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REVIEW

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Massive pulmonary thromboembolism in a pediatric patient with eosinophilic granulomatosis with polyangiitis: a case-based review emphasizing management

Batuhan Küçükali[1*], Merve Yazol[2], Çisem Yıldız[1], Büşra Acun[1], Nuran Belder[1], Nihal Karaçayır[1], Merve Kutlar[1], Pelin Esmeray Şenol[1], Zühre Kaya[3], Deniz Gezgin Yıldırım[1] and Sevcan A. Bakkaloğlu[1]

Abstract

Background Pediatric patients with Eosinophilic Granulomatosis with Polyangiitis (EGPA) are at an increased risk of arterial and venous thromboembolism (AVTE). Although the exact mechanisms underlying AVTE remain unclear, eosinophils play a pivotal role in AVTE.

Main body Current guidelines lack evidence-based recommendations, particularly concerning anticoagulant and antiplatelet treatments for this condition. Herein, we document a pediatric EGPA patient with deep venous thrombosis presenting with massive pulmonary thromboembolism during a relapse, treated with immunosuppressive and anticoagulant therapy to raise awareness among clinicians. Additionally, we performed a literature review to highlight various aspects of pediatric AVTE. Moreover, we evaluated the management strategies employed for the patients identified in the literature review and summarized the current practice guidelines regarding pediatric EGPA patients with AVTE to provide recommendations to clinicians on the management of this challenging complication.

Conclusions Most AVTE events occur during periods of high disease activity. Notably, EGPA patients with VTE often present with thrombocytopenia due to consumption, a finding not typically expected during disease exacerbation. Venous thrombosis generally requires both anticoagulation and immunosuppressive treatment. Although our review indicates a favorable prognosis for AVTE, the small number of reported cases prevents us from drawing definitive conclusions. Future studies should explore the efficacy of mepolizumab and other eosinophil-targeted therapies for AVTE, in addition to investigating the roles of anticoagulation and antiplatelet treatments.

Keywords EGPA, Thromboembolism, Pulmonary embolism, Churg-Strauss Syndrome, Eosinophilic Granulomatosis with Polyangiitis

*Correspondence:
Batuhan Küçükali
Batuhan.kucukali@gazi.edu.tr
[1] Department of Pediatric Rheumatology, Faculty of Medicine, Gazi University, Ankara, Besevler 06500, Turkey
[2] Department of Pediatric Radiology, Faculty of Medicine, Gazi University, Ankara, Turkey
[3] Department of Pediatric Hematology, Faculty of Medicine, Gazi University, Ankara, Turkey

Introduction

Eosinophilic granulomatosis with polyangiitis (EGPA), previously known as Churg-Strauss syndrome, is a rare vasculitis affecting small to medium vessels, histologically characterized by tissue eosinophilia, necrotizing vasculitis, and eosinophil-rich granulomatous inflammation [1, 2]. EGPA is extremely rare in children, with the largest case series only reaching fourteen patients [3–6].

The clinical manifestations of EGPA may be divided into three phases: a prodromal allergic phase characterized by a history of asthma and allergic rhinitis, an eosinophilic phase with peripheral eosinophilia resulting in cardiomyopathy and gastroenteritis, and a vasculitic phase causing glomerulonephritis, peripheral neuropathy, and purpuric skin lesion [1]. However, these phases are not strictly separated [1]. Furthermore, the presence of anti-neutrophil cytoplasmic antibody (ANCA) is particularly associated with vasculitic findings, while its absence is associated with eosinophilic findings [1]. ANCA is detectable in 40% of adult cases, usually against myeloperoxidase (MPO) [1, 5]. In 2022, the American College of Rheumatology (ACR) and the European Alliance of Associations for Rheumatology (EULAR) introduced classification criteria for EGPA [7]. However, these criteria are not specific for children [1]. Additionally, the clinical findings in children with EGPA differ from those in adults, with children exhibiting higher rates of pulmonary infiltrates and cardiac disease, while adults show higher rates of mononeuritis multiplex and myalgia [5].

Considering the rarity of the disease and the heterogeneity in clinical findings, the diagnosis and management of EGPA present significant challenges for clinicians, especially with pediatric patients. The ACR introduced management recommendations in 2021, and EULAR in 2022 [8, 9]. However, there are no recommendations specifically targeting pediatric patients, as their clinical manifestations and treatments may differ [1, 5]. Furthermore, the recommendations did not address the management of thromboembolic events, despite the evidence that patients with ANCA-associated vasculitis (AAV) have an increased risk of these events [10, 11]. Herein, we describe a pediatric EGPA patient presenting with disease flare and massive pulmonary thromboembolism, and review the current literature, highlighting this significant condition and the treatment modalities to supplement the guidelines.

