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A Rare Case of Unresectable Solitary Extramedullary Plasmacytoma at the Shoulder Region: Case Report

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Abstract

INTRODUCTION: Plasmacytoma is a solitary neoplastic lesion marked by a localized accumulation of monoclonal plasma cells without an evidence of a systemic proliferation. Plasmacytoma may arise in intramedullar (bone) or extramedullar (soft tissue). Extramedullary plasmacytoma is rare, with the most frequent locations in the nasal cavity and nasopharynx. Here, we describe a case of solitary extramedullary plasmacytoma of the shoulder region.

CASE REPORT: A 49-year-old male patient presented with right shoulder mass that was solid, fixed, ulcerated, and poorly circumscribed, with size 20 × 15 × 10 cm. Shoulder X-ray and MRI results revealed a soft-tissue mass on the right shoulder with the destruction of lateral part of the right clavicle. Biopsy results suggested Non-Hodgkin Lymphoma; however, first panel of immunohistochemical (IHC) analysis showed negative staining of LCA, CK, CD20, and CD3, with high Ki67. Second panel of IHC revealed negative staining of ALK, TdT, and CD79a; meanwhile, CD138 was strongly expressed. Further hematological, biochemical, and radiological examinations that revealed no systemic involvement supported the diagnosis of solitary extramedullary plasmacytoma. Patient received radiotherapy treatment 60 Gy in 30 fractions and zometa within 4 weeks interval and showed remarkable response of the therapy.

DISCUSSION: Solitary extramedullary (soft tissue) plasmacytomas (SEP) are less common than solitary bone plasmacytoma (SBP), yet it has a better prognosis since the majority can be cured by local radiotherapy.

CONCLUSION: We report a case of SEP of the shoulder that showed remarkable response of therapy. In this case report, radiotherapy was shown to be a highly effective modality to treat a patient with solitary extramedullary plasmacytoma.

Introduction

Plasmacytoma is a solitary neoplastic lesion characterized by a localized accumulation of monoclonal plasma cells without an evidence of a systemic proliferation [1]. Clinically, solitary plasmacytoma (SP) is distinguished from multiple myeloma (MM) by the nonexistence of hypercalcemia, anemia, renal insufficiency, and also skeletal bone lesions. Plasmacytoma and MM may occur intramedullary (bone) or extramedullary (soft tissue). Solitary extramedullary plasmacytoma (SEP) is epidemiologically rare, representing only 4% of all plasma cell neoplasms [2]. Secondary SEP frequently occurs due to a direct infiltration from adjacent intramedullary MM [3]. The solitary extramedullary plasmacytoma (SEP) is commonly seen in the head and neck region, with most frequent locations in the nasal cavity and nasopharynx [4]. It is unusual for this neoplastic condition to present in the shoulder as a primary solitary lesion. Thus, in this case report, we describe our findings in a patient with SEP of the shoulder region.

Case Report

We present a 49-year-old male who came to the hospital with a complaint of a mass on his right shoulder that had been there for 1.5 years. The bulge was initially around the size of a ping-pong ball and mildly painful upon movement. One month before admission, patient complained of rapid growth of the mass and worsening pain, especially after activity and during night-time.

History of cancer within the family and weight loss was denied. On clinical examination, the mass on the right shoulder measured approximately 20 × 15 × 10 cm, solid and fixed with ill-defined borders. Shoulder radiographic result revealed soft-tissue mass over the

right shoulder region with the destruction of lateral part of the right clavicle (Figure 1). There was no other positive finding on systemic investigation.

A fine-needle aspiration biopsy (FNAB) of the mass was performed and revealed hypocellular smear of inflammatory cells with scattered of plasma cells. A tissue biopsy was then performed and revealed a diffused growth pattern of monotone tumor cells resembling Non-Hodgkin Lymphoma, large cell type (Figure 2). Initial panel of immunohistochemical (IHC) markers, including LCA, CK, CD20, CD3, and Ki67, was examined to confirm the diagnosis and determine the subtype. However, those markers were negatively stained, while the proliferation index of Ki67 was high. Considering the numerous features of plasma cells in FNAB, further panel of antibodies, including ALK, TdT, CD79a, and CD138, were used to determine the definitive diagnosis (Figure 2). In the second panel of IHC examination, only CD138 was strongly expressed in the membrane of the tumor cells, suggesting plasmacytic differentiation, so that final diagnosis of plasmacytoma was established.

