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A RARE CASE OF PARANEOPLASTIC POLYMYOSITIS

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Abstract
The association of inflammatory myopathies such as dermatomyositis and polymyositis with malignancies has been well described in literature. The present case report discusses a rare presentation of polymyositis as a paraneoplastic manifestation of an unknown malignancy in an elderly patient. The study underscores the need to conduct careful clinical investigation for the prompt diagnosis and treatment of a paraneoplastic syndrome, thereby to reduce the associated morbidities and mortality.

Introduction

Polymyositis (PM) is an idiopathic inflammatory myopathy characterized by proximal muscle weakness and inflammation, and increased levels of skeletal muscle enzymes. It is one of the common paraneoplastic manifestations, and elderly patients with PM may have a more severe disease course than their autoimmune counterparts.[1]

Case Report

A 60-year-old male patient presented to the clinic with sudden onset of limbs weakness and he was wheelchair bound. He had cough with expectoration, which was not responding to antibiotics. His systemic examination was normal and no lymphadenopathy or organomegaly was reported. His clinical investigation revealed: Creatine phosphokinase (CPK)-12000 IU/L, serum glutamic oxaloacetic transaminase (SGOT)-122 IU/L, serum glutamic pyruvic transaminase (SGPT)-222 IU/L, creatinine-2.2 mg/L, C-reactive protein (CRP)- 34 mg/L. Complete blood count and levels of complements 3 and 4 were within the normal range. He was negative for anti-neutrophil cytoplasmic antibody (ANCA) and anti-nuclear antibody (ANA). Ultrasonography of abdomen showed small contracted kidney on the right side and left normal sized kidney Unfortunately, he had an insect bite over the right calf and developed cellulitis, which spread rapidly to the thigh. He was put on high dose amoxicillin clavulanate 1.2 gm once daily for 3 days. However, due to the minimal response, the antibiotic was changed to pipercillintazobactum (4.5 gm once daily). He developed severe pancytopenia with high grade fever on the same day of antibiotic substitution and started desaturating. His chest X-ray showed pnemonitic patch on the right side and small one on the left (Fig.1 and 2). with scarring, and electromyogram revealed inflammatory myopathic potentials. A provisional diagnosis of idiopathic inflammatory myositis with chronic kidney disease was made and was started on pulse methyl prednisolone (1 gm for 5 days). The patient subsequently received prednisolone (60mg/week) and methotrexate (10mg/week) as steroid sparing medication. Rapid improvement in symptoms was noted and he was able to walk independently within 4 weeks.

Bone marrow aspiration showed hypoplasia and the administration of granulocyte-macrophage colony-stimulating factor (GMCSF) was started. His counts improved within 2 days of GMCSF treatment, but the fever persisted. The patient was diagnosed with drug-induced bone marrow suppression and the antibiotics were substituted with meropenam, ornidazole, and fluconazole. The results of patient's blood/sputum culture and gram stain were negative. Since the patient was rapidly desaturating and his weakness increased, imummunoglobulin therapy (400mg/kg body weight) was initiated. The patient improved rapidly and the oxygen saturation attained normal levels. Since his chest lesions persisted on repeated chest X-ray, a CT scan was performed. The imaging showed bilateral ground glass opacities with homogenous coin lesions suggestive of metastasis (Fig.3 and 4). The primary source of metastasis could not be found, but the probable source was speculated to be prostate or testis. Before referring the patient to an

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oncologist, he was succumbed to death due to sudden desaturation and a massive cardiac arrest, which could not be resuscitated.

