

# Cutaneous involvement by multiple myeloma presenting as erythematous indurated plaques at the site of cardiac pacemaker insertion



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**Key words:** clinicopathology; cutaneous; extramedullary plasmacytoma; multiple myeloma; pacemaker; plaque; plasmacytoma.

## INTRODUCTION

Cutaneous involvement by multiple myeloma (MM) is a rare extramedullary manifestation consisting of a proliferation of malignant plasma cells in the skin. This has sometimes been termed “secondary cutaneous plasmacytoma.” It most commonly affects the trunk and extremities; however, there have been rare cases involving surgical sites and sites of trauma. We describe an interesting case of cutaneous involvement by MM at the site of pacemaker insertion. To our knowledge, only 3 cases of cutaneous plasmacytoma arising at a pacemaker site have been published. Recognition of this rare manifestation is important as it has implications for treatment planning and portends a poor patient prognosis.

## CASE REPORT

A woman in her 70s presented with a 1-month history of erythematous plaques over central portion of her chest and at a recent surgical site. The patient reported associated fevers and night sweats despite a recent course of intravenous and oral antibiotics. The patient had a known background of kappa IgG MM, which had been diagnosed 2 years earlier, and cardiogenic syncope requiring a permanent pacemaker (PPM), which had been inserted 5 months ago. Physical examination revealed 2 indurated, ill-defined, erythematous-to-violaceous plaques (Fig 1). The lesions were nontender with no overlying erosion or ulceration. No other lesions were

### Abbreviations used:

MM: multiple myeloma  
PPM: permanent pacemaker

noted on the skin or mucous membranes, and no palpable lymphadenopathy was noted. An incisional biopsy was performed for histopathology and tissue culture (Fig 2).

Histopathologic examination revealed medium-to-large malignant plasmacytoid cells with enlarged nuclei, prominent nucleoli, and moderate amounts of eosinophilic-to-basophilic eccentric cytoplasm. The dominant population exhibited a plasmablastic morphology with smaller numbers of admixed mature plasma cells. The cells were CD138<sup>+</sup>, CD56<sup>+</sup>, CD20<sup>-</sup>, CD79a<sup>-</sup>, and kappa light chain-restricted on immunophenotyping. Tissue cultures for bacterial, fungal, and atypical organisms were negative. A diagnosis of cutaneous involvement by the patient's known MM was made.

Further history taking revealed that the recent surgical site was the result of a pacemaker removal procedure. The initial treating team interpreted the systemic symptoms, raised inflammatory markers, and erythematous plaques overlying the previous PPM as medical device infection, and the patient had undergone device removal and new pacemaker insertion as well as a prolonged course of oral antibiotics.

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Funding sources: None.

Abstract accepted for presentation at the 53rd Australasian College of Dermatologists' Annual Scientific Meeting (virtual), Adelaide, SA, Australia, April 9-11, 2021.

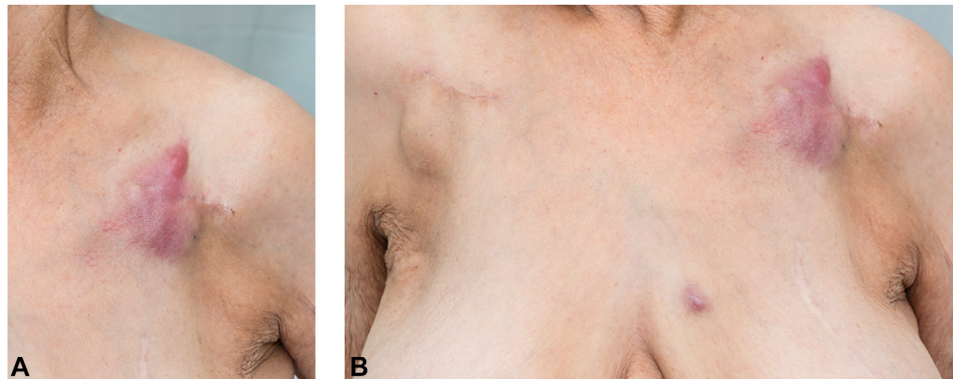
IRB approval status: Not applicable.