Further hematological, biochemical, and radiological work-up were scheduled to exclude systemic manifestation of MM. Hematology investigation was performed and revealed normal hemoglobin level with high leucocyte (19.35 × 10^3/μL) and neutrophil count (73.9%). Erythrocyte sedimentation rate (ESR) (74 mm/h), C-reactive protein (CRP) (72 mg/ml), and lactate dehydrogenase (LDH) (905 U/l) were slightly increased.

Biochemical parameters for serum creatinine, blood urea, total protein, calcium, and serum protein electrophoresis were within normal limits. Urinary examination for myeloma protein was also negative. Bone marrow aspirate showed that plasma cells constituted 4% of the marrow cells. Because the skeletal survey did not reveal any osteolytic lesions, the investigation results leaned more toward solitary plasmacytoma. However, we encountered an unusual presentation where both soft tissue and bone involvement were present at the mass site. A contrast magnetic resonance imaging (MRI) of right shoulder was then performed to evaluate the extension of the mass. MRI results revealed a solid mass that originated from soft tissue of subcutis, infiltrating the trapezius and proximal part of the deltoid muscles (Figure 1). Imaging work-ups that aimed at the detection of distant organ involvement such as thorax CT-scan and abdominal ultrasonography were done and revealed no abnormality.

Based on the above clinical, pathological and laboratory findings, the patient was diagnosed with SEP of the right shoulder. The patient received radiotherapy of 60 Gy in 2 Gy fractions. This dose fractionation was chosen due to massive size and ulcerated tumor and its location did not place any vital organ at risk. Adaptive radiotherapy was implemented during the course of radiation with two plan due to rapid regression of the tumor (Figure 3).

The patient also received 4 mg Zometa treatment with 4 weeks interval. After radiotherapy, the patient was closely followed-up and tumor size was remarkably decreased as shown in Figure 4. After 1 year of follow-up, the patient could return to his normal

