**Case Report**

**Rib Plasmacytoma and IgA Multiple Myeloma with Hyperviscosity Syndrome**

Vitorino Modesto dos Santos MD PhD1,2, Eduardo Flávio Oliveira Brito MD2, Bruno César Silva Paz MD1, Cristina Tavares Leal MD3

**Abstract**

Solitary bone plasmacytoma (SBP) can progress to generalized myeloma if not treated early. The elderly population is increasing and delays in diagnosis of plasma cell malignancies are frequent among them. Hyperglobulinemia of multiple myeloma (MM) plays a role in hyperviscosity syndrome (HVS).

A 65-year-old woman with hypertension and diabetes mellitus was admitted due to loss of appetite, muscle weakness, breathlessness and discrete expectoration, without fever. Chest X-ray showed an abnormal shadow projection on the right lung field, while computed tomography (CT) revealed an osteolytic mass at the sixth rib. There were more than 50% of plasma cells in the bone marrow samples and high IgA levels according to serum electrophoresis.

Rib plasmacytoma and overt IgA-producing myeloma with HVS were diagnosed, but treatment was unsuccessful.

Case studies may enhance the awareness about this ominous condition, which may develop unnoticed, particularly in elderly patients with renal insufficiency, and can pose difficulties with diagnosis in primary care settings.

**Keywords:** Chest wall, myeloma, plasmacytoma, solitary rib tumor

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**Introduction**

Solitary plasmacytomas are characterized by a bone or soft-tissue mass, less than 5% of plasma cells in the bone marrow, and the absence of anemia, hypercalcemia, or renal dysfunction.1 These rare tumors can mimic primary chest wall tumors. Solitary plasmacytoma of the bone (SPB) has a good outcome if treated early with surgery or radiotherapy. Unrecognized SBP can progress to multiple myeloma (MM).1 In patients with MM, hyperglobulinemia and cell aggregation can be a source of hyperviscosity syndrome (HVS), which can develop unrecognized and may be mistaken as concomitant renal insufficiency.2,3

**Case Report**

A 65-year-old woman with arterial hypertension and diabetes mellitus presented to the Emergency Department with muscular weakness and loss of appetite, associated with dyspnea on physical exertion of one-month duration. She had productive cough with hyaline expectoration, but denied any fever. There were palpitation episodes, mental disorientation, and inversion of the sleep pattern. Her hypertension and diabetes were controlled with atenolol 50 mg (bid), losartan 50 mg (qd), diamicron 30 mg (qd), and metformin 850 mg (bid). She denied tobacco smoking, alcoholism or illicit drug use. Previous chest radiography showed an opacity projection in the right lung field (Figures 1A and 1B) for which she received a course of levofloxacine, without improvement. Physical examination was remarkable for pale skin, muffled heart sounds and bilateral lung crackles. The BMI was 26.5 kg/m², and she had an abdominal circumference of 90 cm, blood pressure of 130×80 mmHg, and heart rate of 78 bpm. There was no organ enlargement or edema.

Laboratory data are shown in Table 1. The high pro-BNP level was indicative of congestive heart insufficiency and diuretic administration affected her renal function. Hemodialysis was began (3×/week) with improvement in renal function. Insulin and erythropoietin were also administered. The remainder of the laboratory analysis revealed normal fibrinogen with a highly elevated erythrocyte sedimentation rate. Except for the presence of 51% plasma cells (normal: 3%-6%), with binucleated cells and blasts, the bone marrow aspirate was unremarkable (Figure 1D).

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sessions of plasmapheresis were performed. Hemodynamic tolerance was initially good, but there was irreversible pulmonary acute edema after the third session.

Discussion

This elderly woman had chronic renal failure, respiratory disturbance, and heart failure. Blood tests showed severe anemia with elevated levels of calcium, ferritin, pro-BNP and β2-microglobulin (β2-m), and low intact parathyroid hormone (iPTH). Clinical management focused on the improvement of heart and renal functions. Thorax CT was suggestive of rib malignancy. During hospitalization, she complained of visual disturbance and lumbar pain, with paraparesis. Laboratory data strengthened the hypothesis of HVS associated with malignancy. Differential diagnoses of stroke, brain metastases, and intracranial hypertension were eliminated.