Case presentation

A 17-year-old male patient previously diagnosed with EGPA presented to our clinic with shortness of breath, palpitations, and malaise for two days. The patient denied any fever, sputum, cough, chest pain, or foreign body aspiration.

The patient was diagnosed with EGPA three years ago. He initially presented with bloody sputum, and further evaluations revealed pulmonary consolidations with a halo sign. In his medical history, he had been treated for asthma and allergic rhinitis for nine years. The patient's family history was unremarkable. Peripheral eosinophilia, p-ANCA positivity, proteinuria, and renal biopsy exhibiting pauci-immune focal necrotizing crescentic glomerulonephritis led to EGPA diagnosis according to ACR/EULAR classification criteria [7]. He received pulse corticosteroids (CS), cyclophosphamide (CYC), and rituximab during induction treatment due to his life-threatening condition. During the induction treatment, the patient experienced right popliteal vein thrombosis without any evident hematological or anatomical underlying cause. This was managed with subcutaneous low-molecular-weight heparin (LMWH) at a dose of 1 mg/kg twice daily for six months. Consequently, the management was challenging due to frequent relapses, and the patient could not access healthcare for three months due to the 6th of February-Türkiye earthquake, one of the most devastating ones [12, 13]. The patient presented to our clinic for follow-ups after the earthquake. The patient's management has been summarized in Fig. 1.

Physical examination yielded a 1 cm difference between his calves due to right popliteal venous thrombosis in 2021, and tachycardia and tachypnea without abnormal heart and lung auscultation findings. Initial differential diagnoses included infections such as pneumonia and myocarditis due to the immunocompromised status of the patient, disease flares with cardiac or pulmonary involvement and pulmonary embolism (PE).

The initial laboratory tests resulted in leukocytosis (16.7×10^9/L), mild thrombocytopenia (120×10^9/L), elevation of CRP (33 mg/dL, normal range: 0–5), high-specific troponin T (40 ng/L, normal range: 0–14) and excessively elevated D-dimer (6.69 µg/mL, normal range: 0–0.5) levels. Electrocardiography, echocardiography, and arterial blood gases were within normal range. Additionally, the chest X-ray did not reveal any pathological findings. The patient underwent pulmonary computed tomography angiography, which revealed massive pulmonary thromboembolism, as shown in Fig. 2.

The patient was managed with subcutaneous LMWH 1 mg/kg twice daily, with dose adjustments planned according to anti-factor Xa levels to be maintained between 0.5–1 IU/mL. Doppler ultrasonography of lower extremity veins revealed right distal femoral and popliteal vein thromboses. Infectious screening did not identify any microorganisms or PCR positivity. Additionally, anti-phospholipid antibodies and lupus anticoagulants were negative. The proteinuria and hematuria were significantly increased compared to the last visit (from 24 mg/m^2/hour to 64 mg/m^2/hour). The disease flare-up was evident with severe renal involvement. Methylprednisolone at 1000 mg daily for three days and CYC at a dose of 500 mg every two weeks for six doses were initiated. Additionally, compression stockings were utilized during his inpatient clinic treatment.

The patient responded to the treatment excellently. All of his symptoms subsided, and D-dimer levels decreased