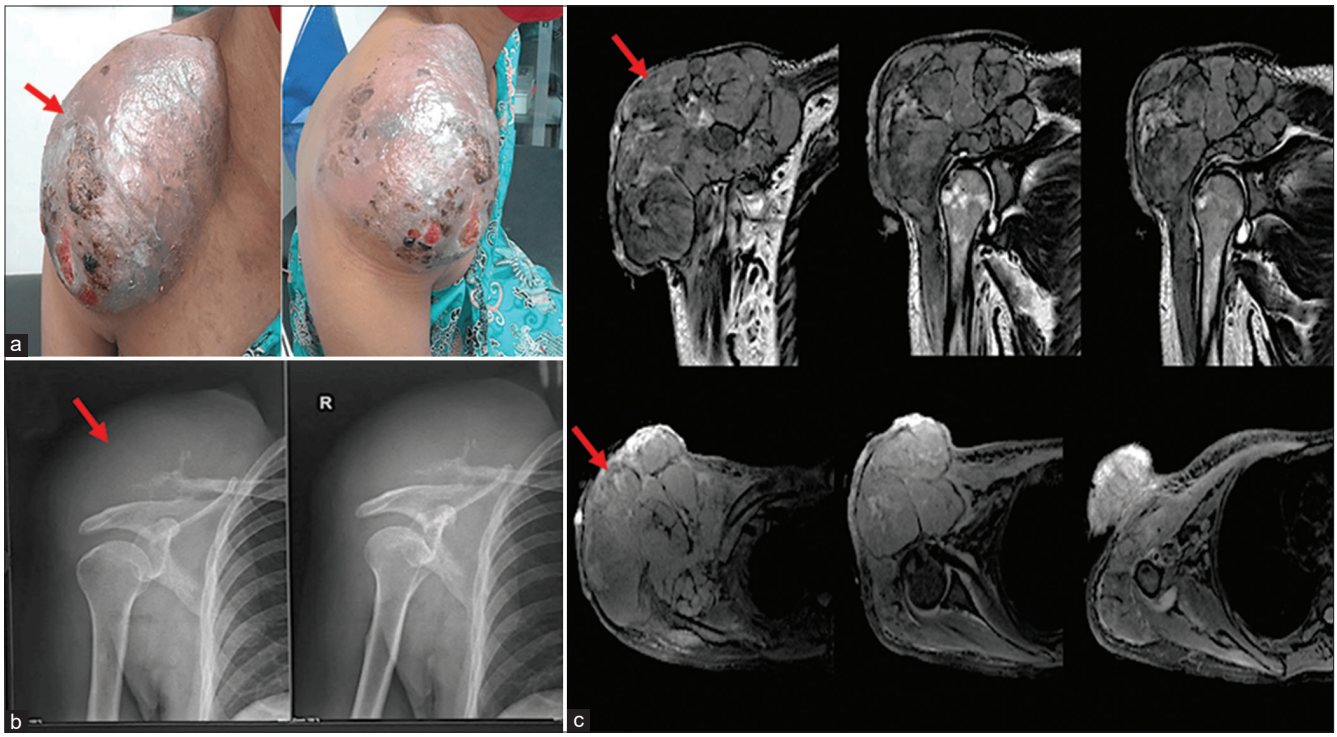

Figure 1: Clinical and radiology features of the patient. (a) Solid, fixed, and ill-defined border mass of the right shoulder, approximately 20 × 15 × 10 cm. (b) Shoulder X-ray revealed soft-tissue mass of the right shoulder region with the destruction of lateral part of the right clavicle. (c) MRI revealed a solid mass that arose from soft tissue of subcutis, infiltrating the trapezius, and proximal part of the deltoid muscles

