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Progressive mantle cell lymphoma presenting with intractable pruritus

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To the Editor:

Mantle cell lymphoma is a rare type of non-Hodgkin lymphoma that most commonly presents with either lymphadenopathy or extra-nodal disease [1]. Skin manifestations of mantle cell lymphoma are extremely rare [2]. Here we report a case of progressive mantle cell lymphoma presenting with intractable pruritus and complete resolution after chemotherapy.

A man in his 60s presented with severe and progressive generalized pruritus refractory to multiple courses of systemic corticosteroids. Three years prior, he was diagnosed with cutaneous marginal zone B cell lymphoma of mucosa-associated lymphoid tissue of the right upper arm, that was treated with radiation. Staging CT scans demonstrated multiple enlarged abdominal and pelvic lymph nodes diagnosed as concomitant mantle cell lymphoma. Treatment of his mantle cell lymphoma was deferred owing to its asymptomatic nature and absence of end-organ damage from compression. The patient was followed with serial imaging and laboratory studies. He also presented with a mild rash characterized by erythematous papules and macules, but this was easily treated and quickly resolved with systemic corticosteroids. However, the pruritus persisted and increased in intensity over time. Phototherapy with narrowband UVB was tried without relief. Mycophenolate mofetil initially helped but ultimately led to an erosive dermatitis and was discontinued. Azathioprine was considered but not favored given his history of indolent lymphoma.

Follow-up staging PET/CT demonstrated significant progression of the patient's mantle cell lymphoma with a significant increase in the size of his enlarged abdominal and pelvic lymph nodes. At this time, the patient also developed fatigue, myalgias, and increasing inguinal lymphadenopathy. Given his bulky stage IIIA disease, the patient started chemotherapy with rituximab, dexamethasone, cytarabine, and oxaliplatin (R-DHAX). The patient's pruritus abated completely after one cycle of chemotherapy. He has since completed his fourth cycle of R-DHAX with a significant decrease in his lymphadenopathy on repeat CT. He was enrolled in a study investigating allogeneic stem cell transplantation in the treatment of mantle cell lymphoma.

Malignancy-associated pruritus has been reported in several hematological malignancies, including Hodgkin lymphoma, non-Hodgkin lymphoma, cutaneous T-cell lymphoma, and leukemia [3]. There are two main mechanisms by which malignancy can cause pruritus: either a local reaction to malignancy or a paraneoplastic itch in response to an internal malignancy [3]. Because our patient's pruritus was generalized, the pathogenesis was more likely paraneoplastic. The estimated prevalence of paraneoplastic itch in non-Hodgkin lymphoma is 15% [3]. By comparison, its prevalence in Hodgkin lymphoma is 25% [3]. Furthermore, intractable pruritus can indicate the progression of Hodgkin lymphoma and precedes the initial diagnosis in some cases [4,5]. The relationship between pruritus and non-Hodgkin lymphomas is poorly described

owing to the heterogeneous nature of this group of lymphomas.

Mantle cell lymphoma is a rare subtype of B-cell non-Hodgkin lymphoma with a heterogeneous clinical presentation and course [1]. Mantle cell lymphoma's most common presentation is symptomatic bulky lymphadenopathy or extra-nodal disease requiring systemic chemotherapy [1]. Mantle cell lymphoma may also follow a more indolent course with some providers electing for observation [1]. Cutaneous involvement by mantle cell lymphoma is extremely rare but has been reported in the literature [6]. Previous case reports of cutaneous manifestations of mantle cell lymphoma report solitary nodules, erythematous papules, plaques, and maculopapular rashes [2,6]. Mantle cell lymphoma presenting as a hypersensitivity reaction to mosquito bites has also been described [7]. The most commonly affected sites include the trunk, face, and upper extremities [6]. In one case report, a patient presented with generalized lymphadenopathy and a maculopapular rash over the extremities and back that progressed to bullae and ulcers [2]. He was later diagnosed with mantle cell lymphoma and his skin lesions resolved after five cycles of chemotherapy [2]. Our case differs in that our patient did not develop bullae or ulcerative lesions and the primary symptom was intense pruritus.

Chronic pruritus is associated with an increased risk of undiagnosed hematologic and bile duct malignancies; however, the overall risk is very low [8]. Broad screening for malignancy in patients with chronic pruritus is unnecessary [8]. However, it is essential to assess these patients with a thorough history, physical exam, complete blood count, and comprehensive metabolic panel [8].

In sum, our patient presented with generalized pruritus refractory to corticosteroids, intensified as his mantle cell lymphoma progressed, and resolved with chemotherapy. This suggests that his pruritus was related to his lymphoma. Whereas pruritus may be a presenting sign for hematologic malignancies, it should also prompt evaluation for recurrence or progression of otherwise quiescent disease.

Potential conflicts of interest

The authors declare no conflicts of interest.

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