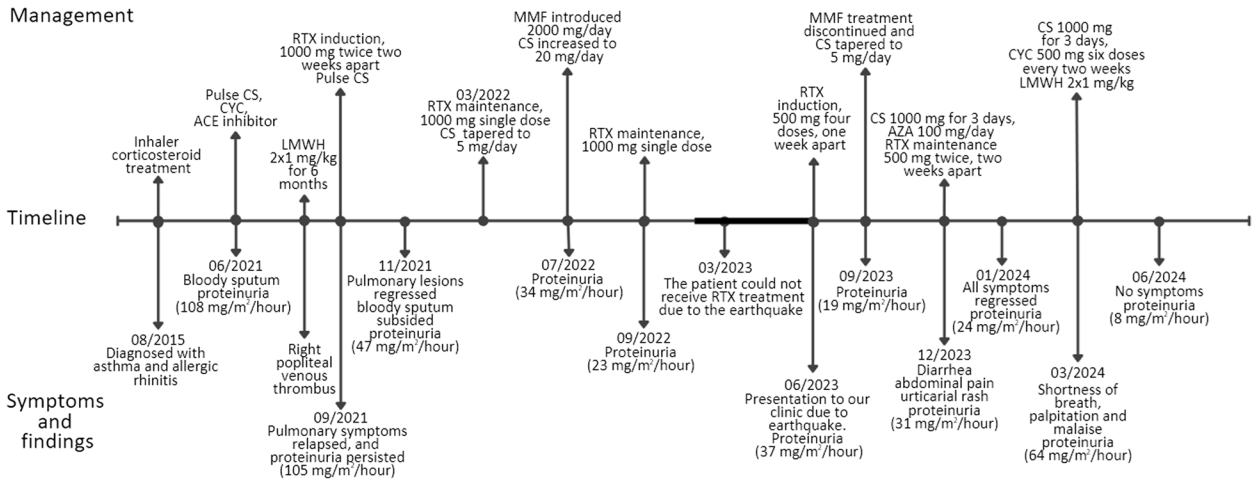

Fig. 1 The patient's clinical progress and management. CS: Corticosteroid, CYC: Cyclophosphamide, ACE: Angiotensin converting enzyme, LMWH: Low-molecular weight heparin, RTX: Rituximab, MMF: Mycophenolate mofetil, AZA: Azathioprine

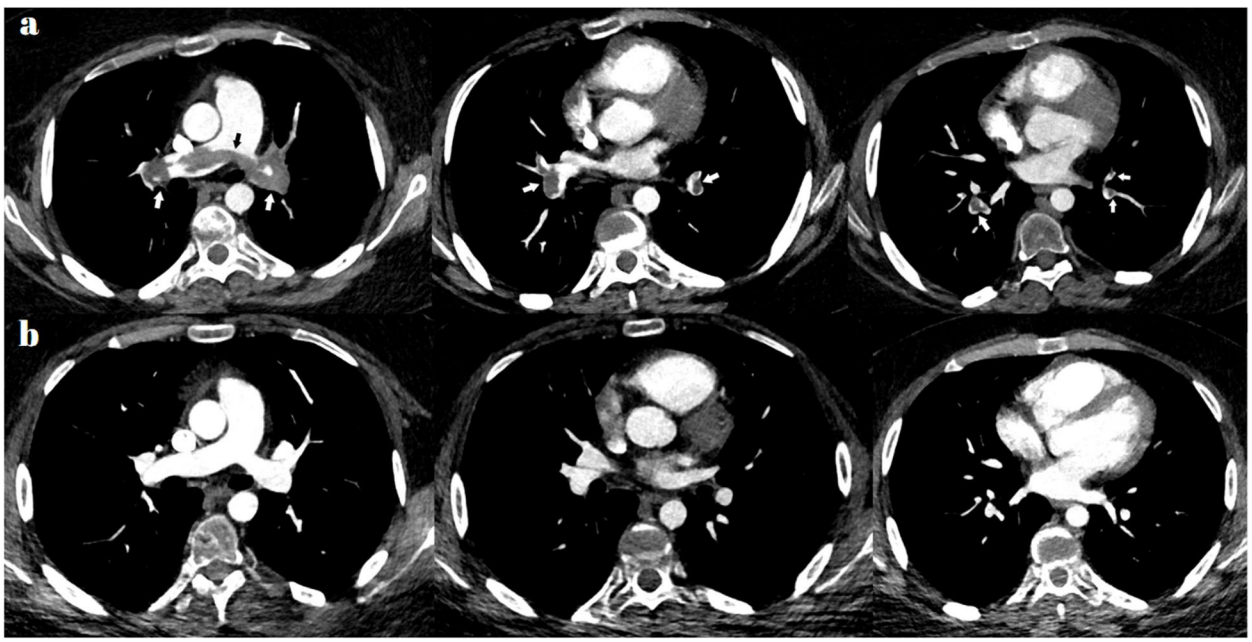

Fig. 2 **a** On pulmonary CT angiography, filling defects (white arrows) extending from the pulmonary trunk to the proximal parts of both pulmonary arteries in a saddle shape (black arrow) and continuing to the segmentary and subsegmentary branches (white arrows) at lower levels. **b** In follow-up CT examination, it is noteworthy that the thrombus and filling defects in the pulmonary artery and its branches have disappeared

significantly by the third day of the anticoagulant treatment. The patient had no symptoms three months after the treatment. Moreover, the proteinuria decreased to 8 mg/m^2/hour. We planned to switch his LMWH treatment to rivaroxaban and taper the CS dose from 10 mg to 7.5 mg daily, and transition to maintenance treatment with rituximab and avacopan.

