Case Report

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A Rare Case of Pulmonary Complications in a Middle-Aged Female Patient Suffering from ANCA Vasculitis

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SUMMARY

We present a case of a middle-aged female patient administered to our pulmonary clinic with high fever, pharyngeal pain, thoracic pain induced with chest movements, dyspnea, night sweating and anorexia. On auscultation, we found unilateral crackle sounds and ronchi at the right lung base. Blood tests showed increased white blood cells (WBCs), CRP, urea and creatinine. Immune blood tests were ordered and the patient had increased ANCA and ANA auto-antibodies. An ANCA-associated renal vasculitis was confirmed. Our patient stayed in the hospital for 8 days. Further investigation included bronchoscopy, a thorax CT scan and renal ultrasound. The patient received vasculitis specific treatment and was discharged.

Abbreviations: WBCs: White Blood Cells; ANCA: Antineutrophilic Cytoplasmic Antibody; MPA: Microscopic Polyangiitis; MPO: Myeloperoxidase; PR3: Proteinase 3

Background

Antineutrophilic cytoplasmic antibody (ANCA) vasculitis is an autoimmune disease leading to inflammation and damage of blood vessels [1]. It is divided into three main categories including granulomatosis with polyangiitis (Wegener's granulomatosis), microscopic polyangiitis (MPA) and eosinophilic granulomatosis with polyangiitis (EGPA-Churg-Strauss syndrome) [2]. ANCA vasculitis affects mainly small blood vessels. Most of the patients have MPO-ANCA and PR3-ANCA antibodies although there are cases of patients who have not. Wegener's granulomatosis involves mainly medium and small blood vessels of upper and lower respiratory tract and then blood vessels of glomerulum. Churg-Strauss syndrome involves mainly respiratory tract with symptoms such as asthma and eosinophilia, but glomerulonephritis may also be present. Microscopic polyangiitis affects arterioles, venules and capillaries of different organs including lungs too.

Case Presentation

A 49-year-old female patient was admitted to our pulmonary department with severe pharyngeal and thoracic pain, dyspnea, night sweats, anorexia and high fever up to 39 degrees Celsius. The patient who was a non-smoker had no allergies. In the past she had one episode of post streptococcal arthritis 5 years ago which was treated with penicillin. She experienced pharyngeal and chest pain later on, initially during the night first and later on during the day. Our patient underwent an ECG test and echocardiography which were normal, but troponin and D-dimer tests were positive. The cardiologists prescribed aspirin plus low molecular weight heparin. However, our patient did not start this treatment immediately due to low levels of hemoglobin and hematocrit. Lung scintigraphy was done for estimation of lung blood supply without any complications so differential diagnosis of pulmonary embolism was excluded. On auscultation we found unilateral crackle sounds and ronchi located on right base mainly. Chest X-ray showed a few opacities into the right lung's base. Blood samples showed high levels of WBCs (18.000mm^3), CRP 12.2 mg/dl urea 87mg/dl and creatinine 2.8mg/dl. Mantoux test was negative.

Blood gas analysis showed a pO_2-61mmHg, pCO_2 26mmHg and pH-7,49 (fIO2-0,21). Our patient was negative to Covid-19. Immediately after the admission into the hospital she started ceftriaxone,

long-acting bronchodilators and oxygen therapy. Due to low hemoglobin, we proceeded to blood transfusion. Evaluation of the kidneys with renal ultrasound was negative. Further tests were ordered for immunoglobulins, proteins, ANA and ANCA antibodies. Our patient was positive to ANA and ANCA antibodies and to myeloperoxidase (MPO). During her hospitalization, our patient underwent bronchoscopy to obtain biopsy samples. Bronchoscopy didn't show any sign of obstruction. Bacterial cultures and cytology were negative. A CT scan of thorax and abdomen were done as well. Our patient had normal lymph nodes without fluid collection.

Investigations

Described in the course of the case report.

Differential Diagnosis

Differential diagnosis in our patient included pulmonary embolism, lung abscess, bacterial or viral pneumonia, inherited abnormal airway malformations. Lung scintigraphy excluded pulmonary embolism while CT scan and bronchoscopy excluded inherited lung complications. Bacterial cultures and cytology obtained via bronchoscopy were negative.

Treatment

Medication included ceftriaxone, proton pump inhibitors for stomach protection, bronchodiallators, NSAIDs and oxygen therapy. After the patient was treated for her pulmonary symptoms, we proceeded to treatment of her vasculitis in cooperation with our rheumatologists. Her treatment included high dose of glucocorticoids together with cyclophosphamide as a remission therapy and then rituximab for remission maintenance.

Outcome and Follow Up

During the last days of her hospitalization, our patient had no fever or pain. Her symptoms improved and was discharged taking only maintenance treatment for vasculitis remission.

