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LETTER TO THE EDITOR

A clinical case of eosinophilic granulomatosis with polyangiitis manifestation with a tumor in the pericardium in combination with high titers of serum immunoglobulin G4

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Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare systemic disease which is histopathologically characterized by eosinophilic infiltration, extravascular granulomas and necrotizing vasculitis with predominantly small and medium vascular involvement.[1] An accurate diagnosis of EGPA is often difficult due to clinical manifestations similar or overlapping with chronic eosinophilic pneumonia, hypereosinophilic syndrome, other primary systemic vasculitis, and hyperimmunoglobulin G4 syndrome.[2]

A 25-year-old female patient presented to a rheumatologist with complaints of severe weakness, shortness of breath with mild physical exertion, palpitations, low-grade fever in the evening, tightness of the chest, and weight loss by 6 kg within the past four months. She reported that she felt herself ill for six months. Initial complains were shortness of breath, fever, and pressure pain behind the sternum. The patient consulted a cardiologist, and echocardiography revealed a neoplasm in the anterior mediastinum (Figure 1). The hypodense structure without clear contours, semicircular in the circumference of the non-coronary and left coronary sinus of the aorta, spreading and circularly enveloping the mouth of the left coronary artery, causing a narrowing of the lumen up to 70 to 80%

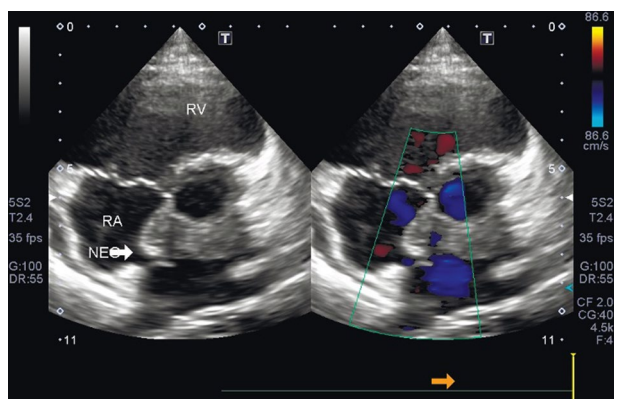

Figure 1. An echocardiographic image showing neoplasm in the anterior mediastinum.
NEO: Neoplasm; RV: Right ventricle; RA: Right atrium.

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was detected by intravenous contrast-enhanced computed aortography. A similar structure was identified along the pericardial layers in the circumference of the mouth of the pulmonary artery trunk (Figure 2). Due to the impossibility of the radical removal of the neoplasm to clarify the diagnosis, it was decided to perform a biopsy of the mass fragment. Stenting of the left coronary artery was performed. Laboratory test results were as follows: anemia (hemoglobin: 104 g/L); eosinophilia (relative numbers and absolute) 12%, 1.3-1.5×10^9/L; erythrocyte sedimentation rate 53 mm/h, and C-reactive protein 31 g/dL. Detected serum immunoglobulin G4 (IgG4) in a high titer-1,030 mg/dL (normal range: 3 to 201 mg/dL). Antineutrophilic cytoplasmic antibodies to myeloperoxidase (ANCA-MPO) were in high titer: 5.2 AI. On multislice computed tomography (MSCT) of the thorax with bolus enhancement, the contrast was accumulated along the periphery and in both lungs with multiple ring-like foci, close to the density of "ground glass" appearance.

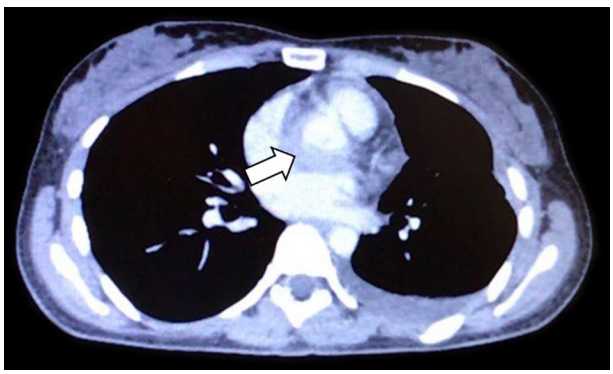

Figure 2. Computed tomography image of mediastinum. A neoplasm between aorta and pulmonary artery, as well as in transverse sinus, tightly adjacent to main vessels.

According to the pathohistological biopsy evaluation, fragments of connective tissue of varying degrees of maturity, focal fibrinoid necrosis, hyalinosis were observed and fibrous tissue was diffusely infiltrated with extravascular granuloma-like lymphohistioplasmacytic cells with an admixture of eosinophilic leukocytes, granulocytes with cellular infiltrates, among which there was focal deposits of fibrinoid masses (fibrinoid necrosis) (Figure 3). An immunohistochemical study revealed a positive expression of IgG4 in the individual plasma cells and in cellular infiltrates (Figure 4). Based on these findings, the patient was diagnosed with ANCA-associated vasculitis, EGPA.