As with the present case, the classical triad of HVS may be incomplete, and mild or atypical clinical signs and symptoms of HVS may develop unnoticed. This often occurs in patients with

Table 1. Routine laboratory data of a 65-year-old woman with rib plasmacytoma and IgA-producing myeloma with hyperviscosity syndrome (HVS).

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Day 1</th>
<th>Day 9</th>
<th>Day 14</th>
<th>Day 16</th>
<th>Day 22</th>
<th>Day 43</th>
<th>Normal range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Erythrocytes</td>
<td>1.81</td>
<td>2.91</td>
<td>3.08</td>
<td>2.26</td>
<td>2.02</td>
<td>2.32</td>
<td>4.4–6.0 ×10¹²/mm³</td>
</tr>
<tr>
<td>Hemoglobin</td>
<td>5.9</td>
<td>9.5</td>
<td>9.7</td>
<td>7.7</td>
<td>6.6</td>
<td>8.9</td>
<td>11.1–16.1 g/dL</td>
</tr>
<tr>
<td>Hematocrit</td>
<td>17.8</td>
<td>28.3</td>
<td>30.1</td>
<td>22.0</td>
<td>19.8</td>
<td>25.7</td>
<td>39–53%</td>
</tr>
<tr>
<td>MCV</td>
<td>98</td>
<td>97</td>
<td>98</td>
<td>97</td>
<td>98</td>
<td>94</td>
<td>80–100 fl</td>
</tr>
<tr>
<td>Leukocytes</td>
<td>7.2</td>
<td>5.9</td>
<td>6.2</td>
<td>4.9</td>
<td>4.9</td>
<td>6.6</td>
<td>4.0–11.0 ×10⁹/mm³</td>
</tr>
<tr>
<td>Platelets</td>
<td>301</td>
<td>213</td>
<td>197</td>
<td>132</td>
<td>96</td>
<td>134</td>
<td>150–450 ×10⁹/mm³</td>
</tr>
<tr>
<td>ESR</td>
<td>ND</td>
<td>ND</td>
<td>134</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>≤ 15 mm/hr</td>
</tr>
<tr>
<td>Pro-BNP</td>
<td>17,311</td>
<td>14,134</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>≤ 125 pg/mL</td>
</tr>
<tr>
<td>β2-m</td>
<td>ND</td>
<td>ND</td>
<td>141.8</td>
<td>ND</td>
<td>ND</td>
<td>35.2</td>
<td>0.7–1.8 mg/L</td>
</tr>
<tr>
<td>Sodium</td>
<td>133</td>
<td>138</td>
<td>140</td>
<td>143</td>
<td>137</td>
<td>130</td>
<td>135–145 mmol/L</td>
</tr>
<tr>
<td>Potassium</td>
<td>4.3</td>
<td>4.2</td>
<td>4.0</td>
<td>4.5</td>
<td>3.5</td>
<td>3.6</td>
<td>3.5–5.2 mmol/L</td>
</tr>
<tr>
<td>Calcium</td>
<td>1.45</td>
<td>1.56</td>
<td>1.56</td>
<td>1.53</td>
<td>1.55</td>
<td>1.32</td>
<td>1.16–1.32 mmol/L</td>
</tr>
<tr>
<td>Phosphorus</td>
<td>4.1</td>
<td>4.3</td>
<td>4.5</td>
<td>7.2</td>
<td>6.1</td>
<td>3.9</td>
<td>2.3–4.7 mg/dL</td>
</tr>
<tr>
<td>Magnesium</td>
<td>2.3</td>
<td>2.1</td>
<td>2.5</td>
<td>3.0</td>
<td>2.3</td>
<td>2.4</td>
<td>1.6–2.6 mg/dL</td>
</tr>
<tr>
<td>iPTH</td>
<td>8.6</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>11–67 pg/mL</td>
</tr>
<tr>
<td>Ferritin</td>
<td>9,145</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>13–150 ng/mL</td>
</tr>
<tr>
<td>Urea</td>
<td>136.5</td>
<td>77.5</td>
<td>120.5</td>
<td>129.0</td>
<td>111.0</td>
<td>64.4</td>
<td>10–50 mg/dL</td>
</tr>
<tr>
<td>Creatinine</td>
<td>4.2</td>
<td>3.1</td>
<td>4.9</td>
<td>5.6</td>
<td>2.2</td>
<td>3.9</td>
<td>0.7–13 mg/dL</td>
</tr>
<tr>
<td>Glucose</td>
<td>248</td>
<td>145</td>
<td>100</td>
<td>96</td>
<td>122</td>
<td>100</td>
<td>70–100 mg/dL</td>
</tr>
</tbody>
</table>

