A Surgical Case of Solitary Plasmacytoma of Rib Origin with Biclonal Gammopathy

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Localized solitary plasmacytoma of the bone (SPB) is a rare disease and is characterized by only one or two isolated bone lesions with no evidence of disease dissemination. A previously healthy 44-year-old male was admitted for evaluation of an abnormal radiographic shadow in the left middle lung field with symptoms of left back pain. Radiological evaluation revealed a peripheral opacity in the left chest wall, which was highly suspected to be a chest wall tumor. CT-guided transcutaneous needle biopsy of the tumor was performed and the specimens showed a monomorphous population of mature plasma cells. The bone marrow biopsy findings revealed no evidence of myeloma and bone scanning revealed only abnormal accumulation in the left seventh rib. He had mild M-proteins in a urine sample and Bence-Jones protein was detected. Immunoelectrophoresis revealed mild biclonal gammopathy of Bence-Jones protein of both the κ and λ light-chain types. Under a diagnosis of solitary bone plasmacytoma, preoperative radiation therapy with doses of 40 Gy for the tumor was performed. He underwent complete en bloc resection of the chest wall, including one-third of the left sixth and seventh ribs, the intercostal muscle and the parietal pleura. The protein abnormalities in the urine sample disappeared following surgical resection. Adjuvant chemotherapy using melphalan and prednisolone was performed. He is doing well without evidence of tumor recurrence 2 years following his initial diagnosis.

Key words: solitary plasmacytoma – biclonal gammopathy – chest wall – rib

INTRODUCTION

Primary malignant tumors of the bony chest wall are rare and localized solitary plasmacytoma of the bone (SPB) is a rare disease that accounts for only about 5% of malignant plasma cell tumors (1,2). SPB is characterized by only one or two isolated bone lesions with no evidence of disease dissemination and has been considered to be curable with radiotherapy and surgical resection and such treatment is sufficient to achieve long-term survival (3). On the contrary, SPB has been considered to be a genetic abnormality which could lead to the development of multiple myelomas (4). In this paper, a patient with a chest wall tumor is reported in whom solitary plasmacytoma originating in the rib was surgically treated with preoperative radiation therapy followed by adjuvant chemotherapy.

CASE REPORT

A previously healthy 44-year-old male was admitted for evaluation of an abnormal radiographic shadow in the left middle lung field with symptoms of left back pain. Computed tomography (CT) and magnetic resonance imaging (MRI) of the chest revealed a peripheral opacity in the left chest wall, which was highly suspected to be a chest wall tumor (Fig. 1). No associated mediastinal lymphadenopathy or intrapulmonary infiltration was evident. Bronchoscopy revealed no endobronchial abnormalities and no malignant cells were identified. A diagnosis of bone plasmacytoma was confirmed by CT-guided transcutaneous needle biopsy of the tumor. The specimens showed a monomorphous population of mature plasma cells with slight atypia (Fig. 2). The following evaluations were carried out: complete blood count with differential and platelet counts; erythrocyte sedimentation rate; serum chemistry; immu-
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noglobulin levels; serum β-2-microglobulin; serum and urine protein electrophoresis; immunoelectrophoresis and quantification of light-chain excretion; a skeletal bone survey and an iliac bone marrow aspiration biopsy. The bone marrow biopsy findings revealed no evidence of myeloma and bone scanning revealed only abnormal accumulation in the left seventh rib. He had mild M-proteins in a urine sample and Bence-Jones protein was detected. Immunoelectrophoresis revealed mild biclonal gammopathy of Bence-Jones protein of both the κ and λ light-chain types (Fig. 3). The patient fulfilled the diagnostic criteria for SPB (5): (a) a radiological lytic bone lesion with histological confirmation of plasma cell histology; (b) clinical and radiological evidence of a solitary lesion on skeletal survey; (c) an absence of myeloma cells and plasmacytosis of less than 10% in a bone marrow examination; (d) an absence of anemia, hypercalcemia or renal involvement; and (e) <2.0 g/dl M-protein in the serum. Moreover, no CD45 abnormality using Blast–Gating bone marrow aspiration was found.

Under a diagnosis of solitary plasmacytoma of the bone, preoperative radiation therapy with doses of 40 Gy for the tumor was performed. However, this was not effective in reducing the size of the tumor and he underwent surgical resec-

Figure 1. CT (top) and MRI (bottom) revealed a chest wall tumor extending into the surrounding tissues.

Figure 2. Biopsied specimens showed minimal dysplasia. Plasma cells were mature and mitoses were rare (H&E, ×50).

Figure 3. Immunoelectrophoresis revealed mild biclonal gammopathy of Bence-Jones protein of both the κ and λ light-chain types.
The protein abnormalities in the urine sample disappeared following complete en block surgical resection. Adjuvant chemotherapy was performed using melphalan 6 mg/m² p.o. daily on days 1–4 and prednisolone 40 mg/m² p.o. on days 1–4, given every 8 weeks for 1 year. The postoperative course was uneventful and a repeat skeletal survey was performed.