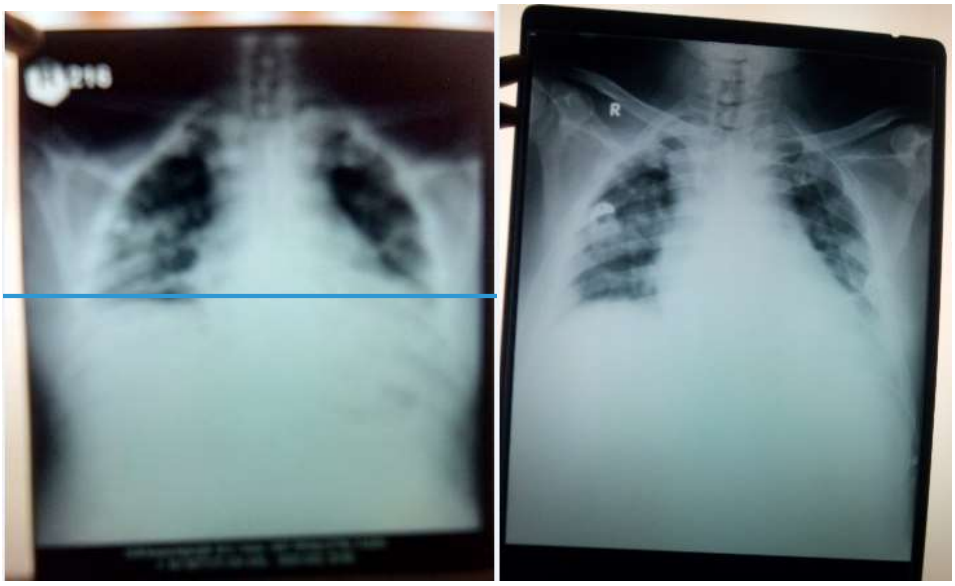

Fig 1 and 2: chest X-ray showing pnemonitic patch on the right side and small one on the left

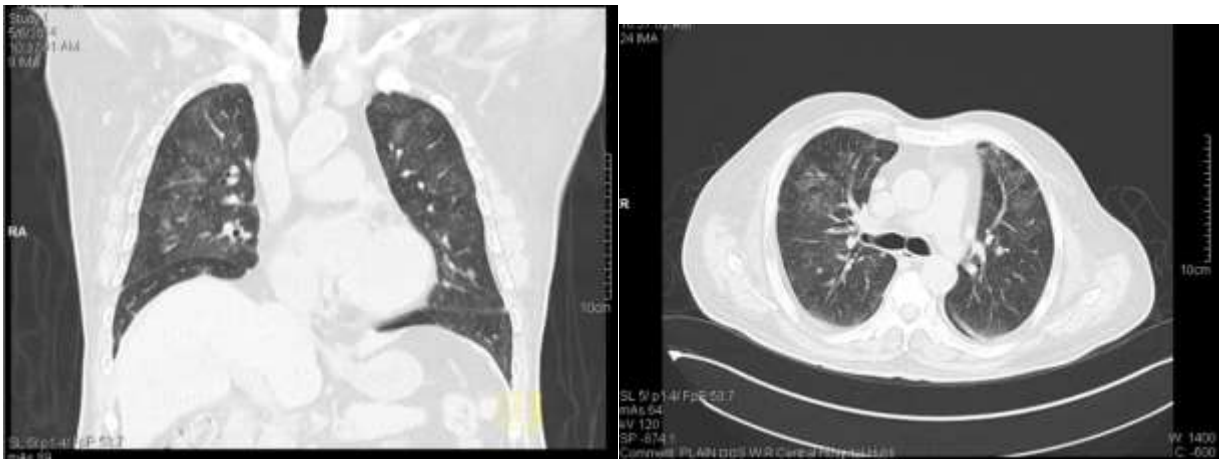

Fig. 3 and 4: CT scan showing bilateral ground glass opacities with homogenous coin lesions, suggestive of metastasis

Discussion

Paraneoplastic syndrome (PNS) refers to signs/symptoms that occur remotely from a malignant neoplasm, caused by the altered immune response to the malignancy.[2] There are very few studies dealing with the prevalence and incidence of PNS in the overall cancer population.[1]

Gabrilovich et al. has highlighted the significance of considering refractory PM as a possible PNS in patients at risk for lung cancer. The study has reported a presentation of biopsy-proven PM of 3 months duration in an elderly patient with squamous cell carcinoma of the lung, confined to a single hilar lymph node.[3] Similarly, Schuster et al. have reported a case of paraneoplastic PM associated with mesothelioma. The case study also underscored the importance of focused diagnostic evaluation for the prompt recognition of PNS.[4]

A population-based cohort study involving 537 subjects has concluded that the risk for malignancy is high in biopsy-proven dermatomyositis and PM.[5] A literature review by Ungprasert and co-authors have reported that age-appropriate cancer screening along with a careful history and physical examination can be performed in patients with idiopathic inflammatory myopathies.[6]

In regular clinical practice, it has been noted that the elderly patients respond very minimally to steroid treatment for PM. In contrast, the current patient had demonstrated a good response to steroids. But his condition had been deteriorated after the administration of pipercillin tazobactum. After conducting several diagnostic tests, CT scan had given a clue to the metastasis. However, the patient had succumbed to death before initiating further work-up to diagnose the malignancy.

The present case underscores the need for considering PM as a potential presentation of PNS, especially in elderly patients. The diagnostic evaluation in such patients should be focused on high degree of suspicion and conducting through clinical examination related to respective tumor entities for the prompt diagnosis of the malignancy. Prompt recognition and treatment of PNS may help in the diagnosis of neoplasia, thereby to reduce the associated morbidities and mortality.

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