Received: 24 May 2022 | Revised: 24 August 2022 | Accepted: 20 September 2022

DOI: 10.1002/ccr3.6445

CASE REPORT

Clinical Case Reports Open Access WILEY

Granulomatous polyangiitis misdiagnosed as hematogenous lung abscess: A case report

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Abstract

Granuloma polyangiitis (GPA) is a necrotizing granulomatous inflammation, which is a systemic autoimmune disease that mainly affects the upper respiratory tract, lungs, and kidneys. Clinically, the clinical manifestations of GPA vary greatly, and it is extremely easy to be misdiagnosed. We report a 60-year-old man with granulomatous polyangiitis with dysuria and parotid gland enlargement as the first symptom. As the condition worsened, he gradually developed symptoms in multiple systems such as persistent high fever, nosebleeds, hemoptysis, skin rash, and multiple cavities in the lungs. Due to high fever, hemoptysis, and the gradual expansion of lung cavities, the suspected hematogenous lung abscess was not well controlled and transferred to the respiratory department. The manifestation of involvement was finally confirmed by skin biopsy. Biopsy was performed on the skin of the lower extremities with a dark red rash with multiple microprojections. Pathology indicated neutrophilic infiltration and necrosis of small vessel walls, and granuloma formation. Blood anti-protease 3 antibody (PR3) was positive Monotherapy with prednisone. Body temperature gradually returned to normal; hemoptysis and nasal bleeding disappeared; parotid gland enlargement and dysuria relieved, and lung cavities gradually reduced. When anti-infective treatment is ineffective, we should consider the presence of some non-infectious diseases, especially when multiple systems are involved; biopsy should be performed as soon as possible. Granulomatous polyangiitis has various manifestations. Parotid glands and prostate may also be the first organs involved, not limited to common targets such as the respiratory tract and kidneys.

KEYWORDS

antineutrophil cytoplasmic antibody, granuloma polyangiitis, lung abscess, parotid gland, prostate

1 | INTRODUCTION

Antineutrophil cytoplasmic antibody (ANCA) associated vasculitis is a systemic autoimmune disease that primarily affects small and medium-sized blood vessels (i.e., capillaries, venules, arterioles and arterioles). Anca-associated vasculitis includes three phenotypes: granuloma polyangiitis (GPA, also known as Wegener granuloma), microscopic polyangiitis (MPA) and eosinophilic granuloma polyangiitis (EGPA).[1] GPA is a necrotizing granulomatous inflammation, mainly affecting the upper respiratory tract, lungs, and kidneys. Clinically, the clinical manifestations of GPA vary greatly, and it is extremely easy to be misdiagnosed. We report a case of GPA with prostate, parotid and groin involvement as the first symptom.

2 | CASE DATA

2.1 | Basic information

The patient was a male of 60 years old, who was admitted to the hospital on May 22, 2020, due to the "Finding of a mass for a month above the surgical incision for the treatment of an inguinal hernia performed 5 years ago." The patient underwent tension-free repair of the right inguinal hernia at the local hospital 5 years ago due to the right inguinal hernia, and recovered well after the operation. One month ago, a mass was found above the hernia incision, with tenderness. Twenty days ago, the patient started to experience difficulty urinating and was admitted to the Department of Urology at the First Affiliated Hospital of an anonymous medical university. Prostate biopsy was indicated inflammation. During hospitalization, the patient had a high fever (up to 39.0°C), with swelling and pain in the right parotid area (Figure 1), and the swelling was gradually increasing; the mass above the hernia incision was further enlarged with sinus formation; no obvious abnormality was seen in the chest radiograph. After 1 week of anti-infective treatment with cefoperazone and sulbactam, there was no improvement in fever, and the masses in the parotid area and above the hernia were further enlarged. Since abscesses were considered, the patient was transferred to the Department of General Surgery of our hospital for further diagnosis. The patient lost about 10 kg in the past 1 month.

2.2 | Admission examination

One palpable mass of about 4 cm in diameter could be seen in the right maxillofacial region, with hard texture, clear boundary, normal skin, mild tenderness without no redness, and the skewing of the corner of the mouth to the left could be visible. The breath sounds were rough in both lungs, and no dry and moist rales could be heard. A mass with a diameter of about 2 cm could be palpable above the incision in the right groin area, with soft texture and mild tenderness, and no redness and swelling could be visible; the boundary with surrounding tissues was clear, and the skin temperature was normal.

