferation of plasma cells occurs locally with the absence of systemic involvement [1]. These tumours can be sub-classified into two groups: solitary plasmacytoma of bone (SBP) and extramedullary plasmacytoma (EMP) [2]. Solitary plasmacytoma of bone is an osseous tumour of proliferating plasma cells, manifesting as a mass lesion alone or as multiple myeloma [3]. Extramedullary plasmacytomas are found in the head and neck region involving the nasopharynx and the nasal cavity [4].

According to the international myeloma working group, solitary plasmacytoma is defined as a biopsy-proven solitary lesion of the bone or soft tissue with evidence of clonal plasma cells without bone marrow involvement (without clonal plasma cells), normal skeletal survey and MRI of the spine except the primary solitary lesion and absence of end-organ damage including hypercalcemia, renal insufficiency, anaemia or bone lesions. When clonal bone marrow plasma cells are found in less than 10% of cases, this entity is called solitary plasmacytoma with minimal marrow involvement [5]. With time, most of the plasmacytomas evolved into multiple myeloma or multiple solitary

lesions, while only a few can remain without further transforming [2].

Solitary plasmacytoma consists of 70% of all plasmacytomas and generally arises from the body of vertebrae, pelvis, and femur. Thoracic and lumbar vertebrae with sacral involvement were identified as the commonly affected regions [2,6]. Thus, the clinical presentation varies according to the site of involvement.

Case presentation

A 45-year-old woman who was previously healthy presented to the medical ward with gradually worsening lower back pain and bilateral lower limb numbness for 4 weeks duration. Later, she developed faecal incontinence. There was no history of trauma or any constitutional symptoms such as fever, loss of appetite or loss of weight.

On admission, bilateral lower limb power was grade 4+, and exaggerated knee jerks and diminished ankle jerk response were detected in both lower limbs. Sensory nervous system evaluation identified impaired sensory perception in L1- L3, L5 and S1- S3 levels suggestive of saddle anaesthesia. Babinski response was negative. No abnormalities were detected in the neurological examination of upper limbs and other system examinations.

Her investigations (Table-1) revealed normocytic normochromic aneamia in her full blood count (FBC). Her erythrocyte sedimentation rate (ESR) was elevated with normal c-reactive protein (CRP)

Table 1: Biochemical findings of the patient

Investigations	
Full Blood Report	
White Cell Count (ml^{-1})	$10.1x10^3$
Hemoglobin (g/dL)	11
Platelets (ml^{-1})	$343x10^3$
Erythrocyte Sedimentation rate (mm/hr)	60
C-reactive protein (mg/dL)	6
Serum Procalcitonin	0.23
AST (u/L) (Aspartate aminotransferase)	236
ALT (u/L) (Alanine aminotransferase)	257
ALP (u/L) (Alkaline phosphatase)	88
Total Bilirubin (umol/L)	4.43
Plasma albumin (g/L)	41.21
Plasma globulin (g/L)	29.39
Serum Creatinine (umol/L)	85.5
Corrected calcium (mmol/L)	2.27

Her blood picture showed normocytic normochromic red cells with a mild degree of rouleaux formation and neutrophil leukocytosis.

Non-contrast MRI spine revealed a mass lesion involving S1 and S2 vertebral bodies and left sacral ala with measurements of 4.7cm x 3.4cm x 6.9cm, which has produced moderate canal stenosis, cauda compression and left side exiting sacral foramina stenosis with nerve root compression at S1/S2 and S2/S3 (Figure 1A). Contrast-enhanced CT showed evidence of lytic lesions, confirming the diagnosis (Figure 1B).