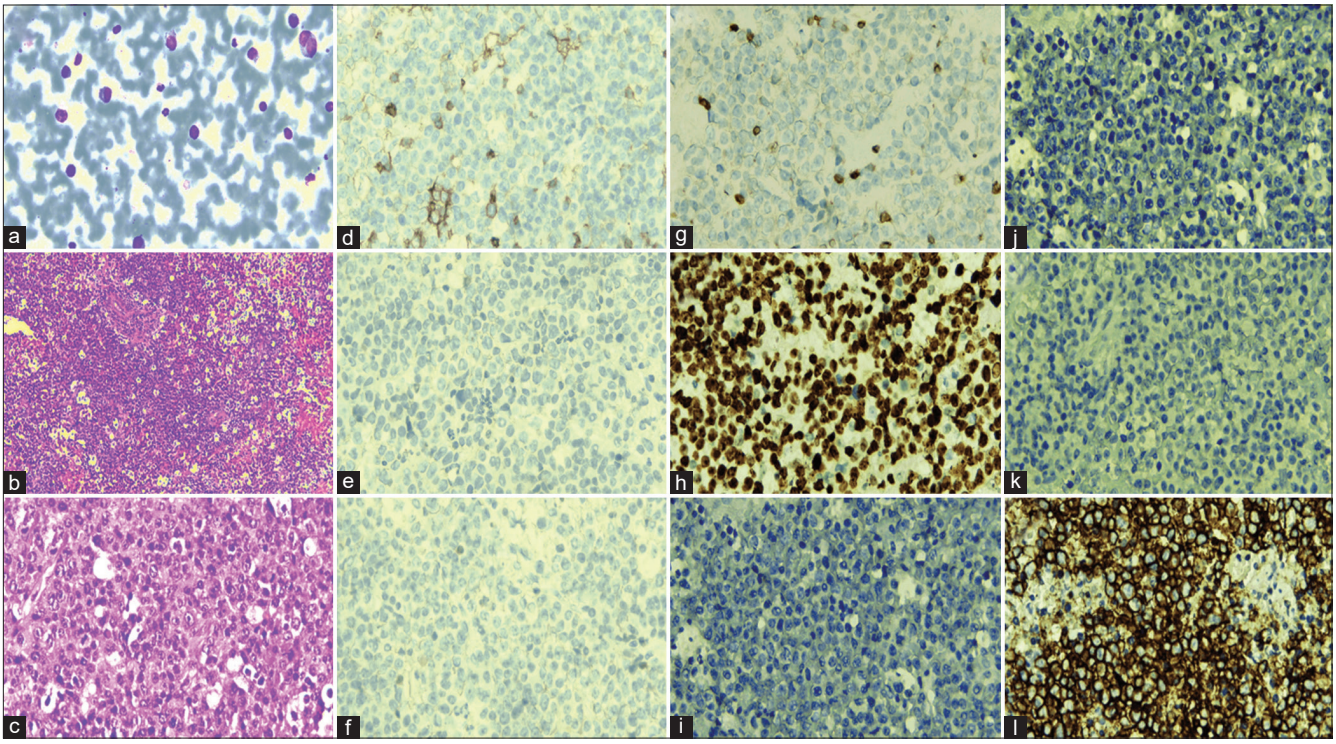

Figure 2: Cytomorphology, histopathology, and immunohistochemical profile. (a) FNAB result showed hypocellular smear of inflammatory cells with scattered plasma cells feature. (b) Histopathology result revealed tumor in diffused growth pattern with wide necrosis, infiltrated by numerous inflammatory cells. (c) Tumor cells were large with round to oval nuclei and prominent nucleoli. Macrophage tangible bodies were scattered, resembling starry sky appearance. IHC examination revealed. (d) CD45 negative. (e) CK negative. (f) CD20 negative. (g) CD3 negative. (h) Ki67 was highly expressed in nuclei. (i) ALK negative. (j) TdT negative. (k) CD79a negative. (l) CD138 was strongly expressed in membrane of tumor cells

daily activity as a farmer with the functional QuickDASH Score of 9.1. Biochemical parameters evaluation such as renal function test and calcium levels remained normal throughout the course of the treatment and follow-up.

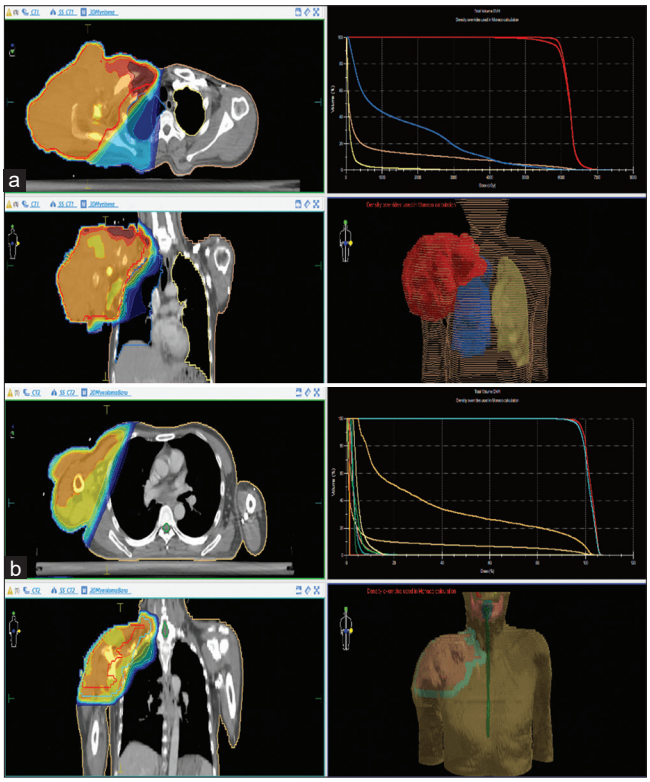

Figure 3: Radiotherapy planning with 3DCRT scheme; (a) first planning 60Gy/30 fractions, (b) adaptive planning implemented after 15 fractions

Discussion

Solitary extramedullary (soft tissue) plasmacytomas (SEP) are less common than Solitary Bone Plasmacytoma (SBP), yet it has a better prognosis since the majority can be cured by local radiotherapy [5]. Although SEPs can arise throughout the body, however almost 90% of them arise in the head and neck area, especially in the upper respiratory tract including the nasal cavity, sinuses, oropharynx, salivary glands and larynx [6], [7]. Approximately 10% of extramedullary plasmacytomas occur in the gastrointestinal tract [8]; there are also reports about pancreatic and liver involvement [9]. A variety of other sites that can rarely be involved include testis, bladder, urethra, breast, ovary, lung, pleura, thyroid, orbit, brain, skin, adrenal glands, retroperitoneum, central nervous system, spleen, and the lymph nodes [10], [11], [12], [13]. In this case, the mass was in shoulder region, which is quite rare.

A monoclonal paraprotein can be detected in the serum and/or urine in fewer than 25% of patients. Local recurrence rates of < 5% have been quoted after

