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Rapid Progress in Diagnosis of ANCA-associated Vasculitis

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Abstract

Diagnosis of ANCA associated vasculitides (AAVs) represents a big burden in the medical field because of clinical features diversity. Wegener's granulomatosis or granulomatosis with polyangiitis (GPA), Churg-Strauss syndrome or eosinophilic granulomatosis with polyangiitis (EGPA), and microscopic polyangiitis (MPA) are the main small-vessel vasculitis. GPA presents in the head and neck, and the Sinonasal cavity is among the most common areas affected. EPGA is occurring within patients with a history of asthma, whereas MPA is known by poor immune complexes markers in histopathology section. In addition, kidney and lung commonly involved in patients with MPA, this may sequentially involve the nervous and musculoskeletal systems. Moreover, skin, heart, eyes, and intestine can be involved. Due to of those varieties and overlapping in signs and symptoms the proper diagnosis is still an urgent need in AAVs management. Although detection of ANCA autoantibodies and histopathology reporting still essential tools for AAVs investigation, it has been reported that they are not enough for definitive diagnosis.

Keywords: Diagnosis; Anti-neutrophil cytoplasmic autoantibody (ANCA); ANCA-Associated Vasculitis (AAVs); granulomatosis with polyangiitis (GPA); Eosinophilic granulomatosis with polyangiitis (EGPA); microscopic polyangiitis (MPA)

Introduction

Anti-neutrophilic cytoplasmic antibodies (ANCA) associated vasculitis (AAVs) is a part of chronic autoimmune inflammatory disorders affected blood vessels which usually affect small-sized vessels. AAVs include three types, granulomatosis with polyangiitis (GPA), eosinophilic granulomatosis with polyangiitis (EGPA), and microscopic polyangiitis (PMA). Despite it is common to misdiagnose them due to their overlapping with other disorders; the advancement in diagnostic approaches may contribute somehow to solve the problem of false diagnosis. In this minireview, we tried to report and summarize the most important tools in diagnosis of AAVs three types.

GPA Diagnosis Update

GPA diagnosis needs proper examination and writing down of clinical features to exclude the confusing clinical presentation. In addition, the laboratory analysis of serum ANCA autoantibodies and histopathology report of tissue specimen is not usually adequate for final diagnosis. Even though the early diagnosis of GPA is hard because of symptoms diversity and confusing histological reports, but these still important to proper management for this confused and aggressive disease. The symptoms in ear, nose, and larynx may be the first clinical features of GPA vasculitis. Thus, an attention should pay to think about GPA from otolaryngologist, particularly in otitis media cases which received treatment by antibiotics for a long period without positive respond. In orbital or sinonasal biopsies from GPA patients, increased IgG4+ plasma cells can be seen; this may cause misdiagnosis with IgG4-related disease [1]. Build on the above, the ideal path for GPA diagnosis aimed to identify, characterize and evaluate ENT signs and symptoms in GPA patients to reach earlier identification for the disease. Meanwhile, we are in need to do comparative studies that explain the diagnosis and prognosis of GPA by sinonasal imaging even if the sinus imaging in GPA gives nonspecific findings. Unfortunately, the available works of literatures were not outstanding to introduce a conclusion

lead us to exact diagnostic tools. In 2011, the American College of Rheumatology (ACR) proposed classification criteria for GPA as a following:

a) Nasal or oral inflammation: Painful or painless oral ulcers or purulent or bloody nasal discharge.

b) Abnormal chest radiograph: Pulmonary nodules, fixed pulmonary infiltrates or pulmonary cavities.

c) Abnormal urinary sediment: Microscopic haematuria with or without red cell casts.

d) Granulomatous inflammation: Biopsy of an artery or perivascular area shows granulomatous inflammation.