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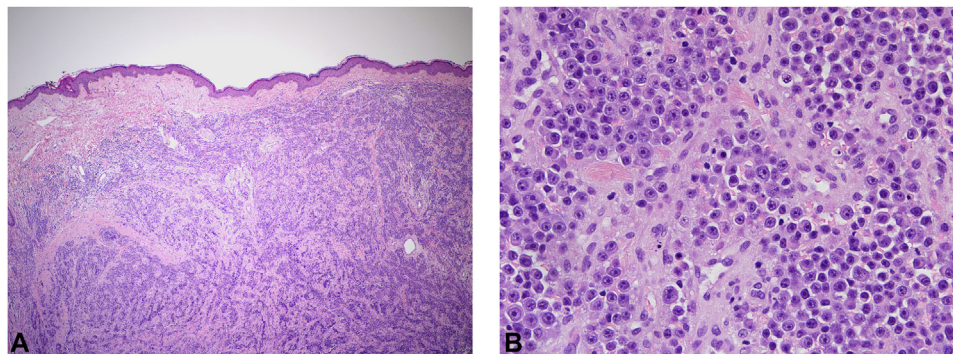
JAAD Case Reports 2021;12:54-7.  
2352-5126

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<https://doi.org/10.1016/j.jidcr.2021.04.013>



**Fig 1.** **A**, Clinical image of a 50 × 70-mm erythematous indurated plaque over the site of the previous pacemaker insertion. **B**, Also, shown is the replacement pacemaker on the contralateral aspect of the chest wall and a 12 × 10-mm erythematous nodule of the left breast.



**Fig 2.** Cellular infiltration of the dermis with medium-to-large plasmacytoid cells with enlarged nuclei, prominent nucleoli, and eccentric cytoplasm. (**A** and **B**, Hematoxylin-eosin stain; original magnifications: **A**, ×40; **B**, ×400.)

Following the diagnosis of cutaneous involvement, a bone marrow biopsy was performed that demonstrated the progression of the patient's MM. Treatment was escalated to the CyBORd regimen (cyclophosphamide, bortezomib, and dexamethasone). Unfortunately, after 2 treatment cycles, serial positron emission tomography imaging demonstrated rapid progression of the disease with extensive metastases. The patient clinically deteriorated over the next 2 months, with the persistence of the cutaneous lesions, and ultimately succumbed to her illness. The patient passed away 4 months after the emergence of the cutaneous lesions.

## DISCUSSION

Cutaneous involvement by MM is a rare extramedullary manifestation occurring in approximately 1% of the patients.<sup>1</sup> It presents as nodules or plaques, which can be erythematous, violaceous, or skin-colored. In most cases, the condition occurs in patients with known MM and is associated with

relapse or progression of the disease. Less commonly, it can be the presenting feature of MM. Cutaneous involvement by MM, as with other extramedullary involvement, portends a poor prognosis with significantly reduced overall survival and progression-free survival.<sup>1,2</sup> The median survival from the time of cutaneous plasmacytoma occurrence is 8-9 months.<sup>1,2</sup>

Common sites involved are the trunk and extremities; however, rare cases involving surgical sites such as pacemaker insertion or fracture sites have been described in the literature.<sup>3-5</sup> The mechanism for these occurrences at sites of trauma is not well understood, though it may potentially involve local inflammatory mediators released following surgery, which contribute to the clonal expansion of a plasma cell population.<sup>3</sup>

In 1976, Hamaker et al<sup>5</sup> reported the first case of cutaneous plasmacytoma in a pacemaker pocket, which occurred 16 months after insertion (Table D). Plasmacytoma was the presenting feature of MM in

