Cutaneous Polyarteritis Nodosa in Newly Diagnosed MGUS: a coincidence or association?

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BACKGROUND

- Polyarteritis Nodosa (PAN) is a rare systemic necrotizing medium-size arteries vasculitis, majorly associated with Hepatitis B infection and rarely with hematological disorders like hairy cell leukemia.
- We present a case of cutaneous PAN in association with monoclonal gammopathy of undetermined significance (MGUS).

CASE

- A 60-year-old male with a history of hypertension, chronic leukocytosis, and anemia (etiology unclear at presentation) presented with altered mental status. He was also noted to have non-healing bilateral lower extremity wounds.
- Vital signs on presentation were blood pressure of 92/58 mm Hg, pulse rate of 112 bpm, respiratory rate of 20, saturating 94% on room air.
- Initial lab evaluation was relevant for creatinine of 2.88 mg/dl, white blood count of 50 k/mcL (from baseline 22.3k/mcL) with differential showing 92 % neutrophils, 4 % lymphocytes and no band forms, and hemoglobin of 9.0 g/dL (baseline 9.4).
- He was treated for possible sepsis secondary to underlying skin wounds with vancomycin and piperacillin-tazobactam. He did not respond to antibiotics and had worsening renal function and leukocytosis.
- An elliptical biopsy of a non-healing left calf lesion showed small cell vasculitis with negative direct immuno-florescence. Further testing with serologies and tissue stain favored a diagnosis of cutaneous polyarteritis nodosa

Figure 1. Bilateral (a) right and (b) left lower extremity necrotic, ulcerating wounds with black-brown eschar in a patient with PAN.

Courtesy: Abousy M, Byrd A, Succaria F, et al. (February 04, 2021) Burn Center Management of Severe Necrotic Cutaneous Polyarteritis Nodosa in a Patient With a History of Thymoma. Cureus 13(2): e13134. doi:10.7759/cureus.13134

DECISION-MAKING

- His antibiotics were stopped because of low concerns for infectious etiology.
- Pulse dose steroids were initiated and an improvement in mental status and vital signs were noted.
- Due to worsening renal function , a renal biopsy was performed which showed acute tubular injury, chronic hypertensive changes with no evidence of glomerulonephritis or vasculitis.
- He was screened for multiple myeloma given his AKI and anemia. SPEP revealed M spike of 0.5 g/dL. Serum immunofixation revealed a free kappa light chain of 10.17 mg/dl, free lambda light chains of 5.94 mg/dl, and kappa/lambda fluid LC ratio of 1.71.
- A subsequent bone marrow biopsy revealed CD20 B cells accounting for 5-10 % of marrow cells. CT head and CT chest, abdomen, and pelvis did not show any lytic bone lesions. These findings were consistent with MGUS. He was then treated with prednisone and rituximab for cutaneous PAN.

CONCLUSION

- Given the association of PAN with hematological disorders, the presence of MGUS in our patient could have a possible relation to the development of cutaneous PAN. This case highlights the importance of considering screening for plasma cell dyscrasias in such patients.

Conflict of Interest: None