FIGURE 1 Swollen parotid glands.

2.3 | Diagnosis and treatment

Chest CT scan (Figure 2) showed multiple nodules in both lungs, and the larger one was 2.9×2.6 cm. Leukocytes, procalcitonin, and C-reactive protein were all elevated (Table 1). Considering that the secondary infection of the hernia patch caused multiple blood-borne abscesses throughout the body, the General Surgery Department immediately gave piperacillin and tazobactam 4.5 g q12h ivgtt, while parotid area tumor incision and drainage were performed, with drainage of yellow slightly turbid liquid. After anti-infective treatment and parotid area incision and drainage for 4 days, the mass in the parotid area was significantly reduced in size, but the patient continued to have fever every day above 39.0°C without chills. On May 26, the antibacterial drugs were switched to meropenem 1.0 g, Q8h, ivgtt and linezolid 600 mg, Q12h, ivgtt, and the blood culture was performed. As of May 29, the patient continued to have a high fever, coughing up a small amount of white sputum and bloodshot sputum. Re-examination of Chest CT revealed that the number of lung nodules increased and enlarged, accompanied by formation of small cavities (Figure 2). Considering that the

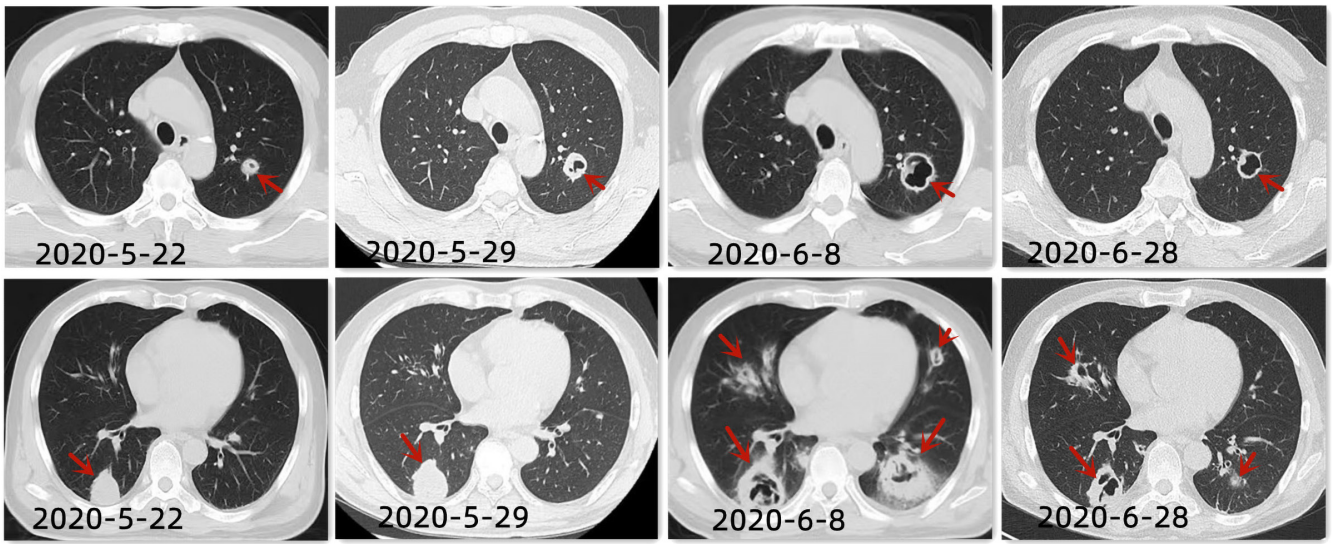

FIGURE 2 Chest CT showed multiple pulmonary nodules and cavities, after glucocorticoid treatment, the lesion was significantly reduced.

blood-borne lung abscesses, the anti-infective treatment with meropenem combined with linezolid was given to the patient continuously, but the patient's body temperature was still above 39.0°C, and the blood in the sputum was more than before, with intermittent nosebleeds.