MCV = medium corpuscular volume; ESR = erythrocyte sedimentation rate; Pro-BNP = pro-brain natriuretic peptide; β2-m = β2 microglobulin; iPTH = intact parathyroid hormone; ND = not done. Abnormal data are in bold.
chronic renal failure. Asymptomatic hyperviscosity-related reti-

cnal changes have been described in a Japanese woman with MM
and renal failure associated with HVS. Nevertheless, we strongly
believe that arterial hypertension caused the fundoscopy changes
seen in our patient.

Monoclonal hypergamaglobulinemia of MM is a main cause
of HVS, which is due to changes in size, shape and polymerization
of the immunoglobulin molecules. The relationship between
HVS and high serum levels of IgA has been studied in patients
with MM, and serum viscosity depends more on the high amounts
of paraprotein than on the ratio between monomeric and dimeric
IgA. This patient presented with neurologic, ocular and cardiovas-
cular symptoms as seen in 2%–6% of patients with MM and HVS.
If the paraprotein is IgA, respective serum levels are usually more
than 6 g/dL. 

Primary chest wall tumors can mimic pulmonary conditions
on plain chest radiography. The differential diagnosis of costal plas-
macytoma includes chondrosarcoma, lymphoma, metastases, os-
teosarcoma, fibrosarcoma, neuroectodermal tumor, Ewing sarcoma,
histiocytoma, chondroma, chondromyxoid fibroma, chondroblas-
toma, giant cell tumor, fibrous dysplasia, lipoma, and bone infarct-
tion. In this case, the diagnosis of rib plasmacytoma and MM was est-
ablished by CT image, β2-m, and histopathology data. After
diagnosis, the debilitated patient who had renal failure and HVS
was treated with dexamethasone and plasmapheresis without
success. Plasmapheresis was the treatment of choice because each
session has been shown to reduce hyperviscosity in 20%–30% for
session, and usually a maximum of three sessions are necessary.

Of note was the unsuspected development of MM with the hypo-
thetical origin in a solitary costal arch mass, which was mistaken
by a pulmonary affection. Unrecognized development of MM is
not uncommon. SPB may precede the development of general-
ized myeloma, and could be predictive of an ominous progression.

The aim is to emphasize the possibility of unnoticed evolution
of a solitary rib plasmacytoma to overt IgA MM associated with
HVS. We believe that case reports may enhance the suspicion in-
dex about this condition in primary care settings.

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Table 2. Electrophoresis of serum proteins and immunoglobulins of a 65-year-old woman with rib plasmacytoma and IgA-producing myeloma with hyperviscosity syndrome (HVS).

<table>
<thead>
<tr>
<th>Serum proteins</th>
<th>Levels (normal range)</th>
<th>Immunoglobulins</th>
<th>Levels (normal range)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total proteins</td>
<td>12.6 (6.4–8.3) g/dL</td>
<td>IgG</td>
<td>204.3 (952–1538) mg/dL</td>
</tr>
<tr>
<td>Albumin</td>
<td>19.4 (55.8–66.1) %</td>
<td>IgA</td>
<td>8917 (153–359) mg/dL</td>
</tr>
<tr>
<td>Alfa1 globulin</td>
<td>2.4 (2.9–4.9) %</td>
<td>IgM</td>
<td>0.0 (73–171) mg/dL</td>
</tr>
<tr>
<td>Alfa2 globulin</td>
<td>5.1 (7.1–11.8) %</td>
<td>Kappa</td>
<td>2990 (625–1668) mg/dL</td>
</tr>
<tr>
<td>Beta1 globulin</td>
<td>2.5 (4.7–7.2) %</td>
<td>Lambda</td>
<td>39.7 (368–803) mg/dL</td>
</tr>
<tr>
<td>Beta2 globulin</td>
<td>1.9 (3.2–6.5) %</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gama globulin</td>
<td>70.6 (11.1–18.8) %</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>