Search strategy

We performed a search on PubMed Medline using the following keywords: ((EGPA) OR (Eosinophilic granulomatosis with polyangiitis) OR (Churg-Strauss) OR (Churg Strauss)) AND ((embolism) OR (thrombus) OR (thromboembolism) OR (emboli) OR (infarct) OR (thrombosis)) and reviewed the current English literature from inception to July 2024, regarding pediatric EGPA cases with thromboembolism. In addition,

reference lists of the initially found reports were hand-searched to identify additional relevant reports. Age, gender, clinical findings, treatment modalities, treatment response, and outcomes of patients were recorded for each patient. We summarized the search strategy in Fig. 3.

Discussion

We documented a pediatric patient with EGPA who experienced deep vein thrombosis (DVT) of the lower limb during induction treatment and massive pulmonary thromboembolism during a relapse to highlight the high risk and significance of thromboembolic events in pediatric EGPA patients and raise awareness among clinicians.

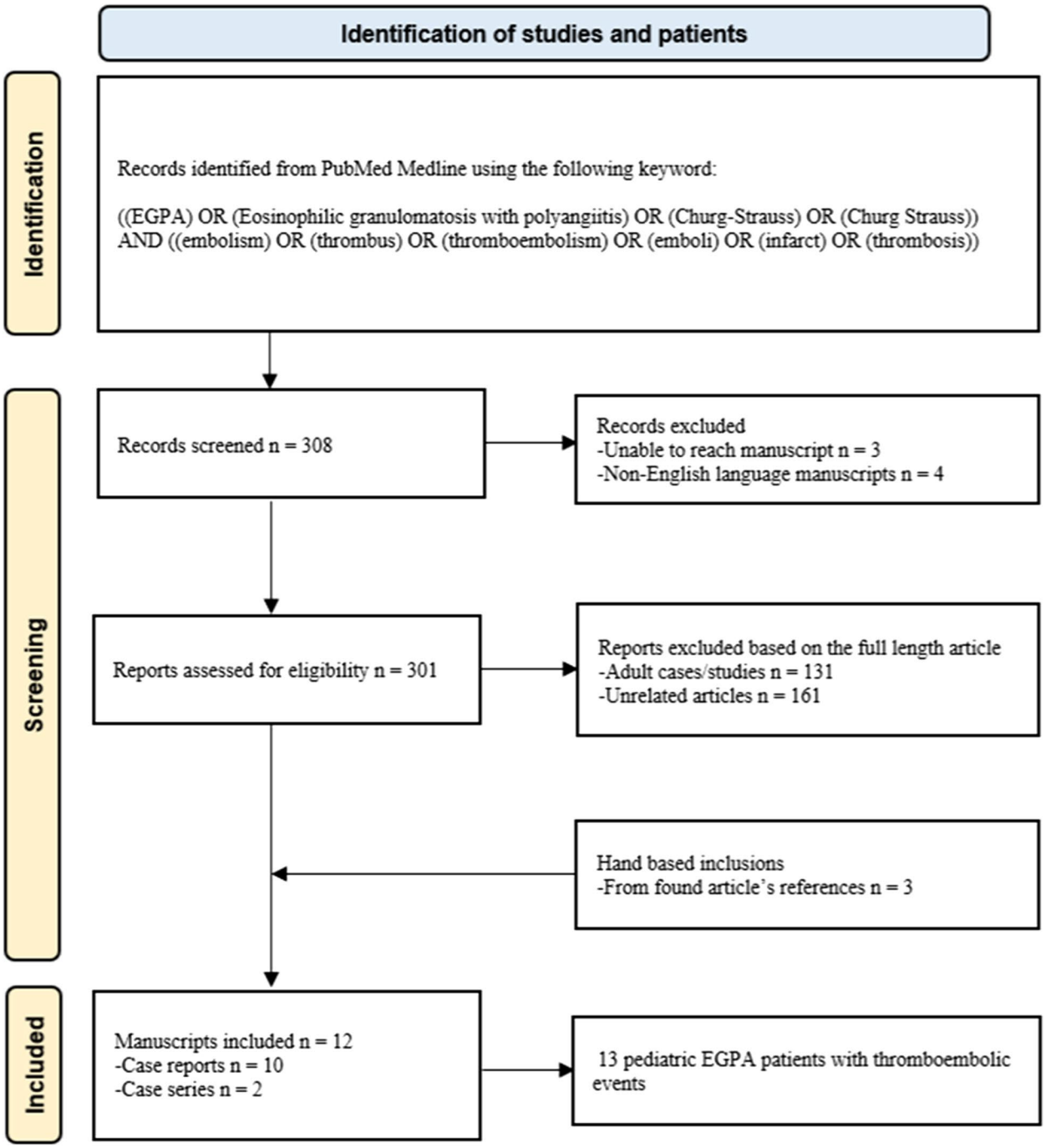

Fig. 3 Search strategy for literature review

In the literature search, we identified 13 pediatric EGPA patients with thromboembolic events. The treatment modalities varied between centers. Therefore, we summarized treatment choices and offered recommendations on the management of thromboembolic events in pediatric EGPA.

A high risk of arterial and venous thromboembolic events (AVTE) in EGPA patients have been shown by various authors [11, 14, 15]. Furthermore, some experts have shown that EGPA causes the highest risk of venous thromboembolism (VTE) and pulmonary embolism (PE) among other subtypes of AAV [15]. Hypothesis explaining the mechanisms of increased risk of thrombosis include endothelial injury due to vasculitis, defective fibrinolytic activity, hypereosinophilia and eosinophil extracellular traps and immunothrombosis, presence of antiphospholipid antibodies, and renal disease leading to the loss of anti-thrombotic factors [16–18]. There are numerous molecular pathways that are required to be elucidated to conclude the exact pathophysiological mechanism leading to thrombosis. However, eosinophils play a pivotal role in the pathogenesis of EGPA, differentiating it from other AAV subtypes and vasculitides, and current findings point to hypereosinophilia as a major player in the pathogenesis of AVTE [1, 17–20]. Treatments targeting interleukin (IL)-5, including mepolizumab, have been utilized in patients with hypereosinophilic syndromes presenting with AVTE [21]. Therefore, EGPA patients with consistent hypereosinophilia presenting with AVTE may benefit from treatments targeting eosinophils, such as monoclonal antibodies against IL-5, IL-13 and IL-4 receptor alpha. Future studies should be more focused on these targeted drugs instead of other immunosuppressive agents.