**Table I.** A summary of case reports of cutaneous plasmacytoma involving pacemaker insertion sites

Author	Demographics	Presenting feature	Known MM at time of diagnosis	Prognosis
Hamaker et al <sup>5</sup> (1976)	48-year-old man	Violaceous lesion over pacemaker site and pearly gray subcutaneous tissue, which occurred 16 months after insertion.	No. MM was diagnosed at the time of presentation of plasmacytoma.	Rapidly growing lesions treated with radiotherapy and surgery. Death within 3 months due to progressive disease.
Sasaki et al <sup>6</sup> (1992)	74-year-old woman	Mass overlying pacemaker that occurred 6 months after insertion.	Yes, diagnosed 18 months earlier.	Initial response to chemotherapy, then re-enlargement was observed several weeks later. Subsequent surgical treatment and death within 10 months due to aggressive disease and heart failure.
Li et al <sup>3</sup> (2013)	89-year-old man	Nontender erythematous plaque that occurred 12 months after insertion.	No. MM was diagnosed 4 months after plasmacytoma presentation.	Initially, complete remission with surgery and radiotherapy had been achieved. Two months later, recurrence at the site of new pacemaker insertion and progression to MM. Received systemic therapy; however, died due to progressive disease (time not specified).
Choong et al (present study)	76-year-old woman	Erythematous indurated plaque at pacemaker site that occurred 5 months after insertion.	Yes, diagnosed 2 years earlier.	Escalation of systemic therapy without change in the lesion. Death due to progressive disease within 4 months of the emergence of cutaneous lesions.

MM, Multiple myeloma.

this patient and manifested as a violaceous mass overlying the PPM site.<sup>5</sup> The patient's disease rapidly progressed following the emergence of the plasmacytoma, and the patient died 3 months later. Similarly, in 2013, Li et al<sup>3</sup> described a case of plasmacytoma occurring in a patient without known MM (Table D). Interestingly, despite initial response to surgery, device replacement, and radiotherapy, the cutaneous plasmacytoma recurred at the site of the new replacement pacemaker 2 months later, and a diagnosis of MM was established.<sup>3</sup> As in our case, Sasaki et al,<sup>6</sup> in 1992, reported a case of plasmacytoma presenting as a lesion

overlying the PPM site in a patient with known MM (Table 1). Their patient passed away 10 months after the emergence of the plasmacytoma.

Other cutaneous plasma cell and B-cell neoplasms can also present as erythematous plaques or nodules and are differential diagnoses for cutaneous involvement by MM. Extranodal marginal zone B-cell lymphoma is a rare non-Hodgkin lymphoma that primarily affects the skin. It can present with solitary or multifocal red to brown plaques or nodules distributed over the trunk and arms. Biopsy demonstrates a lymphocytic infiltrate in dermal or superficial subcutaneous tissue with marginal zone cells and a characteristic immunophenotype with BCL-2 positivity and BCL-6 negativity. Cutaneous plasma cell granuloma is a rare benign inflammatory tumor that often presents with an asymptomatic erythematous-to-hyperpigmented solitary dermal nodule. Biopsy demonstrates a well-circumscribed, nonencapsulated dermal infiltrate of polyclonal plasma cells, which may extend to the subcutaneous tissue. It is often associated with a prominent sclerosing collagenous stroma.

Other differential diagnoses for cutaneous involvement by MM include primary cutaneous plasmacytoma, plasmablastic lymphoma, and lymphoplasmacytic lymphoma. The correlation of clinical and histopathologic features helps distinguish these; however, diagnosis can be challenging, as cutaneous MM involvement has varied morphologic appearances, including plasmacytic, plasmablastic, and lymphoplasmacytic variants.<sup>2,7</sup> Plasmablastic features were seen in 40% of cases in a series by Panse et al,<sup>7</sup> in 2021, as was seen in our patient. Interestingly, a case of plasmablastic lymphoma occurring in a pacemaker pocket has been described.<sup>8</sup>

This case highlights the importance of considering the rare manifestation of cutaneous involvement by MM and the possible predilection it has for surgical

sites or sites of trauma. In our patient, earlier recognition may have avoided unnecessary pacemaker replacement operation and multiple courses of antibiotics. This disease manifestation carries a poor prognosis and may prompt discussions regarding end-of-life care planning with these patients.

We thank the late patient for granting permission to publish these images. We also thank Jean Iacobelli, FRCPA and Nathan Harvey, FRCPA from PathWest (Nedlands, Western Australia) for their expertise in interpreting and producing the histopathology images.

#### Conflicts of interest

None disclosed.

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