This clinical case is of great interest to rheumatologists, cardiologists, and cardiac surgeons given the atypical manifestation of EGPA with heart damage in the form of a tumor-like mass in the pericardium. According to the literature, EGPA may be accompanied by an increased level of serum IgG4. Several studies

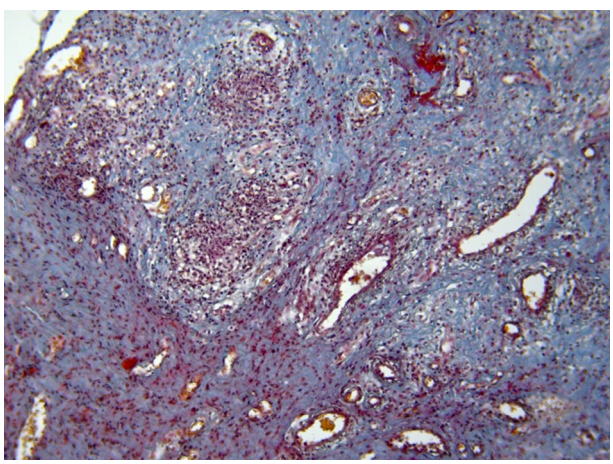

Figure 3. Among connective tissue of varying degrees of maturity, there are areas of fibrinoid changes, fibrinoid necrosis, a large number of extravascular granuloma-like lymphohistioplasmacytic infiltrates with an admixture of eosinophilic leukocytes, granulocytes. Masson's trichrome stain, ×100 magnification.

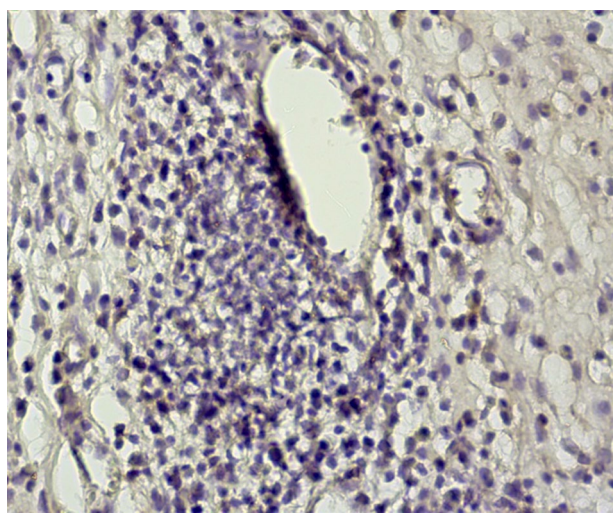

Figure 4. Positive expression of IgG4 in areas of cellular infiltrates in individual plasma cells. Immunohistochemical study with IgG4, magnification ×400.

IgG4: Immunoglobulin G4.

have shown elevated serum IgG4 levels[3] and/or tissue infiltration with IgG4-positive plasma cells in the pathological examination of biopsy material in patients with EGPA.[4,5] Severe eosinophilia and elevated immunoglobulin E (IgE) levels, which are the typical features of EGPA, are also often observed in patients with IgG4-assosiated disease (IgG4-AD).[6,7]

In conclusion, the atypicality of this clinical case is the differential diagnosis between IgG4-AD and ANCA-associated vasculitis, given the high titers of serum IgG4; however, the absence of characteristic immunohistochemical data made it possible to exclude this diagnosis. According to the literature, it can be concluded that EGPA and IgG4-AD may overlap in the course of the disease and, to some extent, the development of one disease may affect the onset of another disease due to a similar pathogenesis.

Declaration of conflicting interests

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REFERENCES

1. Jennette JC, Falk RJ, Bacon PA, Basu N, Cid MC, Ferrario F, et al. 2012 revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. Arthritis Rheum 2013;65:1-11.
2. Mahr A, Moosig F, Neumann T, Szczeklik W, Taillé C, Vaglio A, et al. Eosinophilic granulomatosis with polyangiitis (Churg-Strauss): Evolutions in classification, etiopathogenesis, assessment and management. Curr Opin Rheumatol 2014;26:16-23.
3. Vaglio A, Strehl JD, Manger B, Maritati F, Alberici F, Beyer C, et al. IgG4 immune response in Churg-Strauss syndrome. Ann Rheum Dis 2012;71:390-3.
4. Ayuzawa N, Ubara Y, Keiichi S, Masayuki Y, Hasegawa E, Hiramatsu E, et al. Churg-Strauss syndrome with a clinical condition similar to IgG4-related kidney disease: A case report. Intern Med 2012;51:1233-8.
5. Sato S, Morimoto J, Oguchi Y, Umeda T, Kawamata T, Rikimaru M, et al. A case of eosinophilic granulomatosis with polyangiitis complicated with a IgG4 related disease like symptoms. Case Reports Immunol 2018;2018:3763084.
6. Meridor K, Levy Y. Immunoglobulin G4-Related disease presenting with clinical similarity to Churg-Strauss syndrome. Isr Med Assoc J 2019;21:124-5.
7. Della Torre E, Mattoo H, Mahajan VS, Carruthers M, Pillai S, Stone JH. Prevalence of atopy, eosinophilia, and IgE elevation in IgG4-related disease. Allergy 2014;69:269-72.