There were very scarce data on pediatric EGPA patients who presented with AVTE in the literature. Clinical findings of the 14 patients were demonstrated in Table 1, and characteristics of AVTE and treatments utilized are summarized in Table 2 [3, 4, 22–31]. Notably, detailed data of three patients could not be reached [3, 26, 27]. Furthermore, disease involvements of pediatric cases with AVTE were compared with pediatric EGPA patients documented by Gendelman et al. and their literature review in Table 3 [4]. Although mortality and renal involvement rates were slightly higher in the AVTE group, no significant differences were found between the groups. However, in adult studies, skin, pulmonary, and renal involvement was associated with a higher risk of thromboembolism [10]. Our conflicting results with the literature may be due to the limited number of cases or differences between the characteristics of adulthood and childhood EGPA. High disease activity was observed in 78% of our patients, and AVTE occurred during the presentation in those patients, which is compatible with the literature [10, 32]. Clinicians should be very diligent regarding AVTE for patients with EGPA during presentation and high disease activity. Additionally, the patients are generally immobilized during their presentation or have high disease activity due to being in inpatient clinics, which is a risk factor for venous thrombosis [33]. American Society of Hematology (ASH) recommends utilization of LMWH or fondaparinux for acutely ill patients for VTE prophylaxis in adults [34]. However, ASH does not address pediatric patients [34]. Therefore, future studies should focus on the prophylactic utilization of anticoagulant treatment or compression stockings for pediatric EGPA patients with a high AVTE risk or a history of AVTE during presentation and high disease activity.

Documented cases had thrombosis in various sites of the vascular system. The most common thrombosis sites are cerebral, intracardiac, DVT of the lower limb, and pulmonary arteries. Notably, the hepatic vascular system and limb arteries can also be involved [25, 29]. Therefore, clinicians should be aware of that pediatric AVTE generally presents with stroke and dyspnea, but rare involvements may occur.

Thrombocytosis is a common presentation finding in patients with inflammation, including vasculitides. However, four out of six pediatric EGPA patients exhibited thrombocytopenia during a VTE event [25, 28, 30]. Notably, none of the arterial thrombosis patients had thrombocytopenia. Although the association of thrombocytopenia and VTE in EGPA patients was highlighted in the literature with case reports previously, the significance of thrombocytopenia was proven with the case series in our literature review [35, 36]. Furthermore, this thrombocytopenia during VTE may be due to consumption and may serve as a distinguishing feature compared to disease exacerbations, where thrombocytosis is typically expected [35]. Clinicians should promptly evaluate the patient regarding VTE if thrombocytopenia is detected in a patient with EGPA.

