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CASE REPORT

Polyarteritis Nodosa: an unusual case of paraneoplastic process in renal cell carcinoma

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ABSTRACT

Polyarteritis Nodosa (PAN) is a small and medium vessel necrotizing vasculitis that can affect any system in the human body. Rarely, PAN can be the primary manifestation of an underlying malignancy. The association between malignancy and vasculitis is an ongoing topic of research. Our patient's presentation suggests malignancy may be a trigger for acute onset vasculitis and therefore once a malignancy is identified, therapy should be targeted towards treating the malignancy rather than the vasculitis alone.

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Polyarteritis Nodosa (PAN); paraneoplastic process; renal cell carcinoma; malignancy; tumor antigens; vasculitis

1. Introduction

Polyarteritis Nodosa (PAN) is a small and medium vessel necrotizing vasculitis that can affect any system in the human body. Typical presentations of the panarteritis include skin changes, GI system involvement, polyneuropathies, and acute renal failure as a result of an autoimmune process. Rarely, PAN can be the primary manifestation of an underlying malignancy, likely from a paraneoplastic process. The association between malignancy and vasculitis is an ongoing topic of research. We report one such case of a patient with biopsy-proven PAN resulting in loss of lower extremities, testicles, and upper extremity digits, in the setting of Renal Cell Carcinoma (RCC).

2. Case report

Fifty-four-year Caucasian male with past medical history significant for hypertension, alcohol abuse and tobacco abuse presented to an outside facility with bilateral upper and lower extremity pain and right lower quadrant abdominal pain. He described the pain in extremities as pins and needles sensation. Walking and exposure to cold exacerbated the pain in the lower extremities, and rest and warmth relieved it. The extremity pain was initially believed to be frostbite as patient had cold exposure for multiple hours with improper footgear prior to admission. A CT abdomen was performed for the sharp right lower quadrant pain, and appendicitis was noted. CT abdomen incidentally also noted a right renal lesion located in the lateral aspect measuring 2.6 cm x 2.7 cm x 2 cm. He underwent an appendectomy and was transferred to our hospital. On physical exam, the upper extremities had peripheral cyanosis, right upper extremity digits were extremely tender to touch and patient was unable to flex the right fifth digit at proximal or Manuscript – anonymous distal interphalangeal joints. He had 2+ bilateral lower extremity pitting edema and skin manifestations consistent with livedo reticularis. There was partial loss of light touch sensation up to his ankles bilaterally, as well as some loss of motor function of toes bilaterally. At this time, possibilities of presenting symptoms for upper and lower extremities included cold exposure leading to frostbite, thromboembolic phenomena, non-infectious endocarditis, vasculitic disease, and infectious etiologies. On labs, the patient was noted to have leukocytosis at 26 K with a neutrophilic predominance at 89.1%. ESR and CRP were both elevated at 90 and 31.1, respectively. ANA was negative. Test for hepatitis-B virus, hepatitis-C virus and human immunodeficiency virus were negative. ABIs of lower extremities were noted to be normal. Ultrasound arterial doppler of upper extremities showed severe ischemia at the digits bilaterally: Right radial brachial index of 1.08, waveforms were unobtainable in the digits with a finger brachial index of 0, indicating severe ischemia. Left radial brachial index of 1.07, waveforms were unobtainable in the digits with a finger brachial index of 0, indicating severe ischemia. During the hospital-stay, patient also developed severe testicular pain for which an ultrasound was performed. Ultrasound showed bilateral testicular infarction. At this time, the pathology from the appendix returned from the outside facility and was notable for vasculitic changes. Given overall presentation, the patient met American College of Rheumatology criteria for Polyarteritis Nodosa with the following features: livedo reticularis of the lower extremities, polyneuropathy, vasculitis of the appendix

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on surgical pathology and bilateral testicular infarction as seen on the ultrasound. The patient was started on a regimen that included high-dose steroids of Solu-Medrol 125 q.6 hours IV, therapeutic anticoagulation with heparin, and cyclophosphamide. The patient reported improvement of pain in extremities and testicular pain within 2 days of starting the above medications. The patient was to be discharged on prednisone 80 mg daily, bactrim doublestrength tablets daily for PJP prophylaxis and alendronate 70 mg weekly for osteopenia. In regards to the right renal mass, pathology resulted as clear cell carcinoma and patient subsequently underwent cryoablation of the mass.

