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CASE REPORT

Intracerebral Hemorrhage with Churg Strauss-Syndrome: Multidisciplinary Collaboration and Literature Review

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Objective: To explore the clinical characteristics and treatment outcomes of intracerebral hemorrhage in eosinophilic granulomatosis with polyangiitis (EGPA).

Methods and Patient Presentation: We report an 18-year-old student of EGPA complicated with intracerebral hemorrhage. The laboratory tests showed a continuous increase in eosinophils. The CT of head and chest showed cerebral hemorrhage and pulmonary infiltration.

Interventions: The patient received an intravenous infusion of methylprednisolone 1g/(kg·d) and cyclophosphamide for 3 days, followed by oral prednisone 1 mg/(kg·d).

Outcomes: At discharge, the patient's head and chest CT showed obvious absorption of intracranial hematoma and improvement of pulmonary infiltration. We reviewed 40 previously published cases of EGPA with intracerebral hemorrhage focusing on the clinical features and treatment of intracerebral hemorrhage caused by EGPA.

Conclusion: For the cases of EGPA complicated with intracerebral hemorrhage, we should timely differentiate diagnosis and recognition. Early diagnosis with aggressive immunosuppressive therapy can help improve the prognosis of patients EGPA with intracerebral hemorrhage. When a patient is affected by EGPA, it is essential to remain vigilant for signs of Central Nervous System involvement. The treatment with glucocorticoids and cyclophosphamide is effective in managing EGPA.

Keywords: Churg Strauss-Syndrome, eosinophilic granulomatosis with polyangiitis, eosinophilia, intracerebral hemorrhage

Introduction

Churg-Strauss Syndrome (CSS) was known as Eosinophilic Granulomatosis with Polyangiitis (EGPA). The name of CSS is derived from the first case of EGPA reported by Churg and Strauss in 1951.[1,2] EGPA is a disease associated with anti-Neutrophil Cytoplasmatic Antibody associated vasculitides, alongside granulomatosis with polyangiitis and microscopic polyangiitis.[3] It is a rare systemic disease characterized by asthma, transient pulmonary infiltration, eosinophilia, and systemic vasculitis.[4] EGPA progresses rapidly and has a variety of clinical manifestations, involving multiple organs and systems.[5] The respiratory tract and lungs are the earliest and most easily involved,[6] and most of the first symptoms are wheezing seizures and nasosinusitis symptoms, followed by the nervous system. Nervous system involvement is common in EGPA, especially peripheral neuropathy,[7,8] which is also included in the classification criteria for EGPA. In contrast, EGPA rarely affects the central nervous system, Hemorrhagic stroke in EGPA is extremely rare.[9] In patients with EGPA, most patients show with upper respiratory tract and lung involvement, such as nasosinusitis, asthma, and wandering pulmonary infiltrating shadow, and some patients may show peripheral neuropathy, skin damage, and heart involvement. Among, Eosinophilia is one of the cardinal features in EGPA.[10] Recent studies have shown that the global incidence of the disease is 2.5 cases per 100,000 adults per year,[11] with incidence peaking at ages 30–40 or 55–64.[12]

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However, the pathogenesis of EGPA remains unclear, and it is currently believed to be related to environmental, genetic factors and immune disorders.[10]

Before 1990, the EGPA could be diagnosed when patients developed asthma, the number of eosinophils in peripheral blood exceeded 1.5×10^9 /L, and two organs were affected.[9] This diagnosis only relies on clinical manifestations, without histopathological diagnosis, is simple and easy to use, and is widely used by clinicians. After 1990, the diagnostic criteria for EGPA mainly refer to the classification criteria proposed by the American College of Rheumatology in 1990. According to the Diagnostic criteria of the American College of Rheumatology (ACR),[1] EGPA can be diagnosed when four or more of these conditions are satisfied. ACR criteria: Presence of four or more of the following: 1) eosinophilia (more than 10%), 2) asthma, 3) pulmonary infiltrates, 4) paranasal abnormalities, 5) neuropathy, and 6) extravascular eosinophilia on biopsy.[13] In 2012, Chapel Hill consensus pointed out that the definition of EGPA should include eosinophilic granulomatous inflammation in tissues, necrotizing small vasculitis, blood eosinophilia, and asthma.[14] With the deepening of EGPA research, more specific MIRRA classification criteria were proposed in clinical studies in 2017.[15] The main criteria were: (1) history of asthma; (2) Eosinophilia (peripheral blood eosinophilia ratio > 10% or absolute value > 1.0×109 /L). Secondary criteria (meeting at least 2 criteria) were: (1) Positive biopsy: perivascular eosinophilic infiltration; (2) Neuropathy: single or multiple neuropathy; (3) pulmonary infiltration; (4) sinus lesions; (5) Cardiac color ultrasound or cardiac magnetic resonance imaging suggested cardiomyopathy; (6) glomerulonephritis; (7) Hematuria, proteinuria; (8) Alveolar hemorrhage diagnosed by alveolar lavage fluid; (9) Skin purpura; (10) Positive ANCA (MPO or PR3) test. In 2018, China "Multidisciplinary Expert Consensus on Diagnosis and Treatment of EGPA": a. Asthma-like lesions (wheezing, cough, chest tightness and dyspnea); b. Eosinophilia (≥10% or absolute value ≥1.5*109/L); c. Multiple or single peripheral neuropathy; d. Non-fixed pulmonary infiltration; e. Sinusitis; f. Extravascular eosinophilic infiltration. The Meeting 4 or more of the 6 classification criteria can be diagnosed as EGPA. In this paper, we report a case of left frontotemporal parietal cerebral hemorrhage, and summarize and review previous reports of cerebral hemorrhage and EGPA.