The patient was then transferred to the intensive care unit of our department on June 02 and the antibiotics were adjusted to vancomycin 1.0 g Q12h ivgtt combined with imipenem and cilastatin sodium 0.5 g q6h, ivgtt. Hemoptysis was still getting worse, about 50 ml per day. The patient developed shortness of breath at rest, and the blood oxygen saturation was 92%–95% with oxygen inhalation at 3 L/min; the high fever was still persistent. On June 04, a bronchoscopy (Figure 3) showed bronchial mucosa hemorrhage with granulation formation. Because the patient had bleeding and low blood oxygen saturation, his family members did not agree to perform biopsy, so the basal segment of the inferior lobe of the right lung was lavaged and the lavage was submitted for metagenomic next-generation sequencing (mNGS) and culture for bacteria and fungi; at the same time, anti-candida treatment with fluconazole was given to the patient. On June 07, mNGS reported human herpesvirus type IV, no fungi or bacteria were found, and no mycobacterium tuberculosis was found. However, human herpesvirus type IV infection could not fully explain the patient's current condition. Since no fungi or bacteria were found, it is more likely that the patient was with a systemic non-infectious disease. Re-examination with enhanced CT (June 09, Figure 2) showed: right parotid gland lesions, possibly abscesses; paranasal sinusitis and sinusitis; multiple lung nodules, possibly inflammatory lesions, more severe and extensive than before; bilateral pleural hypertrophy; a small amount of pleural effusion on the left side; a cyst in the tail of the pancreas; significantly reduced spleen density; irregular thickening of the bladder wall, possibly cystitis; right inguinal abscess. On June 08, dark red rashes slightly raised from the skin scattered on both lower extremities were found (Figure 4). Otolaryngology consultation: polyp in the left middle nasal passage, bilateral nasal mucosa congestion and edema. Since nasal cavities, sinuses, parotids, lungs, bladder, prostate and skin were involved, the patient was considered to have a higher possibility of systemic disease, so immune-related tests (antineutrophil cytoplasmic antibody [ANCA], antinuclear antibody, antinuclear antibody profile, complements, immunoglobulins, rheumatoid factors, etc.) were further performed. A fusiform incision of about 1 cm was performed at a larger rash on the right lower extremity, and the rash and subcutaneous tissue were excised, and the specimens were submitted for pathological examination. ANCA report showed: anti-protease 3 antibody (PR3) positive, anti-myeloperoxidase antibody (MPO) negative, anti-glomerular basement membrane antibody (GBM-Ab) negative. Skin histopathology (Figure 4) showed: Infiltration and necrosis of neutrophils in the walls of small blood vessels, infiltration of inflammatory cells dominated by histiocytes, lymphocytes and neutrophils, granuloma formation, and fibrinoid necrosis of the small vascular walls in some areas. These findings were more in line with the acute phase changes of Wegener's granulomatosis.

Based on multi-site involvement, ineffective anti-infective treatment, positive for PR3, and Wegener granuloma seen in pathological examination, granulomatous polyangiitis was diagnosed on June 11, and treatment with glucocorticoids + immunosuppressants + immuno-

TABLE 1 Data of patient laboratory examinations.

Items	Day 1 of anti-infective treatment	Day 4 of anti-infective treatment	Day 8 of anti-infective treatment	Day 11 of anti-infective treatment	Day 17 of anti-infective treatment (Day 1 of hormone therapy)	Day 20 of hormone therapy
C-reactive protein (0–8 mg/L)	127.00	133.93	229.00	202.70	331.00	66.82
Procalcitonin (<0.1 ng/ml)	0.47	0.14	0.17	0.19	1.01	0.14
White blood cells ($3.5–9.5\times10^9$/L)	13.12	11.92	12.87	12.48	16.28	12.11
Neutrophils ($1.8–6.3\times10^9$/L)	10.34	9.92	11.37	10.75	14.92	11.32
Lymphocytes ($1.1–3.2\times10^9$/L)	2.09	0.94	1.00	1.10	0.81	0.48
Monocytes ($0.1–0.6\times10^9$/L)	0.05	0.77	0.48	0.61	0.31	0.30
Eosinophils ($0.02–0.52\times10^9$/L)	0.62	0.25	0.01	0.02	0.22	0.10
Platelets ($125–350\times10^9$/L)	344.00	264.00	321.00	286.00	282.00	258.00
Hemoglobin (130–175 g/L)	120	118	121	126	106	112
Total bilirubin (≤26 μmol/L)	9.8	10.7	12.9	12.3	14.5	9.0
Aspartate aminotransferase (15–45 U/L)	65	67	56	34	26	18
Creatinine (57–111 μmol/L)	69	67	73	77	72	76
Oxygen partial pressure/oxygen absorption concentration	319	311	289	256	218	343
Detection of T cells in tuberculosis infection (≤11 SFCs/2.5×10^5 PBMCs)	–	–	0	–	0	–
Acid-fast bacilli detection	–	Negative	Negative	Negative	Negative	–

