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CASE REPORT

Solitary Plasmocytoma of Bone

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ABSTRACT

Within the plasma cell dyscrasias, solitary plasmacytoma is rare. Solitary plasmacytoma of bone (SPB) is defined as a single bony lesion caused by a monoclonal plasma cell infiltrate, with no evidence of myeloma elsewhere.

All cases reported as multiple myeloma/plasmacytoma, from January 1999 to August 2002 at St. John’s Medical College and Hospital, were reviewed. After applying the diagnostic criteria, three cases conformed to the diagnosis of solitary plasmacytoma (7.5%). In the three cases of SPB, two occurred in males and one in female. The site of occurrence was in the thoracic vertebrae. Although radiotherapy alone is considered adequate, as two patients presenting in the vertebral region had neurological dysfunction, laminectomy was done in addition to radiotherapy. Light microscopy of all three cases showed a dense infiltrate of sheets of plasma cells.

The present study highlights the salient features of SPB. The frustrating aspect in the management of the SPB is the development of peripheral neuropathy, which was present in all three cases and was the presenting symptom in two of the cases as part of POEMS syndrome. Careful microscopy is required in EMP, as it can be confused with other malignancies, particularly lymphomas.

Key words: Solitary plasmacytoma, multiple myeloma

Introduction

Within the monoclonal B-cell proliferations, solitary plasmacytomases are rare. They can present as either extramedullary plasmacytoma or a solitary lesion involving a bone – solitary plasmacytoma of bone (SPB). These entities account for 3% and 5% of all plasma cell myeloma, respectively [1].

SPB is defined as a single bony lesion caused by a monoclonal plasma cell infiltrate, with no evidence of myeloma elsewhere. Despite their rarity, these variants of traditional multiple myeloma are seen regularly in a general oncologic practice, and they require specific diagnostic approaches and therapies that differ from that of standard multiple myeloma. The most common site of SPB is the spine. At presentation, pain is the most common symptom. Patients can present with polyneuropathies alone or as a part of variable constellation of features including polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin changes that has been termed as the POEMS syndrome or Crow–Fukase syndrome.

We undertook the present study to determine the frequency with which solitary plasmacytomases (SPB) are encountered and to analyse the clinical and laboratory features to emphasise the need for specific diagnostic approach.
Materials and Methods

Source of data
Forty reported cases of multiple myeloma or plasmacytoma at the Department of Pathology, St. John’s Medical College, from January 1999 to July 2002, were reviewed. Clinical data and laboratory results were retrieved from the medical records. The following diagnostic criteria were used to define the cases of SPB [1]:

1. Single area of bone destruction that showed monoclonal plasma cell infiltrate on biopsy
2. Normal bone marrow aspirate and biopsy
3. Negative skeletal survey
4. Low concentration of serum or urine monoclonal protein and preserved levels of uninvolved immunoglobulins

Three patients fulfilled the criteria and were diagnosed as SPB.

Table/Fig 1 Summary of clinical features

<table>
<thead>
<tr>
<th></th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
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<tbody>
<tr>
<td>Age</td>
<td>40</td>
<td>65</td>
<td>60</td>
</tr>
<tr>
<td>Sex</td>
<td>M</td>
<td>F</td>
<td>M</td>
</tr>
<tr>
<td>Presenting symptoms</td>
<td>Polyneuropathy</td>
<td>Polyneuropathy</td>
<td>Polyneuropathy</td>
</tr>
<tr>
<td>Examination</td>
<td>Sensory &amp; motor loss</td>
<td>Sensory loss</td>
<td>Motor loss</td>
</tr>
<tr>
<td>Associated findings</td>
<td>Systemic vasculitis Splenomegaly</td>
<td>Hepatomegaly Splenomegaly Renal cyst Diabetes mellitus</td>
<td>Lymphadenopathy Diabetes mellitus</td>
</tr>
</tbody>
</table>

Results

Clinical profile
Among 40 cases of multiple myeloma reviewed, three cases (7.5%) conformed to the diagnostic criteria. Among them two were males and one was a female. All the cases presented clinically as polyneuropathy. The clinical features are summarised in [Table/Fig 1].