Case Presentation

On January 13, 2021, an 18-year-old student suddenly lost consciousness at school. He was admitted to the hospital emergency department for treatment. Prior to admission, he had a persistent fever and headache for more than 10 days. On December 30, 2020 (10 days before hospitalization), he was treated at a small local clinic, where doctors treated his fever with cephalosporin antibiotics and methylprednisolone sodium succinate for injection. After treatment, his fever and headache did not ease. On 12 January 2021 (one day before admission), his headache symptoms worsened. Then, he suddenly lost consciousness, accompanied by convulsions and vomiting. He underwent a head and chest CT scan at a local hospital on the same day. Head CT showed high-density imaging of the left temporal lobe. Chest CT showed lower right pneumonia. On January 13, 2021, he was transferred to our hospital for further treatment.

In 2007, he underwent hernia surgery at a local hospital. History of allergy was not known. History of asthma, rhinitis was also not known. His parents denied that he had a history of high blood pressure, heart disease, diabetes, smoking and taking illegal drugs.

On admission, the patient had a body temperature of 36.5°C. Physical examination revealed blood pressure of 132/69 mmHg and a regular pulse rate of 56 beats/min. Chest auscultation revealed expiratory wheezes in the bilateral lower lung field. There was no palpable enlargement of systemic superficial lymph nodes. Electrocardiogram shows a normal heart rate. Neurosurgical examination: blurred consciousness, misorientation, Glasgow Coma Score: 13 (mild disturbance of consciousness). He is sensitive to light. Stretching the tongue is not biased. The muscle strength of four limbs is □. Muscle tension is normal. And bilateral knee tendon reflex is symmetrical.

Laboratory examinations revealed a white blood cell (WBC) count of 12.31×10^9/L (normal range: 3.5–9.5×10^9 /L), a hemoglobin level of 134 g/L, and a platelet count of 65×10^9 /L (normal range: 125–350 $\times10^9$ /L). The neutrophil count was 10.44×10^9/L (normal range: 2.04–7.5×10^9 /L). The ratio of neutrophils was 84.70% (normal range 51–75%). The lymphocyte count and the ratio of lymphocyte were both reduced, 0.76×10^9/L (normal range 0.8–4.0×10^9/L) and 6.20% (normal range 20–50%), respectively. However, the eosinophilic granulocyte was abnormal. The count of eosinophilic granulocyte was 0.00×10^9/L (normal range 0.02–0.52×10^9/L). The creatinine level was 66 μmol/L (normal range: 53–97

μmol/L), with an increased Hypersensitive C-reactive protein (Hs-CRP) of 127.86 mg/L (≤5 mg/L). Urinalysis showed proteinuria. The patient presented with an increased aspartate aminotransferase of 42 U/L (normal range 15–40 U/L) and increasedγ-glutamyl transpeptidase (76.6 U/L) (normal range ≤73). The procalcitonin (PCT) was 0.13 ng/mL (<0.1 No bacterial infection; 0.1≤PCT<0.25 may be no bacterial infection). The coagulation indexes were all within the normal range. And the other laboratory examinations showed no abnormality.

Treatment Process

The above is the detailed process of clinical examination, treatment and diagnosis of a patient with EGPA complicated with intracerebral hemorrhage reported in this paper, as shown in Table 1. During the diagnosis and treatment of this case, the clinical pharmacist participated in the treatment and made recommendations for diagnosis and treatment. The count of eosinophilic granulocyte gradually increased from the third day of admission. Its peak reached maximum on January 22. The count of eosinophilic granulocyte was more than 10 times the normal range. Changes in eosinophilic granulocyte count and ratio during hospitalization are shown in Figure 1. The cerebrospinal fluid was detected by metagenomic next-generation sequencing (mNGS). No pathogenic microorganism infection was detected by mNGS. The MRA+MRV did not find vascular malformation (Figure 2). However, MRI revealed left inferior turbinate hypertrophy, right maxillary sinus cyst, and sphenoid sinus hypertrophy. It suggested that the patient had a history of sinusitis and allergic rhinitis. MRI and head CT showed left frontotemporal and parietal cerebral hemorrhage with soft tissue swelling in the left frontotemporal region (Figures 3 and 4), and chest CT showed shadows in the lower lobes of both lungs (Figure 5). Clinical pharmacists summarized the above results for a literature search. According to the American College of Rheumatology Criteria,[1,2] EGPA was diagnosed based on clinical symptoms and laboratory findings. He was treated with methylprednisolone and cyclophosphamide and was discharged on February 9. On discharge on February 9, CT of the chest showed that shadow of lower lobe of both lungs was more absorbed than those on admission (Figure 6) and intracranial hematoma was more absorbed than those on admission (Figure 7).