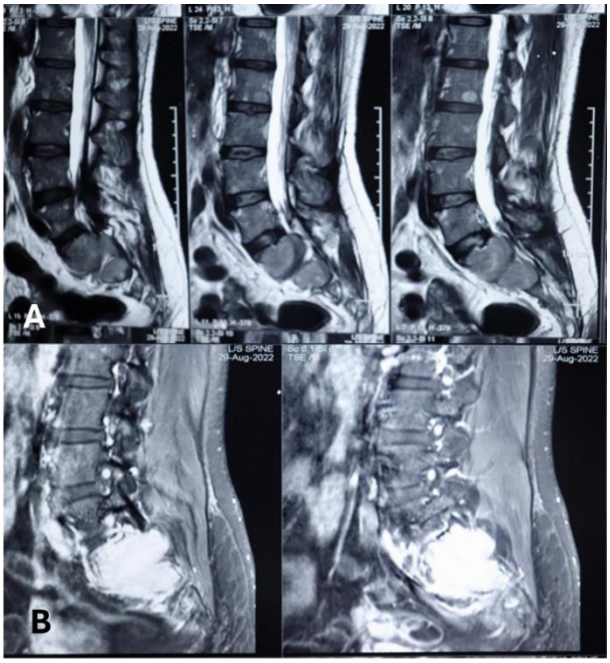

Figure 1: A, non-contrast MRI images of the sacral lesion - T1 hypointense mass lesion at S1 and S2 vertebral level; B, Contract CT - T2 hyperintense mass lesion at S1 and S2 vertebral bodies and sacral ala

Histology from a CT-guided biopsy of the sacral mass showed a neoplasm composed of sheets of atypical cells with eccentric nuclei along with nuclear pleomorphism and occasional mitoses. Immunohistochemical staining with CD 138, demonstrated positivity for lambda light chains. The appearance was compatible with plasmacytoma. However, serum protein electrophoresis did not reveal monoclonal gammopathy, and Serum-free light chain assay was normal.

Moreover, her bone marrow trephine biopsy revealed small focal collections and scattered plasma cells accounting for 8-9% along with hyperplastic erythropoiesis with normoblastic maturation, active

granulopoiesis and normal megakaryopoiesis without evidence of fibrosis or infiltration.

Ultimately, she was diagnosed with solitary plasmacytoma of the sacrum with minimal marrow involvement as she fulfilled the criteria [4]. She was started on radiotherapy, with which she achieved a remarkable improvement in her lower limb numbness and faecal incontinence over six months.

Discussion

Here, we present a clinical case of a female patient coming with insidious onset neurological disability without significant back pain turned out to be a plasma cell tumour without systemic involvement.

Plasma cell tumours are broadly categorized into three groups: multiple myeloma, extramedullary plasmacytoma, and solitary bone plasmacytoma [1]. The Median age of the patients with SBP or EMP is 55 years. The male-to-female ratio of both is 2:1[4]. Whereas the median age of diagnosis of multiple myeloma is 69 years in the United States with 1.5 times commoner in males compared to females [7]. However, our patient, who is a female developed the disease in her mid-forties.

According to the prevailing data, plasmacytoma of the sacrum is uncommon [6].

The usual presentation of plasmacytoma is the pain at the site of involvement along with neurological features of the spinal cord and root compression, depending on the level of lesion [8]. This patient presented with back pain and bilateral lower limb numbness with faecal incontinence and examination showed features suggestive of conus medullaris.

Imaging revealed features of a malignant mass lesion located in the sacral region, which then proved as plasmacytoma with biopsy. Her bone marrow biopsy specimens revealed clonal bone marrow plasma cell proliferation <10%. She neither had lytic bone lesions except for the primary solitary lesion nor end-organ involvement. Thus, she fulfilled the criteria for the diagnosis of solitary plasmacytoma with minimal marrow involvement [5]. Therefore, we think that in patients who are presenting with clinical signs of conus medullaris, solitary plasmacytoma is one of the possible differential diagnoses that can be considered.

Generally, the modality of treatment for solitary plasmacytoma is local radiotherapy with or without surgery depending on the severity and neurological disability [8]. For our patient radiotherapy was started promptly. With that, the patient improved significantly.

However, according to the prevailing data, the prognosis is favourable with radiotherapy with a controlled rate of 88-100% [2] and 5 years estimated disease-free period is 60% and 44% can progress into multiple myeloma within around 13 years of diagnosis [3]. Thus, it is important to assess this patient periodically with time for evidence of transformation into multiple myeloma with repeated bone marrow assessment and serum-free light chain assessment. There had been no cases of solitary plasmacytomas reported below 50 years in a case study series done in 1999 [8].

Due to the rarity of plasma cell neoplasms in a patient below 50 years and uncommon presentation as a sacral mass this case would be unique case reference.

Consent

Informed written consent was obtained from the patient.

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