On examination, none of the patients had anaemia; serum calcium levels were normal with preserved renal function. Radiological examination revealed solitary lesion in the thoracic vertebra in all the three, with rest of the skeletal survey being normal. Two of the patients had visceral organ involvement in the form of splenomegaly and hepatomegaly. Diabetes mellitus was noted in two patients.

Biopsy from the affected site revealed sheets of plasma cells [Table/Fig 2]. Serum electrophoresis was normal except for one patient who had polyclonal increase in gammaglobulins.

Follow up
Among the three patients, one had a follow-up of 3 years. After the laminectomy, which was followed by radiotherapy, there was no improvement. The patient was referred to rehabilitation department. During the follow-up for 3 years, he did not develop any other lesions.

The other two patients are being followed up till date. They are being treated for other systemic complaints such as hypertension and diabetes mellitus.
The clinical features and the results of investigations and the follow-up details are summarised in Table/Fig 1 and Table/Fig 3.

**Table/Fig 3 Result of Investigations**

<table>
<thead>
<tr>
<th>Investigations</th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
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<tbody>
<tr>
<td>Serum electrophoresis</td>
<td>No M spike</td>
<td>Polyclonal increase in gammaglobulins</td>
<td>No M spike</td>
</tr>
<tr>
<td>Bone marrow aspirate</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Bone marrow trephine</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
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</tbody>
</table>

**Discussion**

Solitary plasmacytomas account for 3% of all plasma cell myelomas. Patients presenting with SPB tend to be younger, as compared to the patients with multiple myeloma. The median age of presentation [2] is 37 ± 12 years, with a male predominance similar to the present series. A distinction between solitary plasmacytoma and multiple myeloma is essential, since the treatment protocols, prognosis and course of the disease differ.

The most common presenting symptom of SPB is pain in the area surrounding the lesion. But all the three cases in the study presented with polyneuropathy, which is often found in spinal involvement by myeloma. One of the patients had both sensory and motor loss; other two had isolated sensory or motor loss. Polyneuropathy was associated with organomegaly involving liver, spleen and lymph nodes, with two of the patients having diabetes mellitus. Thus, all the three patients fulfilled at least three criteria for the POEMS syndrome. Therefore, it is evident that it warrants a systemic search for a solitary bone lesion and M component in a case of unexplained polyneuropathy [3].

The polyneuropathy associated with plasma cell dyscrasia is mainly of the demyelinating type, in which pain and autonomic involvement is rare. Although the pathogenesis of the nerve damage is unclear, recent reports suggest that the nerve injury may be mediated by complement [4]. The organ involvement in POEMS syndrome is heterogeneous, with normal liver histology, Castleman disease-like picture in lymph nodes and spleen [3],[4]. Endocrine involvement is diverse, with hypothyroidism being the frequent abnormality encountered [4], but two of the patients had diabetes mellitus. The pathogenesis of endocrinopathy remains unexplained, although it has been postulated that antibodies directed against specific endocrine organs may be involved [3]. None of the patients had an M spike on serum electrophoresis.

On long-term follow-up, overt myeloma develops in approximately 50% of patients with SPB. Three patterns of failure occur: (1) development of multiple myeloma, (2) local recurrence and (3) development of bony lesions in the absence of myeloma [5],[6]. Those patients who develop multiple myeloma differ from the patients of classical myeloma at presentation, in that the former present at a younger age with a male predominance. There is a high incidence of neurological symptoms in cases with spinal involvement. The osteolytic lesions frequently resemble those of giant cell tumours, with absence of any monoclonal component in the serum [2].

Definitive local RT has been the treatment of choice for SPB. Most series agree that a dose of at least 4000 cGy is necessary to eradicate the local disease [1]. About two-thirds of the patients with SPB eventually develop multiple myeloma. The most common pattern of progression consists of new bone lesions, rising myeloma protein levels and development of diffuse marrow plasmacytosis, consistent with a macro-focal pattern of multiple myeloma. The median time for progression is 2 years, but occasionally patients have developed multiple myeloma as long as 15 years after radiotherapy.

Therefore, a patient of SPB should be followed up during and after treatment with serial physical examinations, SPEP and urine electrophoresis studies for evidence of dissemination or reactivation of disease. Any evidence found of a second bony lesion or progression to multiple myeloma should be evaluated fully and treated.

**References**


[4] Miralles GD, O’Fallan JR, Talley NJ. Plasma cell dyscrasia with polynuropathy. The spectrum of