Review of the Literature

We searched the literature published in PubMed, Web of Science, and Embase databases using the MeSH terms or keywords: "Eosinophilic Granulocyte, Eosinophilia, Intracerebral Hemorrhage, Cerebral Hemorrhage, Eosinophilia with Intracerebral Hemorrhage, Churg-Strauss syndrome, Eosinophilic Granulomatosis with Polyangiitis" until October 2024. Case reports and case series of patients with the diagnoses of CSS or EGPA were eligible for inclusion. Publications were excluded if they did not meet the above criteria. As a result, 39 articles described EGPA with intracerebral hemorrhage were identified.[16–54] We systematically reviewed clinical features and therapies of EGPA with intracerebral hemorrhage. Clinical information in the previous reports are summarized in Table 2, including age, sex, clinical features, Systemic involvement, and therapies.

Discussion

EGPA is a rare systemic disease characterized by vascular involvement in multiple organs, such as respiratory tract, cutaneous tissue, gastrointestinal tract, cardiovascular system, paranasal sinus abnormality, arthritis/arthralgia, and peripheral nerves. The patient usually has a history of chronic asthma, long-standing rhinitis, recurrent sinusitis, nasal obstruction, and nasal polyposis. Asthma is one of the most common diseases associated with EGPA, including IgE-mediated allergic asthma, exercise-induced asthma, non-allergic asthma caused by viral upper respiratory tract infection or no obvious trigger, and cough variant asthma.[55] The clinical manifestations of EGPA is characterized by asthma, peripheral eosinophilia, peripheral neuropathy and pulmonary invasion. The hallmark laboratory indicators of the disease are peripheral blood eosinophilia. The CT sinuses and CT chest are beneficial in determining sinus and respiratory involvement, respectively. ANCA was detectable in only 40% of patients, indicating vasculitis.

Granulomatosis with polyangiitis (GPA) is characterized histologically by necrotizing granulomatous inflammation in addition to vasculitis. Although the target organs of GPA and EGPA are similar, the clinical and pathological manifestations of GPA and EGPA are different, which can help to distinguish them. Both of them are easy to invade the respiratory system, but GPA often forms destructive lesions, such as nasal mucosa ulcers and pulmonary cavities. The respiratory

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Table 1 The Clinical Examination, Treatment and Diagnosis Process of EGPA with Intracerebral Hemorrhage

Timeline	Symptoms and Blood test index	Treatment	Suggested inspection	Improves or Worsens	Diagnosis
January 14, 2021	Persistent headache; Persistent fever; Lower right pneumonia; Confusion of consciousness; WBC: 12.31×109/L; Hs-CRP: 127.86 mg/L (≤5 mg/L)	Vancomycin 1 g/q12h, Meropenem 2 g/q8h; hemostasis;	Blood routine examination of various indicators	No improvement	Suspected Intracranial infection, Cerebral hemorrhage, Tumor
January 15, 2021	Confusion of consciousness; Headache; Head CT:the left temporal lobe exhibits a high-density shadow (Figure 4); WBC:9.97×109/L	Vancomycin 1 g/q12h Meropenem 2 g/q8h	Blood routine examination of various indicators; Head CT	Headache worsen	Cerebral hemorrhage; Suspected Inflammatory bleeding due to infection; Suspected Arteriovenous malformation bleeding
January 16, 2021	Confusion of consciousness; Headache; Eosinophilic granulocyte count: 0.67×109/L; MRA: Arteriovenous malformations were absent (Figure 2). Cerebral hemorrhage in the left frontotemporal occipital lobe was reported by MRI (Figure 3). Left inferior turbinate hypertrophy, Right maxillary sinus cyst, sphenoid sinus hypertrophy were reported by MRI.	Vancomycin 1 g/q12h Meropenem 2 g/q8h	Skull magnetic resonance imaging; Blood routine examination of various indicators; Respiratory physician consultation: The cerebrospinal fluid detected by mNGS;	No improvement	Suspected Inflammatory bleeding due to infection
January 17, 2021	Symptoms did not ease. The laboratory tests reported increased eosinophilic granulocyte (1.44×109/L) in Table 2. Chest CT: Shadow of lower lobe of both lungs (Figure 5)	Vancomycin 1 g/q12h Meropenem 2 g/q8h	Blood routine examination of various indicators; Consult a clinical pharmacist: Auto-antibodies detected and Chest CT were recommended	No improvement	
January 18, 2021	Eosinophilic granulocyte count: 1.88×109/L. Negative results were observed for anti-nuclear, anti-extractable nuclear, anti-DNA, and anti-phospholipid. mNGS: Pathogenic microorganisms were not detected.	Clinical pharmacists recommend: iv methyl-prednisolone 1g/d Cyclophosphamide 0.2g/d	Blood routine examination of various indicators	No improvement	EGPA
January 19–25, 2021	The patient's state of consciousness improved and the headache was obvious at night. Eosinophilic granulocyte count: 2.11×109/L.	Methyl-prednisolone 1g/d cyclophosphamide 0.2g/d	Blood routine examination of various indicators	Improvement	
January 26, 2021	Vital signs stable, Clear consciousness; The head CT showed that the absorption was better than that of previous. Laboratory tests reported a lower number of eosinophilic granulocyte count than before.	Methyl-prednisolone 1g/d cyclophosphamide 0.2g/d	Re-examination head CT and Chest CT; Blood routine examination of various indicators	Significant improvement	
January 27-February 8, 2021	The patient was in good spirits and had a good diet.	Oral prednisolone 1 mg/kg·d	Blood routine examination of various indicators	Significant improvement;	
February 9, 2021	No obvious positive signs were found in the specialist examination. Laboratory tests reported that eosinophils had returned to normal. And the chest CT showed that the inflammation of the right lung was more significantly absorbed compared with that at admission (Figure 7). Intracranial hematoma is more significantly absorbed than before (Figure 6).	Oral prednisolone 1 mg/kg·d	Reexamination head CT and Chest CT	Further obvious improvement; disappearance in symptoms and laboratory examination	