In our literature review, twelve out of thirteen pediatric EGPA patients received CS after being diagnosed with AVTE. Immunosuppressive treatments were utilized in eight (61%) patients. CYC was the most common treatment. Although the cause of thrombosis in EGPA involves various molecular pathways, achieving disease remission should be a top priority [16]. Therefore, initiating prompt immunosuppressive treatment is essential. Regarding immunosuppressive selection, the Five Factor Score (FFS) was designed to predict prognosis and can be used to help clinicians choose the most adequate treatment for EGPA patients [37]. However, AVTE was not in the FFS. Emmi et al. recommended adding rare but severe

Table 1 Clinical findings in pediatric EGPA patients with thromboembolic events

Patient	1	2	3	4	5	6	7	8	9	10	11	12	13	14
Year	2024	2022	2019	2008	2011	2013	2016	2008	2016	2005	2016	2016	2011	2006
Author	Our case	Doraiswamy et al.[23]	Liu et al.[29]	Ikemoto et al.[30]	Moradinejad et al.[31]	Gendelman et al.[4]	Sahin et al.[28]	Lu et al.[22]	Ananth et al.[24]	Abdulwahab et al.[25]	Eleftheriou et al.[3]	Eleftheriou et al.[3]	Kumar et al.[26]	Ledden-Klok et al.[27]
Sex	M	F	F	M	F	F	M	F	M	M	F	N/A	M	M
Age at diagnosis, years	14	14	10	13	16	17	14	13	17	13	12	N/A	13	13
Asthma	+	+	+	-	+	+	+	+	+	-	+	N/A	N/A	N/A
Pulmonary involvement	+	+	+	+	+	-	+	+	+	-	-	N/A	N/A	N/A
Renal involvement	+	-	-	-	-	+	-	-	-	-	-	N/A	N/A	N/A
Skin involvement	+	+	+	+	-	-	+	-	+	-	+	N/A	N/A	N/A
Neuropathy	-	-	+	+	-	-	-	-	+	-	-	N/A	N/A	N/A
Cardiac involvement	-	+	-	-	+	+	-	+	-	-	+	N/A	N/A	N/A
GI involvement	+	-	-	-	-	+	-	-	+	+	+	N/A	N/A	N/A
Presence of eosinophil or granulomatous inflammation on biopsy	-	N/A	+	+	-	-	+	-	+	+	+	N/A	N/A	N/A
Other involvement	Upper RT	Upper RT, Sinus mucosal polyp	MSK	MSK	Upper RT, MSK	MSK	-	Ocular	Upper RT	-	-	N/A	N/A	N/A
ANCA positivity	p-ANCA	-	-	-	c-ANCA and p-ANCA	-	-	-	-	-	-	N/A	N/A	N/A
2022 ACR/EULAR classification criteria	+	+	+	+	-	-	+	N/A	+	+	+	N/A	N/A	N/A
Lanham classification criteria	+	+	+	-	-	+	-	N/A	+	-	+	N/A	N/A	N/A

M Male, *F* Female, *N/A* Not available, *GI* Gastrointestinal, *RT* Respiratory tract, *MSK* Musculoskeletal system, *ANCA* Anti-neutrophil cytoplasmic antibody, *ACR* American College of Rheumatology, *EULAR* European Alliance of Associations for Rheumatology

Table 2 Characteristics and treatment modalities of thromboembolic events in pediatric EGPA patients