3. Discussion

Polyarteritis Nodosa (PAN) is a medium and small vessel-necrotizing arteritis that can affect any organ system, although the lungs are usually spared. It predominantly affects middle-aged men and can have varied presentations, such as myalgias, arthralgias, neuropathy, orchitis, skin changes and symptoms of ischemia, due to its propensity to involve multiple organs. PAN has been shown to be associated with hepatitis B in 30–70% of the cases [1]. Rarely, PAN can be a manifestation of a paraneoplastic syndrome, or malignancy can be an underlying triggering factor for the disease. The structural similarity between tumor antigens and blood vessel antigens, as well as the formation of immune complexes with tumor antigens, are postulated mechanisms for this association [2]. A review of the literature revealed that PAN has been associated with multiple myeloma, non-hodgkin's lymphoma, hairy-cell leukemia as well as solid tumors like gastric cancer and lung angiomatoid fibrous histiocytoma [3]. Although RCC presenting primarily as Polyarteritis Nodosa has not been reported in the past, it has been associated with a wide range of paraneoplastic syndromes, including vasculitis. A diagnosis of PAN can be suspected based on the clinical presentation but histologic or radiologic evidence is necessary to confirm the diagnosis whenever possible, especially due to potential side effects of treatment. The American College of Rheumatology established 10 criteria for the classification of PAN in a patient suspected of vasculitis, and at least three or more criteria are needed to make a diagnosis. These criteria have a sensitivity of 82.2% and a specificity of 86.6% [4]. A French study has shown that these may not always be applicable and hence pathologic evidence is helpful in confirming the diagnosis[4]. A limited number of case reports involving vasculitis secondary to RCC are reported in the literature. Of the few case reports available, most describe a form of vasculitis prior to the identification of RCC [5]. Our patient presented with an unusual case of Polyarteritis Nodosa manifesting as multi-organ infarction. He had livedo reticularis of the lower extremities, ischemia of the upper and lower extremity digits, infarction of the testes and appendix as well as polyneuropathy and a biopsy-proven vasculitis to confirm PAN. Eventually, clear cell carcinoma was diagnosed on biopsy of an incidental right renal mass found on the CT abdomen of the patient. Although PAN can be an idiopathic diagnosis, the lack of previous symptoms and the almost fatal presentation with multi-organ infarction led to the suspicion of an underlying trigger, which was confirmed by the diagnosis of RCC. In the case reports authored by Hoeg, RCC was identified post-mortem in two patients who had died from complications of vasculitis [6]. These case reports, along with our patient's presentation, suggests malignancy may be a trigger for acute onset vasculitis and therefore once a malignancy is identified, therapy should be targeted towards treating the malignancy rather than the vasculitis alone. Our patient did respond to immunosuppressive therapy; however, it was not until cryoablation of RCC that his PAN symptoms began to fully resolve. Conclusion This unusual case adds to the previous cases reported discussing the association of renal cell carcinoma with vasculitis and its various presentations. Early detection and timely subsequent eradication of malignancy may result in reducing the amount of immunosuppressive therapy used to treat the vasculitis. This brings up questions regarding paraneoplastic syndromes and cross reacting antigens as possible underlying mechanisms and research into looking at clear cell receptors and previously known antigens triggering PAN, such as HBV, may be worthwhile. Further case studies and retrospective analyses may help in identifying if such an association is more common than expected.

Disclosure statement

No potential conflict of interest was reported by the authors.

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