Abbreviations: WBC, White blood cell; Hs-CRP, Hypersensitive C-reactive protein; CT, Computed Tomography; MRI, Magnetic Resonance Imaging; mNGS:metagenomic, Next-generation Sequencing.

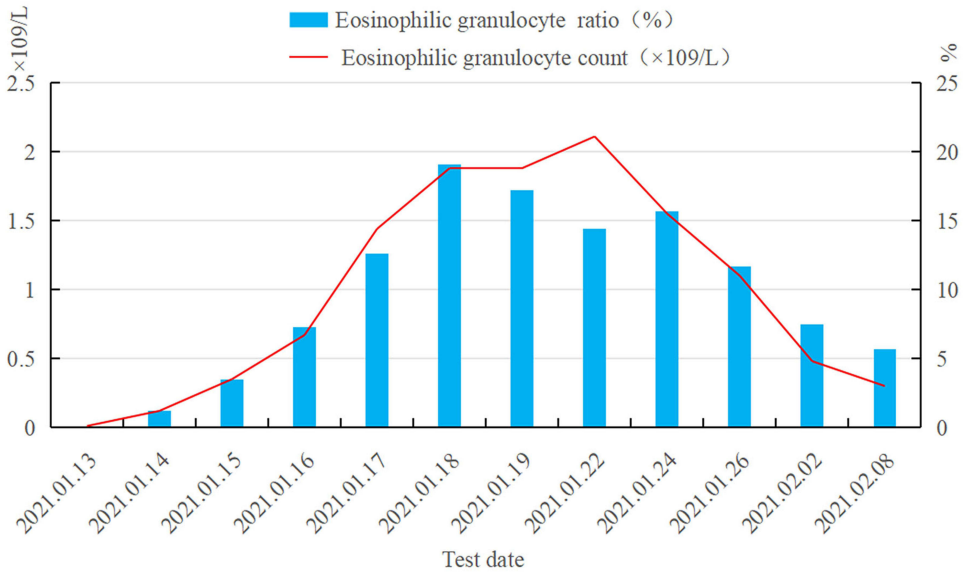

Figure 1 The eosinophil count and ratio changed with time during hospitalization.

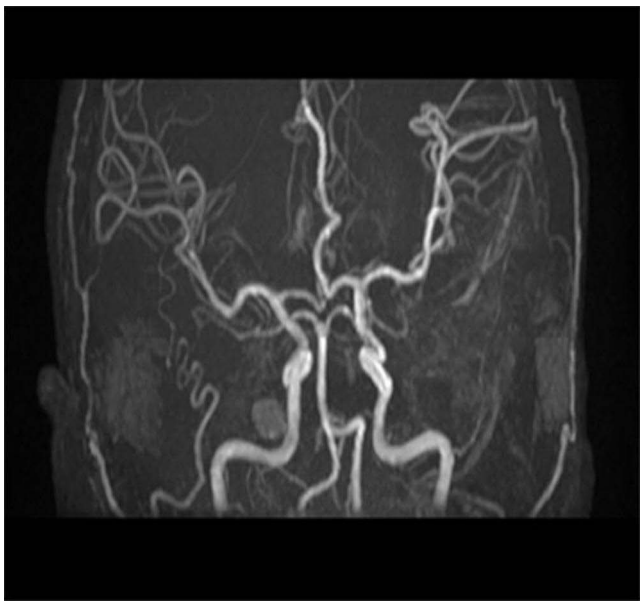

Figure 2 The Magnetic Resonance Angiography of the patient at admission. The MRA did not find vascular malformation at admission.

tract involvement of EGPA is mild, which is manifested as allergic rhinitis, nasal polyps, and transient pulmonary infiltration. Skin lesions were common in patients with EGPA (70%) and GPA (13%). EGPA is prone to invade the heart, while GPA is rare. Renal failure is rare in patients with EGPA, but GPA is common. EGPA also has a better prognosis than GPA and responds well to glucocorticoids.

Hypereosinophilic syndrome has many similarities with EGPA. Both are systemic disorders with increased peripheral blood eosinophils and tissue infiltration by eosinophils. However, the peripheral blood eosinophil count of hypereosinophilic syndrome is higher than that of EGPA. Hypereosinophilic syndrome is often accompanied by diffuse central nervous system damage, hepatosplenomegaly and systemic lymphadenopathy, thromboembolism and thrombocytopenia, while EGPA is rare. Vasculitis and granulomas are rare in the hypereosinophilic syndrome. The response to glucocorticoids was also different between the two groups, and the response to hypereosinophilic syndrome was worse.