Patient	1	2	3	4	5	6	7	8	9	10	11	12	13	14
Author	**Our case**	**Doraiswamy et al.**[23]	**Liu et al.**[29]	**Ikemoto et al.**[30]	**Moradinejad et al.**[31]	**Gendelman et al.**[4]	**Sahin et al.**[28]	**Lu et al.**[22]	**Ananth et al.**[24]	**Abdulwahab et al.**[25]	**Eleftheriou et al.**[3]	**Eleftheriou et al.**[3]	**Kumar et al.**[26]	**Ledden-Klok et al.**[27]
TE event	**DVT PE**	**IC thrombus, cerebral infarct, cerebral venous thrombosis**	**Tibial Radial Ulnar Arterial thrombosis**	**DVT PE**	**Cerebral infarction**	**Cerebral infarction**	**DVT**	**IC thrombus**	**Transverse sinus thrombosis**	**Portal Hepatic splenic SMV thrombosis**	**PE**	**IC thrombus**	**Cerebral infarction**	**Aortic bifurcation IC thrombus**
Thrombocytopenia during event	+	-	-	+	-	**N/A**	+	**N/A**	-	+	**N/A**	**N/A**	**N/A**	**N/A**
Disease activity during event	**A**	**A**	**A**	**A**	**R**	**N/A**	**A**	**A**	**R**	**A**	**N/A**	**N/A**	**N/A**	**N/A**
The time between TE event and diagnosis in years	**S, 3 years**	**S**	**S**	**S**	**5 months**	**N/A**	**S**	**S**	**1**	**S**	**N/A**	**N/A**	**N/A**	**N/A**
Treatment	**LMWH Pulse CS CYC**	**Pulse CS RTX Warfarin Dabigatran (Switched)**	**Pulse CS LMWH Clopidogrel MTX**	**Heparin CS Warfarin (switched)**	**Pulse CS CYC**	**CS AZA CYC**	**Pulse CS LMWH Aspirin**	**CS Anticoagulation Surgery**	**CYC LMWH Warfarin IVIG**	**Heparin Pulse CS CYC Warfarin (switched)**	**CS MMF**	**N/A**	**CS Aspirin**	**CS Heparin Warfarin Surgery**
Outcome	**Recovered**	**Recovered, but deceased due to sepsis after 2 years**	**Recovered**	**Recovered**	**Minimal hemiparesis**	**N/A**	**Recovered**	**N/A**	**Deceased 3 days after thrombosis**	**Recovered**	**Deceased**	**N/A**	**Recovered**	**N/A**

DVT Deep vein thrombosis, *PE* Pulmonary embolism, *IC* Intracardiac, *SMV* Superior mesenteric vein, *N/A* Not available, *A* Active, *R* Remission, *S* Simultaneously, *LMWH* Low-molecular weight heparin, *CS* Corticosteroids, *CYC* Cyclophosphamide, *RTX* Rituximab, *MTX* Methotrexate, *AZA* Azathioprine, *IVIG* Intravenous immunoglobulin G, *MMF* Mycophenolate mofetil

Table 3 Comparison of clinical findings of pediatric EGPA patients with thromboembolism and all pediatric EGPA patients documented until 2013 by Gendelman et al.

	Pediatric EGPA patients with AVTE	Pediatric EGPA patients	p
Number of Patients	14	47	
Age at diagnosis, mean years	13,7	12	
Male/Female ratio	1.16:1	0.36:1	
ANCA positivity (%)	2/11 (18)	6/24 (25)	NS
Pulmonary involvement (%)	8/11 (72)	43/47 (91)	0.088
Renal involvement (%)	2/11 (18)	6/43 (13)	NS
Skin involvement (%)	7/11 (64)	31/46 (67)	NS
Neuropathy (%)	3/11 (27)	18/45 (40)	NS
Cardiac involvement (%)	5/11 (45)	21/47 (45)	NS
GI involvement (%)	5/11 (45)	22/47 (47)	NS
Mortality (%)	3/10 (27)	6/47 (13)	0.179

EGPA Eosinophilic Granulomatosis with Polyangiitis, *AVTE* Arterial or venous thromboembolism, *P P* value, *NS* Non-significant, *ANCA* Anti-neutrophil cytoplasmic antibody, *GI* Gastrointestinal

complications, including retinal artery or vein occlusion, to the FFS [1]. Therefore, clinicians should evaluate each AVTE event on a case-by-case basis, decide on the severity of the event, and initiate immunosuppressive treatment accordingly. DVT may be managed as a non-severe disease presentation, while severe arterial thrombosis, which may lead to loss of limb or sight, requires intensive remission induction treatment for severe disease, including pulse CS along with CYC or RTX [1, 8, 9].