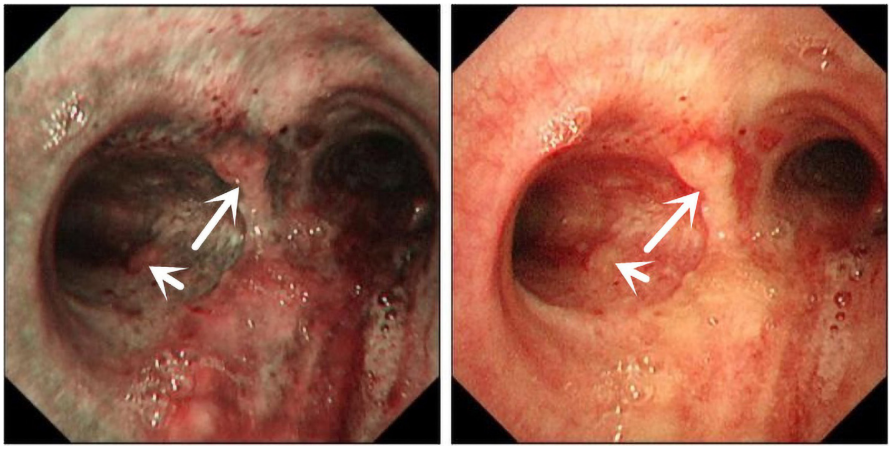

FIGURE 3 The formation of multiple granulomas can be seen under bronchoscopy.

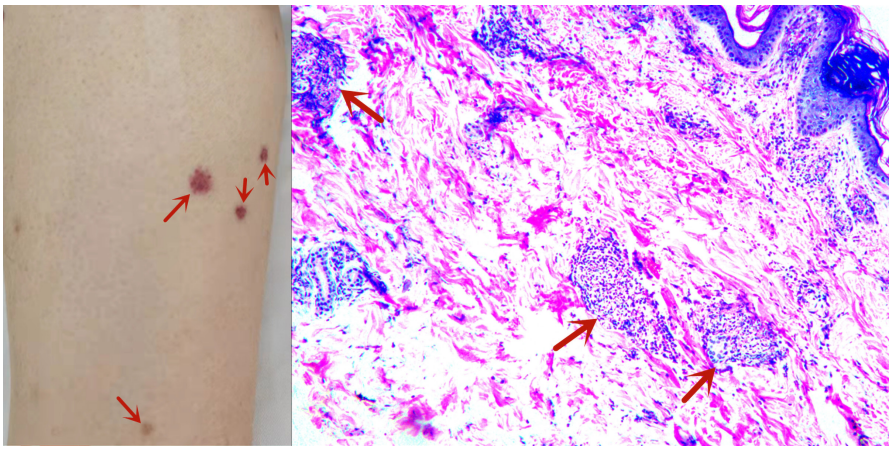

FIGURE 4 Red rash on the right lower limb, pathology of biopsy (HE staining, 10×10): Infiltration and necrosis of neutrophils in the walls of small blood vessels, and granuloma formation.

globulin were to be given to the patient. However, after explaining the medical condition and treatment plan to the patient and his family members, they said that the medical condition continued to deteriorate for a month since they went out to seek medical attention, and they were unwilling to seek any treatment and strongly requested that the patient be discharged from the hospital. Therefore, the patient was advised to try oral prednisone (50 mg/day) for treatment, and contact us at any time.