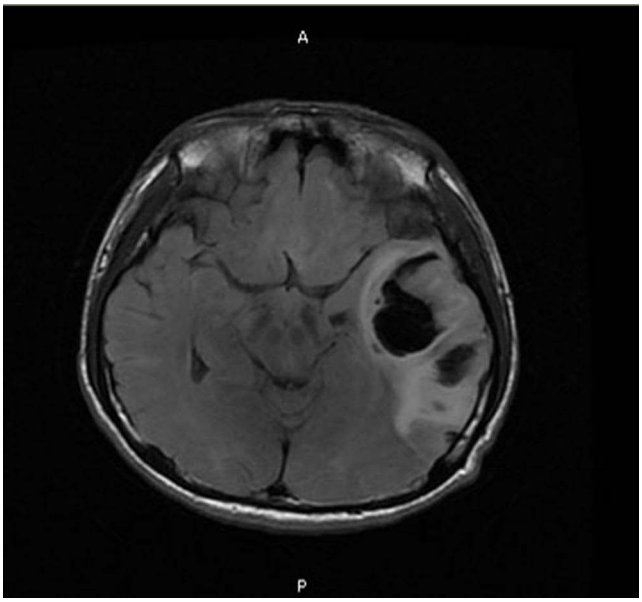

Figure 3 The Magnetic Resonance Imaging of the patient at admission. The MRI showed left frontotemporal and parietal cerebral hemorrhage.

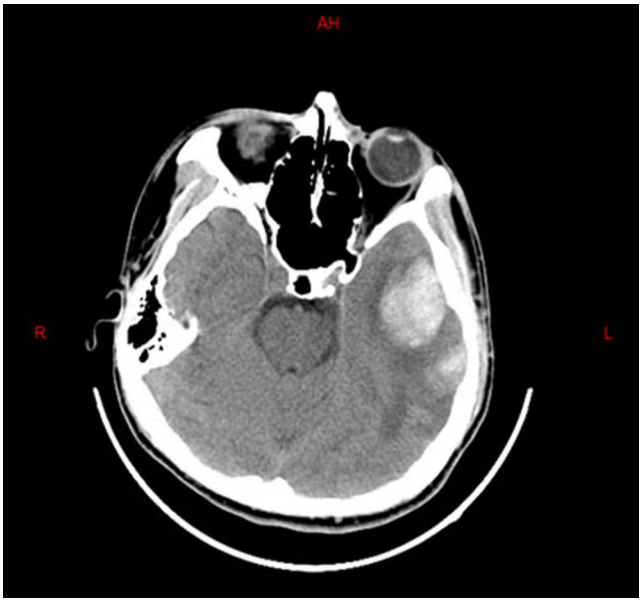

Figure 4 The heat computerized tomography of the patient at admission.

Microscopic polyarteritis (MPA) is characterized histologically by vasculitis without granulomatous inflammation.[56] Common clinical manifestations include rapidly progressive pauci-immune glomerulonephritis and alveolar hemorrhage. MPA is most commonly associated with perinuclear ANCA and antibodies to myeloperoxidase.

The exact pathogenesis of EGPA remains unclear. The possible pathogenesis is that particulate matter and cytokines released by eosinophils, such as eosinophil cationic protein and major basic protein (MBP), cause vascular injury and tissue damage.[1,57] In addition to eosinophil action,[58] it may also be the result of interaction between B and T lymphocytes. This implies EGPA symptoms induced by autoimmunity acting on various tissues of the self.

In Table 2, we reviewed all reported cases of EGPA with ICH. A total of 39 articles reported 40 patients with EGPA complicated with cerebral hemorrhage. The patients had a mean age of 34 and shared clinical features of sinus abnormalities, asthma, elevated eosinophils, and lung involvement. Based on the literature of EGPA-related intracerebral

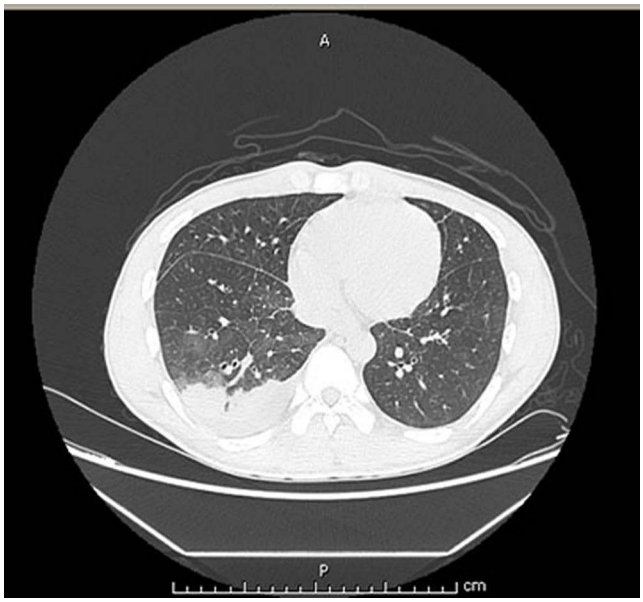

Figure 5 The chest computerized tomography of the patient at admission.

