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Letter to Editor

C-ANCA Positive Vasculitis in a Case of Amyloidosis

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Dear Editor

Thick waxy skin, easy bruising and subcutaneous nodules, plaques and purpura particularly in the periorbital area are common skin manifestations of systemic amyloidosis. Primary amyloidosis may present with vasculitides, especially giant cell arteritis or polymyalgia rheumatica.[1-5] Rarely, it may coexist with necrotizing vasculitis of the central nervous system, giant cell arteritis, or vasculitis of the small intestine.[1, 6-8]

A 68-years-old man was referred to the hematology clinic with purpuric lesions over the skin. He complained of skin ecchymoses lasting for one year. The skin lesions were not itchy. He also reported numbness of both hands and feet for which he underwent surgery with a diagnosis of bilateral carpal tunnel syndrome. He was taking levothyroxine for hypothyroidism since one year ago.

Physical examination revealed purpuric skin lesions distributed over forearms and periorbital area, macroglossia, pitting edema over both legs and bilateral atrophy of thenar muscles. Due to prolonged skin lesions, a skin biopsy was performed which showed vasculopathic tissue pattern with infiltration of eosinophils and subepidermal blistering.

Laboratory tests showed normal hematological indices. ESR 86 mm, Ca 10.5 mg/dl, evidence of monoclonal gammopathy in serum protein electrophoresis, high IgG titer 2923 mg/dl (normal range: 77-1600) and proteinuria (1550 mg in 24 hour-urine) were observed. Serum B12 level was 183 pg/ml. Electrocardiography showed right bundle branch block. Abdominal ultrasound revealed mild splenomegaly (spleen span=132 mm). EMG and NCV was performed that was in favor of bilateral carpal tunnel syndrome. In order to rule out autoimmune vasculitides, Antineutrophil cytoplasmic antibody (ANCA) (c-ANCA and p-ANCA) was checked which the result was positive with the titer of 1/32 for c-ANCA (negative <1/10). Skull-x-ray of the patient showed multiple lytic lesions.

The patient underwent bone marrow aspiration and biopsy which exhibited more than 70% plasma cells. The above mentioned findings confirmed amyloidosis associated with multiple myeloma.

We found c-ANCA positive vasculitis in this case of multiple myeloma associated with amyloidosis. It could be suggested that clinical and para-clinical findings of plasma cell dyscrasia should be searched in any case of vasculitis.

Salvarni and colleagues reported three cases of AL amyloidosis who were preceded by typical symptoms of giant cell arteritis such as headache, proximal muscle weakness and lower extremity claudication.[1]

A 67-year-old man was reported presenting with a one-year history of weakness, fatigue, headache and jaw claudication. Thickened temporal artery was noticeable in that case that he was first diagnosed as giant cell arteritis; however, later on, the patient developed macroglossia, hard nodular structures of lips, buccal

dryness and livedo reticularis in the skin. The lip biopsy was performed from nodular structures. Hematoxylin and eosin staining of the biopsy showed concentric intimal thickening and deposition of large amorphous, eosinophilic material, suggestive of amyloid deposits in the media of the arteries. Histochemical examination was in favor of amyloid angiopathy. Electrophoresis revealed a monoclonal spike (Immunoglobulin G lambda) and free lambda light chains with a kappa/lambda ratio of 0.24. Bone-marrow aspiration showed 30% atypical plasma cells compatible with multiple myeloma.[4]

There is report of a woman with clinical and radiological signs of a right temporal mass suggestive of a brain tumor. She further found to have granulomatous angiitis associated with cerebral amyloid angiopathy confirmed by biopsy.[7]

Auethavekiat and co-workers reported a previously healthy woman who developed a small bowel perforation. Visceral angiography was strongly suggestive of necrotizing vasculitis in all vascular beds, with segmental arterial strictures and fusiform aneurysms. Pathologic examination of the bowel did not show any evidence of vasculitis; however, significant amyloid deposition was remarkable by Congo red staining. She was diagnosed with primary systemic amyloidosis which was masked by necrotizing vasculitis.[8]

AL amyloidosis should be suspected in any patient with a monoclonal gammopathy, proteinuria, edema, weight loss, orthostatic hypotention and typical symptoms of carpal tunnel syndrome. In addition, features of vasculitis in any patient with negative biopsy results for vasculitis should elicit the possibility of primary systemic amyloidosis.

Conflict of Interest: None declared.

References

1. Salvarani C, Gabriel SE, Gertz MA, Bjornsson J, LI CY, Hunder GG. Primary systemic amyloidosis presenting as giant cell arteritis and polymyalgia rheumatica. Arthritis Rheum. 1994; 37(11):1621–6. doi: 10.1002/art.1780371111.
2. Ing EB, Woolf IZ, Younge BR, Bjornsson J, Leavitt JA. Systemic amyloidosis with temporal artery involvement mimicking temporal vasculitis. Ophthalmic Surg Lasers. 1997; 28(4):328–31. **PubMed PMID:** 9101575.
3. Rao JK, Allen NB. Primary systemic amyloidosis masquerading as giant cell arteritis: case report and review of the literature. Arthritis Rheum. 1993;36(3): 422–5. doi: 10.1002/art.1780360320.
4. Emmungil H, Kalfa M, Başarık B, Kahraman H, Tanhan F, Yaman B, et al. Primary systemic Al amyloidosis presenting as temporal arteritis. Case Rep Rheumatol. 2014; 2014: 549641. doi: 10.1155/2014/549641. PubMed PMID: PMC3914325. PubMed Central PMCID: PMC3914325.
5. Estrada A, Stenzel TT, Burchette JL, Allen NB. Multiple myeloma-associated amyloidosis and giant cell arteritis. Arthritis Rheum. 1998 Jul;41(7):1312-7. doi: 10.1002/1529-0131(199807)41:7<1312::AID-ART23>3.0.CO;2-T. PubMed PMID: 9663490.
6. Oweity T, West AB, Stokes MB. Necrotizing angiitis of the small intestine related to AA-amyloidosis: a novel association. Int J Surg Pathol. 2001;9(2):149–54. doi: /10.1177/106689690100900211.
7. Le Coz P, Mikol J, Ferrand J, Woimant F, Masters C, Beyreuther K, et al. Granulomatous angiitis and cerebral amyloid angiopathy presenting as mass lesion. Neuropathol Appl Neurobiol. 1991; 17(2):149–55. doi: 10.1111/j.1365-2990.1991.tb00706.x.
8. Auethavekiat P, Murali NS, Manek NJ. Clinical images: primary systemic amyloidosis masquerading as necrotizing vasculitis. Arthritis and rheumatism. 2004; 50(10): 3400. doi: 10.1002/art.20560.