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Opinion

Granulomatosis With Polyangiitis: A Brief Review on Some Important Clinical Notes

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Abstract

Granulomatosis with polyangiitis or GPA is a rare disease which causes inflammation in the blood vessels. This brief review tries to point to some important clinical notes about this pathology which having knowledge about them, is of importance in approaching the relevant patients with more precision at the bedside.

Keywords: granulomatosis; gpa; polyangiitis; wegener; clinical notes

Introduction

Granulomatosis with polyangiitis or GPA which is formerly known as Wegener granulomatosis is an uncommon disease which affects the blood vessels by causing inflammation in them. Granulomatosis with polyangiitis in many cases has association with manifestations in the neurological system including multifocal neuropathy and ophthalmoplegia. Its peak incidence is at fifty to sixty years of age and its prevalence in males is somehow higher than in females. Typically, it affects the airways and the kidneys. Also, the central nervous system, the eyes, the joints and the skin can be affected. The thyroid, the breast, the heart, the gastrointestinal system, the liver and the parotid gland can be affected by this disease either although, its occurrence in these organs are less common.

Sinus pain, ulcers of the nose or the oral cavity, hoarseness, rhinorrhea, hemoptysis, hearing loss, nasal discharge, earache, dyspnea, cough, myalgia and polyarthralgia may be seen in the granulomatosis with polyangiitis. Also, the affected patients may complain of having malaise, fever, anorexia, night sweats and weight loss. Peripheral neuropathy, ophthalmoplegia and cranial nerves involvement can also be seen in the patients with granulomatosis with polyangiitis. Radiography of the chest may show opacities and nodules either.

In the affected patient's serum, antibodies against the myeloperoxidase and proteinase 3 or PR3 can be found. Finding of such antibodies in the serum of the affected patients is of importance in the diagnosis of the granulomatosis with polyangiitis. Such antineutrophil cytoplasmic antibodies are positive in about ninety percent of the patients whom are in the generalized and active phase of the disease. A biopsy of tissue is a precise method to diagnose the granulomatosis with polyangiitis. A normocytic and normochromic anemia, thrombocytosis, leukocytosis and an elevated rate of the erythrocyte sedimentation can also be seen in the affected patients with granulomatosis with polyangiitis.

Initial severity of the disease defines the mortality rates. In the patients with severe lung hemorrhage or renal failure, the mortality rate would be around twenty five percent. This rate would be lower at first in the patients with generalized and not life threatening antineutrophilic cytoplasmic antibody associated systemic vasculitis. Leflunomide, methotrexate or mycophenolate mofetil may be used for treatment of the patients with mild forms of the antineutrophilic cytoplasmic antibody associated vasculitis. Rituximab or cyclophosphamide in addition to glucocorticoids which would be continued by steroids with lower doses and in addition to methotrexate or azathioprine, may be used for treatment of the patients with severe forms of the disease. In the short term and for the patients with very severe forms of the disease, the plasmapheresis may also be used.

Conclusion

It is important for the clinicians to have knowledge about the granulomatosis with polyangiitis as an antibody mediated autoimmune granulomatous vasculitis disease. Paying enough attention to details is of importance to approach the affected patients with more precision at the bedside.

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