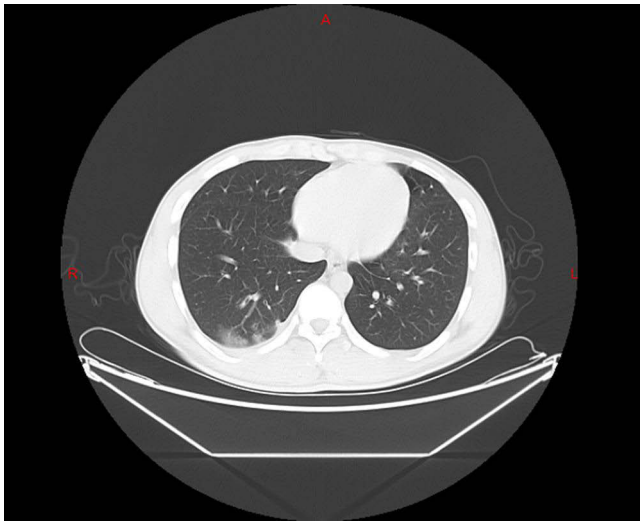

Figure 6 The chest computerized tomography of the patient at discharge.

hemorrhage, it was found that intracerebral hemorrhage was almost always preceded by prolonged asthma and atopic reactions. Twenty-five percent of these patients developed vascular structural abnormalities through MRA. The treatment is the combination of glucocorticoid and immunosuppressant.

First of all, the eosinophil count of the case reported was 2.11×109/L, which was more than 10 times the normal value. However, on admission, the eosinophil count was 0.00×109/L and less than 0.2×109/L. After the third day of admission, eosinophils continued to increase and exceeded the normal range. The possible cause for the result is that the patient had received low-dose glucocorticoids before the hospital admission. Besides, the MRI showed hypertrophy of the left inferior turbinate, right maxillary sinus cyst, and sphenoid sinus hypertrophy, which indicated a past history of allergic rhinitis and asthma. Thirdly, the cerebrospinal fluid testing contained a large Eosinophilic granulocyte infiltrate. In addition, the patient had a history of headache. Moreover, the chest CT scan showed non-fixed pulmonary infiltration. Brain magnetic resonance imaging and magnetic resonance angiography conducted showed no obvious evidence of aneurysm, artery dissection, or vascular spasm. Anti-nuclear antibody, anti-extractable nuclear antigen antibody,

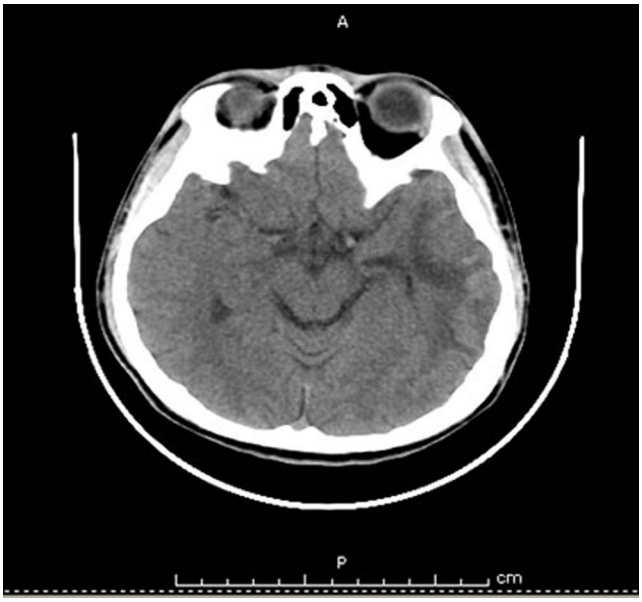

Figure 7 The heat computerized tomography of the patient at discharge.

myeloperoxidase-ANCA, proteinase- 3-ANCA, anti-cardiolipin antibody were all normal. No pathogenic microorganisms were detected by mNGS. Therefore, according to the diagnostic criteria of ACR in 1990 and Chapel Hill consensus in 2012, the patient was comprehensively diagnosed as EGPA with following diagnostic basis: allergic rhinitis, asthma,

Table 2 Clinical Features of Previously Reported Patients with EGPA and Intracerebral Hemorrhage