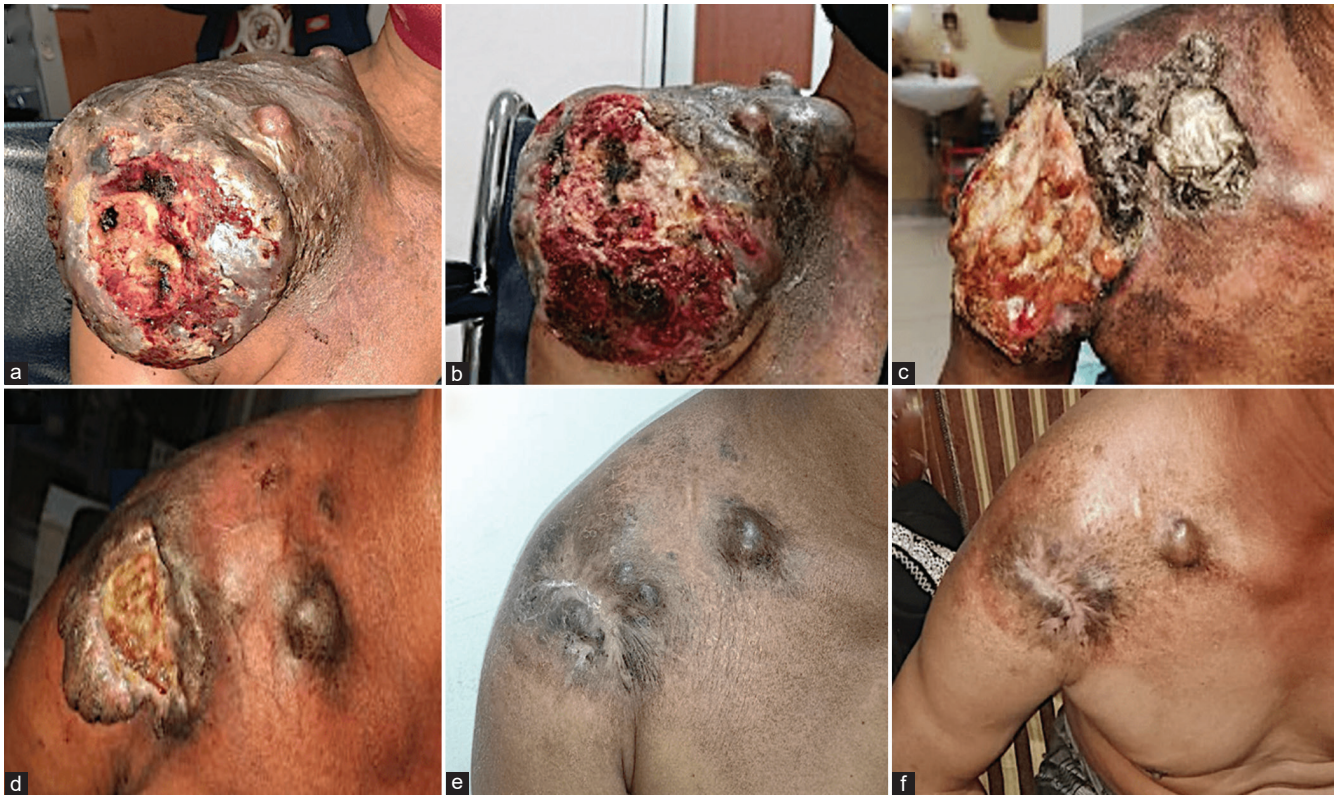

Figure 4: The follow-up figures of the right shoulder mass after undergoing radiotherapy at: (a) 1 month; (b) 2 month; (c) 4 month; (d) 6 month; (e) 10 month; and (f) 1 year, that showed remarkable response of therapy

radiotherapy [6]. The risk of distant relapse appears to be < 30%, that is, significantly less than with SBP. At least two-thirds of patients survive for > 10 years [7].

SEPs are highly radiosensitive tumors so that surgical resection is usually not required. Local control rates of 80–100% are consistently reported with moderate doses of radiotherapy [6], [14]. Radiotherapy alone is the treatment of choice for head and neck SEP, and radical surgery should be avoided. For SEP at other sites, complete surgical removal should be considered if feasible. Patients with involved surgical margins should receive adjuvant radiotherapy. No recommendation for adjuvant radiotherapy can be made for patients who have undergone complete surgical excision with negative margins [15]. For SBP < 5 cm in size, a dose of 40 Gy, in 1.8 to 2 Gy fractions is recommended by NCCN. For SBP greater than 5 cm in size, a dose of 40−50 Gy is recommended. A dose of 40−50 Gy is suggested for SEP as well [16].

Conclusions

We report a case of SEP of the shoulder region that was established by clinical features, histopathology and panel of IHC examination, radiology, and also laboratory examination. In this case report, radiotherapy was shown to be a highly effective modality to treat the patient with SEP.

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