Discussion

ANCA vasculitis is a rare autoimmune disorder affecting small and medium size blood vessels [1]. It is divided into granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis and microscopic polyangiitis affecting different organs and systems but mainly lungs and kidneys as ANCA glomerulonephritis [2] due to their high vascularization including thousands small blood vessels [3]. In some cases it may involve also children [4]. ANCA autoantibodies can be connected with two proteins mainly called proteinase 3 (PR3) and MPO [3]. ANCA vasculitis can be divided based on severity into severe and non-severe and based on the extension into limited or systemic [5]. The triggering of this autoimmune disease can be genetic, environmental or combined with other factors too related to activation with different cellular types such as T-cells [6]. The most severe and deadly clinical manifestations include massive alveolar and glomerular hemorrhage, gastrointestinal, cardiac or central nervous system complications. The evaluation of severity is based on Birmingham Vasculitis Activity score (BVAS) [7]. The therapy is based on a variety of different drugs including glucocorticoids and later rituximab useful for remission of acute symptoms [8-11] as well as methotrexate, mycophenolate, leflunomide and azathioprine for remission maintenance therapy [10,12].

In some cases, in patients with severe symptoms and complications, plasma exchange can be used in order to avoid further complications and disorders especially in patients with acute glomerulonephritis [13,14]. In our case and following guidelines for vasculitis therapy, we started with high dose of glucocorticoids and later cyclophosphamide. Based on the fact that our patient didn't know her situation about vasculitis, we started following this therapy in order to suppress a possible worsening of symptoms of our patient. Later, we added rituximab for remission maintenance. Finally, our patient had an improved clinical and laboratory status and she was discharged from the hospital instructing her to come for re-evaluation after 3 months.

References

1. Abdallah Qasim, Jayesh B Patel (2024) ANCA Positive Vasculitis.
2. Max Yates, Richard Watts (2017) ANCA-associated vasculitis. Clin Med (Lond) 17(1): 60-64.
3. Mark E McClure (2018) Vasculitis Research Fellow, Addenbrooke's Hospital, Cambridge, Rachel B, Jones, Consultant in Nephrology and Vasculitis, Addenbrooke's Hospital, Cambridge ANCA Associated Vasculitis and the Kidney.
4. Mojca Zajc Avramovič, Tadej Avčin, Marina Vivarelli Chapter (2023) Renal Vasculitis in Children. Pediatric Kidney Disease, pp. 707-736.
5. Karla N Samman, Carolyn Ross, Christian Pagnoux, Jean Paul Makhzoum (2021) Update in the Management of ANCA-Associated Vasculitis: Recent Developments and Future Perspectives. Int J Rheumatol 2021: 5534851.
6. Jennette JC, Nachman PH (2017) ANCA glomerulonephritis and vasculitis. Clinical journal of the American Society of Nephrology: CJASN 12(10): 1680-1691.
7. Mukhtyar C, Lee R, Brown D, O Flossmann, C Hall, et al. (2009) Modification and validation of the Birmingham Vasculitis Activity Score (version 3). Annals of the Rheumatic Diseases 68(12): 1827-1832.
8. Mendel A, Ennis D, Go E, Volodko Bakowsky, Corisande Baldwin, et al. (2020) CanVasc consensus recommendations for the management of antineutrophil cytoplasm antibody-associated vasculitis: 2020 update. The Journal of Rheumatology 48(4): 555-566.
9. Yates M, Watts RA, Bajema IM, M C Cid, B Crestani, et al. (2016) EULAR/ERA-EDTA recommendations for the management of ANCA-associated vasculitis. Annals of the Rheumatic Diseases 75(9): 1583-1594.
10. Ntatsaki E, Carruthers D, Chakravarty K, David D Cruz, Lorraine Harper, et al. (2014) BSR and BHPR guideline for the management of adults with ANCA-associated vasculitis. Rheumatology (Oxford) 53(12): 2306-2309.
11. McGeoch L, Twilt M, Famorca L, Volodko Bakowsky, Lillian Barra, et al. (2015) CanVasc recommendations for the management of antineutrophil cytoplasm antibody (ANCA)-associated vasculitides - executive summary. Canadian Journal of Kidney Health and Disease 2: 43.

12. Geetha D, Jin Q, Scott J, Zdenka Hruskova, Mohamad Hanouneh, et al. (2018) Comparisons of guidelines and recommendations on managing antineutrophil cytoplasmic antibody-associated vasculitis. Kidney International Reports 3(5): 1039-1049.

13. Bernhard Hellmich, Beatriz Sanchez Alamo, Jan H Schirmer, Alvise Berti, Daniel Blockmans, et al. (2024) EULAR recommendations for the management of ANCA-associated vasculitis: 2022 update. Ann Rheum Dis 83(1): 30-47.

14. Peter A Merkel, Andre A Kaplan, Ronald J Falk (2024) Granulomatosis with polyangiitis and microscopic polyangiitis: Induction and maintenance therapy. In: Gerald B Appel, Fernando C Fervenza, Albert Q Lam (Eds.).,.

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