Literature	Age	Sex	Site of Hemo rrhage	MRI/CT	Angiography	Eos	SI	PI	Treatment
Maloon A et al, 1985[16]	39	M	SAH	+	Vasculitis	+	BA, PSA, Skin	+	SD: iv PSL, Oral PDS; ID: iv CPM
Chang et al, 1993[17]	47	F	SAH	+	NR	+	BA, PNS, PSA	+	SD: iv PSL, Oral PDS; ID: iv CPM
Liou et al, 1997[18]	27	M	ICH	+	NR	+	BA, PNS, PSA, AR	+	SD: iv MP; Oral PDS; ID: iv CPM
Muraishi K et al, 1998[19]	29	F	SAH	+	Vasculitis	+	NR	NR	SD
Ojeda et al, 2001[20]	48	M	ICH	NR	NR	+	BA, PSA	+	SD: Oral PDS; ID: iv CPM
Calvo-Romero et al, 2002[21]	47	F	SAH	–	Vasculitis	+	BA, PNS, PSA, Skin	–	SD: Oral PDS; ID: iv CPM
Tyvaert et al, 2004[22]	47	F	SAH	+	Vasculitis	+	BA, PNS, PSA, Skin	–	SD: iv MP; Oral PDS; ID: iv CPM
Sakamoto et al, 2005[23]	36	F	SAH	+	Vasculitis, Aneurysm	+	BA, PNS, PSA	–	SD: Oral PDS
Mishra et al, 2007[24]	45	M	SAH, ICH	+	Normal	+	BA, PNS, PSA, AR, Skin	–	SD: Oral PDS; ID: iv CPM
Sheerin et al, 2008[25]	37	F	SAH	+	Vasculitis	+	NR	–	SD: iv MP
Nam et al, 2009[26]	32	M	ICH	+	Normal	+	BA, PNS, PSA, AR	+	SD: Oral PDS; ID: iv CPM
Mencacci et al, 2011[27]	29	M	ICH	+	Normal	+	BA, PNS, PSA, AR, Skin	–	SD: IV MP, Oral PDS
Shimizu Ket al, 2011[28]	60	F	SAH	+	Normal	+	PNS, Arthritis	+	SD: iv PSL; ID: Oral CsA
Halliday et al, 2012[29]	43	M	ICH	+	NR	+	BA, PNS, PSA	+	SD: iv PSL, Oral PDS; ID: iv CPM
Myeong Hoon Go et al, 2012[30]	39	M	SAH	+	IVAD	+	BA, PNS, PSA, Skin, Arthritis	+	SD: iv MP 1 mg/kg; IA: iv CPM
	46	M	ICH	+	Normal	+	PSA, AR	–	iv MP, Oral PDS
Menditto VG et al, 2012[31]	64	F	SAH	+	Aneurysm	+	Skin	NR	Oral PDS
Ito M et al, 2014[32]	68	M	SAH	+	IVID	+	PNS, Arthritis	NR	SD
Taormina G et al, 2014[33]	58	M	SAH	+	Normal	+	BA, PNS, PSA, Skin, AR	+	Oral PDS
Diamanti L et al, 2014[34]	31	F	SAH	+	NR	+	PNS, Skin, Arthritis	NR	iv MP
Sylvain L et al, 2015[35]	43	M	SAH	+	Normal	+	BA, PNS, PSA, Arthritis	+	SD: iv MP; ID: CPM
Sharma SR et al, 2016[36]	6	F	LPLH	+	NR	+	BA, AR, PSA, PNS	+	SD: iv MP; ID: iv CPM
Ullah Z et al, 2016[37]	57	M	BGH	+	Normal	+	BA, SPA, PNS	–	iv MP, Oral PDS
Mattsson G et al, 2017[38]	53	M	ICH	+	NR	+	BA, SPA	NR	iv MP, Oral PDS
Lee MXW et al, 2017[39]	48	F	SAH	+	Aneurysm	+	PNS, Skin	NR	SD: iv MP; ID: CPM

(*Continued*)

Table 2 (Continued).

Literature	Age	Sex	Site of Hemo rrhage	MRI/CT	Angiography	Eos	SI	PI	Treatment
Matsuda S et al, 2018[40]	48	F	SAH	+	NR	+	PNS, Skin, Arthritis	NR	NR
Yamada Y et al, 2018[41]	42	F	SAH, MH	+	NR	+	BA, AR, PNS	+	SD: iv MP; Oral PDS
Hira K et al, 2019[42]	78	F	NR	+	NR	+	SPA	+	SD: iv MP, Oral PDS; ID: CPM
Southam C et al, 2019[43]	56	M	SAH	+	NR	+	PNS	NR	iv MP
Mrackova J et al, 2020[44]	54	M	SAH	+	NR	+	BA, PNS, Skin	NR	NR
Lázaro Romero A et al, 2021[45]	52	F	SAH	+	NR	+	BA, PSA	+	Oral PDS; iv CPM
Burtson KM et al, 2021[46]	41	F	NR	+	NR	+	SPA	+	SD: iv MP
Frikha F et al, 2022[47]	24	M	IVH	+	Normal	+	BA	–	SD: iv MP, Oral PDS; ID: iv CPM
Mino T et al, 2022[48]	69	F	SAH	+	Normal	+	BA, SPA	–	Oral PDS
Mutoh T et al, 2022[49]	45	F	PH	+	Normal	+	BA, SPA, PNS	+	SD: iv MP, Oral PDS; ID: iv CPM
Guerra M et al, 2023[50]	60	M	NR	NR	NR	+	BA, AR, PNS	+	Oral PDS
Mahmood K et al, 2023[51]	72	F	NR	+	NR	+	PSA	+	SD: iv MP; ID: iv CPM
Satake Y et al, 2024[52]	39	M	SAH	+	Normal	+	BA, AR, SPA, Skin	–	SD: iv MP
Nie N et al, 2024[53]	10	F	NR	+	Normal	+	BA, SPA, Skin	+	SD: iv MP; ID: iv CPM
Li G et al, 2024[54]	55	F	ICH	+	Normal	+	BA, SPA, PNS	+	iv MP