After returning to his hometown on June 12, the patient took oral prednisone for treatment every day according to our advice. On June 15, fever was significantly reduced, and hemoptysis was reduced without any nosebleeds. The patient and his family members felt that the treatment was effective, so he continued taking oral prednisone for treatment at home. On June 25, his body temperature completely dropped to normal, and his appetite was significantly improved; there was no hemoptysis, and shortness of breath was completely relieved. The dose of prednisone was adjusted to 40 mg/day and the patient continued taking it. On June 28, the clinical symptoms disappeared completely, and the patient returned to our department again for a CT scan of the chest, which showed that the lesions in both lungs were significantly reduced compared with those in the past (Figure 2). Considering that the therapeutic effect of monotherapy with oral prednisone was good, the patient was recommended to take it at 30 mg/day, with a gradual decrease in the dose and discontinuation eventually. Since then, the patient has been followed up many times over the phone, and the general condition of the patient was good; he had no special discomfort but was unwilling to visit the hospital for follow-up.

3 | DISCUSSION

The cause of GPA is not yet fully understood. At present, it is mainly considered to be an autoimmune-related disease, which is genetically susceptible and can be induced by surrounding environmental factors. In PR3-AAV, genome-wide association studies have revealed its association with class II HLA genes, especially with HLA-DPB1 * 04:01.[2] The main target antigen of cytoplasmic (c)-ANCA is protease 3, and cANCA is of high specificity to GPA, with a direct relationship between the titer of c-ANCA and GPA disease activity.[3] The detection of c-ANCA is of great significance for the diagnosis of GPA. The clinical manifestations of GPA are diverse, with involvement of nose, throat, bronchi, lungs, kidneys, eyes, skin and mucous membranes,

nervous system, joints, etc.[4–6] EGPAs without respiratory system symptoms have been reported recently,[7] such cases will begin to appear in granulomatous polyangiitis. A few GPA presented as glandular involvement,[8] such as parotid gland enlargement, prostate enlargement, salivary gland enlargement, etc. Currently, there are only a very small number of case reports of GPA with parotid gland enlargement or prostate enlargement as the main clinical symptom,[9–14] and the mechanism has not been completely clarified.

The patient in this case started with prostate and parotid gland involvement, which is really rare; in addition, the presence of a mass above the right inguinal hernia patch opening is highly misleading as it could be easily assumed to be the hernia patch rejection and infection, with secondary systemic bloodstream infection leading to abscesses in the lungs and parotid glands. We also made a mistake initially in the diagnosis and treatment of this patient and were misled by the appearance. This case retrospectively according to the latest GPA classification score standard[1] score: nasal bloody discharge (3), endobronchial involvement (2), antiproteinase 3 (anti-PR3) antibody (5), pulmonary cavitation (2), biopsy suggests granulomatous inflammation (2), sinusitis (1), a total of 15 points, met the GPA diagnostic score.

This patient had fever, pulmonary cavitation, and bacterial infection indicators significantly increased, which could be easily mistaken for infectious diseases, but the situation did not improve after adequate anti-infection treatment. At this time, we re-adjusted our thinking and looked for non-infectious disease factors. After biopsy and ANCA examination, we found GPA in time. Our experience is that it is important to consider the presence of non-infectious diseases when anti-infective therapy fails, especially when the patient has clinical symptoms in multiple organs.

Treatment of GPA mainly includes the combination of adrenocortical hormone and immunosuppressive agents, and high-dose intravenous immunoglobulin therapy. The active phase is dominated by hormone pulse therapy plus immunosuppressive agents therapy, but the latest guidelines also mentioned that minimizing exposure to glucocorticoid is essential to improve prognosis.[15] This case was initially treated with oral prednisone (50 mg daily) without immunosuppressive agents, which indicated that some GPA could be cured with conventional dose of glucocorticoid alone.

The clinical manifestations of GPA are lack of specificity, and the early symptoms are mostly atypical. Misdiagnosis or missed diagnosis is easy to occur. It is necessary to jointly improve the understanding of GPA.

AUTHOR CONTRIBUTION

Yong Yang and Xiao-Yue Chang: collected clinical data and wrote manuscripts.

ACKNOWLEDGEMENTS

This article has not been published elsewhere in whole or in part. All authors have read and approved the content and agree to submit for consideration for publication in the journal.

DATA AVAILABILITY STATEMENT

Data can be obtained from the corresponding author upon request.

ETHICAL APPROVAL

There are no ethical/legal conflicts involved in the article.

CONSENT

Written informed consent was obtained from the patient for publication of this case.

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How to cite this article: Yang Y, Chang X-Y. Granulomatous polyangiitis misdiagnosed as hematogenous lung abscess: A case report. *Clin Case Rep*. 2022;10:e06445. doi: 10.1002/ccr3.6445