Figure 4. Immunohistological study of resected specimen with the avidin–biotin complex method, using rabbit anti-human γ (a, ×70), κ (b, ×20) and λ (c, ×20) chain antibodies. The tissue was stained negatively with anti-α, -µ and -γ heavy-chain antibody, but was stained positively with anti-κ and -λ light-chain antibodies.
annually. There was no serum evidence of M-protein. He is doing well without evidence of tumor recurrence or secondary neoplasms, including multiple myelomas and acute leukemia, 2 years following his initial diagnosis.

DISCUSSION

We have reported a rare finding of Bence-Jones \(\kappa\) and \(\lambda\) double gammopathy from the histochemical results for a 44-year-old male with plasmacytoma of rib origin. Primary malignant tumors arising from the bony chest wall are uncommon and SPB is a rare condition of plasma cell dyscrasias (5). Moreover, biclonal gammopathy is unusual. The incidence of SPB has been reported to be approximately 3/100,000 annually (6). SPB is an uncommon disease that accounts for only about 5% of malignant plasma cell tumors and it is less common in the chest wall than with spinal involvement; the thoracic spine is most commonly involved, followed by the lumbar spine (7). Fourteen cases of solitary plasmacytoma of rib origin have been described in the Japanese literature (8,9). The ratio of male to female patients was approximately 1.3:1. The average age on presentation was 59.5 years with a range from 39 to 77 years.

The criteria for diagnosis of solitary plasmacytoma are variable. Histopathological evidence of plasmacytoma, clinical and radiological evidence of a solitary lesion and absence of multiple myeloma features on bone marrow examination are essential. Radiographically, plasmacytoma almost always destroys bone. Recently, with advances in roentgenological diagnosis and related instruments, percutaneous needle biopsy has become a popular technique for the diagnostic evaluation of chest lesions. According to the current recommendations, the detection of a monoclonal component in the serum and/or urine does not exclude a diagnosis of solitary plasmacytoma (5). Approximately half of these patients have monoclonal gammopathy detectable with serum or urine electrophoresis. Moreover, urine electrophoresis is an important test, because it may show abnormalities in a few patients even when the serum electrophoretic pattern is normal.

In previous cases, radiation therapy was used as the primary treatment for solitary plasmacytomas. Mendenhall et al. reported a 6% local failure rate in patients with solitary plasmacytoma treated with doses of 40 Gy or above, in contrast to 31% for doses below 40 Gy (6). Aviles et al. observed that most patients treated with adequate radiation therapy alone will develop multiple myeloma within the first 3 years after diagnosis and treatment (4). The primary methods for treating solitary plasmacytoma according to Bataille and Sany were: surgery with radiation therapy, 95 cases; surgery alone, 15 cases; postoperative chemotherapy without radiation therapy, one case; no treatment, one case; and unknown treatment, two cases (5). They showed that the lowest incidence of progressive disease was observed in patients with peripheral solitary plasmacytoma treated with surgery plus an adequate dose of radiation therapy.

SPB tends to occur later in life, primarily involving marrow-containing bones, and in most cases progresses to a disseminated form indistinguishable from multiple myelomas. Although solitary plasmacytoma does appear to be a separate disease, approximately 2–10% of multiple myeloma patients who present with solitary plasmacytoma develop the more generalized disease at a later date (6). Persistence of M-protein after therapy has been reported to be an indication of a residual tumor or occult dissemination (5,7,10). On the other hand, Jyothirmayi et al. reported that no significant prognostic factors for progression to myeloma could be identified in patients with SPB in their series (11). Frasica et al. observed that there were no significant differences in overall actuarial survival among patients with or without protein abnormalities (7). Kyle et al. demonstrated that the clinical features of biclonal gammopathies are similar to those of monoclonal gammopathies (12). It has been postulated that some biclonal pairs may result from a transformation event in a cell undergoing a variable-region switch from one light-chain class to another. In our patient, the biclonal gammopathies may have resulted from the production of two monoclonal proteins by a single plasma cell clone. The tissue section and tumor cells were stained positively with both anti-\(\kappa\) and -\(\lambda\) light-chain antibodies. However, not all cells stained positively. Stained cells were scattered throughout the field. The reason for this is thought to be that the production of this protein is relatively low and varies from one cell to the next.

The role of adjuvant chemotherapy in preventing progression to multiple myeloma is controversial. Several reports suggest that adjuvant chemotherapy could increase the clearance of M-proteins and delay progression to multiple myeloma (3,13). Holland et al. showed that adjuvant chemotherapy did not affect the incidence of conversion, but did appear to delay conversion to multiple myeloma (13). Seventeen of 32 (53%)
patients with SPB converted to multiple myeloma. They concluded that lesion size, total serum protein levels and the presence of a monoclonal spike on serum electrophoresis may be of prognostic significance in identifying those solitary lesions that ultimately will convert to myeloma. Aviles et al. showed that the use of low doses of melphalan and prednisolone contributed to an improvement in disease-free survival (DFS) and overall survival in patients with SPB, compared with patients who were treated with radiotherapy alone (4). Their results suggest that the use of adjuvant chemotherapy will improve the outcome and prolong the duration of remission and survival. We conclude that complete surgical resection with preoperative radiotherapy, followed by adjuvant chemotherapy, aided the long-term survival and disease-free state of our patient.

References