Abbreviations: MRI, Magnetic Resonance Imaging; CT:Computed Tomography;SI:Systemic involvement;PI, Pulmonary Involvement; Eos, Eosinophilia; SAH, Subarachnoid hemorrhage; ICH, Intracerebral Hemorrhage; IVH, Intraventricular Hemorrhage; PLH, Parietal Lobe Hemorrhage; PLH, Parietal Lobe Hemorrhage; BGH, Basal ganglia Hemorrhage; PH, Putaminal Hemorrhage; SD, Steroid drugs; MP, Methylprednisolone; PSL, Prednisolone; PDS, Prednisone; AR, Allergic rhinitis;PSA, Paranasal sinus abnormality; IVAD, Intracranial vertebral artery dissection; ID, Immunosuppressive drugs; PNS, Peripheral Nervous System; CPM, Cyclophosphamide; CsA, Ciclosporin;BA, Bronchial asthma; NR, Not reported.

peripheral blood eosinophilia (>10%), paranasal sinusitis, transient pulmonary infiltration and eosinophil infiltration of the cerebrospinal fluid. The ACR and the European Alliance of Associations for Rheumatology have published a new EGPA diagnostic criteria that it had a sensitivity of 85% and a specificity of 99%.[59]

EGPA is generally treated with medication. Prednisone 1–2 mg/(kg·d) was used for patients with mild disease. Methylprednisolone 1.0g/d (15 mg/kg) was used for 3 days, then oral prednisone 1–2 mg/(kg·d) was used for patients with rapid disease progression and vital organ involvement. In patients with major organ dysfunction (kidney damage, mononeuritis multiplex), immunosuppressive agents should be added to reduce or prevent irreversible organ damage. Immunosuppressant is mostly cyclophosphamide, generally intravenous infusion. After 6 to 12 weeks, the symptoms are relieved, the hormone should be slowly reduced, and the hormone therapy should be discontinued for one to two years. Prednisone, methylprednisolone, methotrexate, and cyclophosphamide were the most commonly prescribed drugs.

Conclusion

EGPA is rarely associated with intracerebral hemorrhage. For the cases of EGPA complicated with cerebral hemorrhage, it should be diagnosed as early as possible. The systemic symptoms of patient should be investigated after excluding other possible causes of ICH. Evaluation and recognition of EGPA and adequate intervention in the early are the key to treatment and the basis of drug therapy. Timely identification and management can reduce mortality and morbidity. EGPA is treated with corticosteroids and immunosuppressants. Corticosteroids are beneficial for controlling eosinophilia, inflammation, and symptoms of asthma. Immunosuppressive drugs deal with the autoimmune factors, which play an etiological role in the progression of the disease.

Abbreviations

MRI, Magnetic Resonance Imaging; MRA, Magnetic Resonance Angiography; CT, Computed Tomography; MRV, Magnetic Resonance Venography; EGPA, Eosinophilic Granulomatosis with Polyangiitis; SWI, Susceptibility Weighted Imaging; ACR, American College of Rheumatology; DWI, diffusion-weighted imaging; WBC, white blood cell; SI, Systemic involvement; PCT, procalcitonin; PI, Pulmonary Involvement; Hs-CRP, Hypersensitive C-reactive protein; SAH, Subarachnoid hemorrhage; CRP, C-reactive protein; ICH, Intracerebral Hemorrhage; PCT, Procalcitonin; IVH,

Intraventricular Hemorrhage; EOS, Eosinophilia; PLH, Parietal Lobe Hemorrhage; mNGS, metagenomic Next-generation Sequencing; BGH, Basal ganglia Hemorrhage; IVAD, Intracranial vertebral artery dissection; PH, Putaminal Hemorrhage; ID, Immunosuppressive drugs; SD, Steroid drugs; PNS, Peripheral Nervous System; MP, Methylprednisolone; CPM, Cyclophosphamide; PSL, Prednisolone; CsA, Ciclosporin; PDS, Prednisone; BA, Bronchial asthma; PSA, Paranasal sinus abnormality; NR, Not reported; AR, Allergic rhinitis.

Ethics Approval and Consent for Publication

This case was approved by the ethics committee of Inner Mongolia Medical University Ordos School of Clinical Medicine. The case details were agreed to publish by the Ordos Central Hospital. Written and signed consent was obtained from patients to publish her clinical history.

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Author Contributions

All authors made a significant contribution to the work reported. Pu Bai contributed to this manuscript as follows: conception, study design, execution, acquisition of data, analysis and interpretation, Funding acquisition, Writing – original draft and Writing – review & editing. Pu Bai have written the article and substantially revised the article. Peitao Xie contributed to this manuscript as follows: conception, study design, execution, acquisition of data and interpretation. All authors of manuscript have agreed to take responsibility and be accountable for the contents of the article and agreed on the journal to which the article will be submitted. All authors of manuscript have reviewed and agreed on all versions of the article before submission, during revision, the final version accepted for publication, and any significant changes introduced at the proofing stage.

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Disclosure

The authors declare no conflicts of interest.

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