At least two or more of these criteria should present to provide a sensitivity of 88% and a specificity of 92% [2]. On the other hand, a professional radiologist who has the skill to find the relationship between imaging findings and vasculitis knowledge is a very important issue. Indeed, radiologists should have the ability to interpret the laboratory test findings and connect them with imaging features together with the clinical presentation. The accumulation of the above-mentioned parameters related to radiologist may lead to a meaningful differential diagnosis. Recently, a novel procedure in clinical imaging, fluorine-18-fluorodeoxyglucose (FFDG) uptake into GPA lesions in positron emission tomography (PET) had a strategic potential importance in the diagnosis of GPA. But, differentiation between malignant and benign lesion still a challenge [3,4].

EGPA Diagnosis Update

Due to its overlapping with bronchial asthma, diagnosis of EPGA includes many issues. In cases of eosinophilic airways allergy with family history of EGPA, we should think seriously about EGPA. Meanwhile, clinical reports suggest that sensitization to fungi resulting in allergic bronchopulmonary mycosis (ABPM) especially aspergillosis may contribute to EGPA pathogenesis. So, the probability of EGPA coexist is high during the course of disease. Also, EGPA may coexist with GPA in a case referred as polyangiitis overlap syndrome. Additionally, cardiac involvement e.g. Löeffler ndocarditis in EGPA may diagnosed by using a sensitive procedure, cardiac magnetic resonance imaging (CMRI) to detect cardiac lesions. It helps to detect patients who need combining therapy and helps evaluate the therapeutic effect. Furthermore, in small intestines involvement in EPGA, the capsule endoscopy, the small-bowel radiography is convenient procedure for the diagnosis of EPGA lesions in the intestine and the evaluation of their clinical course [4,5].

Lanham's criteria, ACR classification criteria and Chapel Hill definition are classification criteria and definitions commonly used for EPGA diagnosis: Lanham; asthma, eosinophilia > 1.5 ×109 /l, and clinical or pathological evidence of vasculitis involving at least two organs. ACR 1990; asthma, eosinophilia > 10%, neuropathy (mono- or polyneuropathy), non-fixed pulmonary infiltrates, paranasal sinus abnormalities, and extravascular eosinophil infiltration on biopsy [6]. Chapel Hill Consensus Conference 1994; eosinophil-rich and granulomatous inflammation involving the respiratory tract, necrotizing vasculitis affecting small to medium-sized vessels, and associated with asthma and eosinophilia. More important, the eosinophilia and edema of submucosa in EGPA is greater than in eosinophilic pneumonia [4]. Though it is difficult to point specific biomarkers in EGPA especially during times of glucocorticoid use, C-reactive protein to serum albumin ratio may provide a way in the management of AAVs including EGPA [7].

PMA Diagnosis Update

MPA diagnosis is basically performed using clinical presentations, computed tomography imaging, pulmonary or renal tissue sample, and laboratory investigations especially, serum ANCA autoantibodies analysis using immunofluorescence microscopy and ELISA. Actually, an important role can be achieved by chest computed tomography in patients with MPA to identify the abnormalities in a wide range of anatomic areas, including the whole airway. Meanwhile, fascial vasculitis in a patient with MPA can be diagnosed by magnetic resonance imaging. In addition, prostate screening should be performed while screening for malignancies in individuals diagnosed with vasculitis, including MPA, especially in the case of elderly adults with risk factors and alerting symptoms. Moreover, during vasculitis assessment cytomegaloviruses (CMV) should kept in mind because CD28- and CD57+T cells were associated with latent CMV infection but not with GPA or MPA diagnosis [3,4,8].

Conclusion Remarks

The idiopathic etiology of AAVs and uncommon occurrence contributed to increasing diagnosis difficulties. In the last few years, there were a plenty of scientific reports related to AAVs diagnosis and management especially those related to new clinical presentation. However, they couldn't reach the goal of proper diagnostic approaches and treatment to secure AAVs patients. Actually, the need for a new protocol which considers the current researches outputs is an urgent necessity. Finally, we always recommend activating the communication between health workers, especially radiologist, histopathologist, otolaryngologist, laboratorians, rheumatologist, and all internal medicine branches physicians to reach the optimum diagnosis for AAVs.

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