American Society of Hematology established management guidelines regarding pediatric VTE in 2018 [38]. According to these guidelines, patients with cerebral sino/venous thrombosis should receive anticoagulation but not thrombolytics. Furthermore, pediatric patients with symptomatic DVT and PE should receive anticoagulation, while asymptomatic patients may or may not receive treatment since adult data suggest that treatment is not required for most asymptomatic patients [38]. However, the current opinion of most clinicians leans towards treating even asymptomatic pediatric VTE cases. Additionally, ASH recommended the utilization of thrombolysis followed by anticoagulation in pediatric patients with PE and hemodynamic compromise [38]. Our patient did not receive thrombolytics because he was hemodynamically stable, and he recovered promptly with anticoagulation. ASH did not prioritize either LMWH or vitamin K antagonists as first line treatment; both can be utilized [38]. Anticoagulation treatment was initiated in six out of seven patients during VTE. Additionally, one patient received antiplatelet treatment along with LMWH contrary to the recommendations of ASH guidelines. The duration of the anticoagulation was not addressed [1, 8, 9]. ASH recommended that patients with provoked VTE should not receive anticoagulation for longer than 3 months [38]. Although there is no data regarding the duration of anticoagulation in our VTE patients, we recommend that continuing anticoagulation at least until disease remission is achieved due to the increased risk of thrombosis during active disease. However, there is no sufficient evidence regarding the effectiveness and relapse risk reduction of anticoagulation longer than three or six months in pediatric EGPA patients with VTE and inactive disease [8].

EGPA patients with arterial thrombosis and infarctions should be treated as having vasculitic involvement and managed with immunosuppressive agents. There is no available evidence regarding antiplatelet or anticoagulant treatment for AAV patients with arterial thrombosis [39]. However, antiplatelet or anticoagulant treatment may worsen the clinical picture due to hemorrhage risk of already vulnerable inflamed arteries or aneurysms in certain vasculitides, including Behçet's disease [39–41]. Therefore, clinicians should be aware of both the possible risks and the lack of evidence of efficacy before utilizing anticoagulation or antiplatelet treatment for arterial thromboses [39]. Future studies should investigate the efficacy and risks of anticoagulation and antiplatelet treatment for arterial thrombosis in pediatric EGPA patients to establish clinical practice recommendations. In our review, intracardiac thrombus was the most common site, followed by cerebral infarcts. The clinicians utilized anticoagulation for four patients and one of them received antiplatelet along with anticoagulation, and one patient received only antiplatelet treatment. Additionally, two patients required surgical intervention out of seven arterial thrombosis patients.

If immunosuppressive treatment fails, surgical interventions may be performed for patients with thrombosis that may cause severe morbidity or mortality. Notably, surgical intervention should not be performed before immunosuppressive treatment due to the high risk of complications caused by the fragile nature of inflamed vascular structures [40].

The prognosis of AVTE in pediatric EGPA patients may depend on involved vascular system, and timely diagnosis and intervention. Out of nine patients, seven recovered completely, while one with sequela. Additionally, one patient's condition was deteriorated, and he was lost three days after the event. In our review, the prognosis of AVTE was favorable. However, the patient who recovered with sequela received prompt and intensive immunosuppression including pulse CS and CYC for cerebral infarction, yet minimal hemiparesis persisted. Therefore, establishing conclusions on the prognosis would

be ungrounded due to the retrospective nature of this review and limited number of cases.

Conclusion

Pediatric EGPA patients have an increased risk of AVTE. Eosinophils play substantial role in the pathogenesis, and most of the AVTE events occur during high disease activity. Although, there is no evidence available on the use of pharmacological or mechanical VTE prophylaxis in pediatric EGPA patients, those with high disease activity and a high risk or history of VTE may receive prophylaxis. Notably, EGPA patients with VTE exhibited thrombocytopenia, which is not typically expected in vasculitides, likely due to consumption. Venous thrombosis requires anticoagulation treatment, but there is no evidence to recommend duration, and it may be considered as a non-severe disease presentation when deciding on immunosuppressive treatment. However, arterial thrombosis, with a risk of severe morbidity and mortality, can be regarded as severe disease involvement, even though it is not included in the FFS. Future studies should focus on the efficacy of mepolizumab and other eosinophil-targeted treatments for AVTE, as well as anticoagulation and antiplatelet treatments, in pediatric EGPA patients with arterial thrombosis. Although the prognosis of AVTE showed favorable results in our review, with only one patient with minimal sequela and one loss, the retrospective nature of the study and limited number of cases do not allow us to draw definitive conclusions.

Abbreviations

EGPA	Eosinophilic granulomatosis with polyangiitis
MPO	Myeloperoxidase
ANCA	Anti-neutrophil cytoplasmic antibody
ACR	American College of Rheumatology
EULAR	European Alliance of Associations for Rheumatology
AAV	ANCA associated vasculitis
CS	Corticosteroids
LMWH	Low-molecular-weight heparin
CRP	C-reactive protein
PCR	Polymerase chain reaction
DVT	Deep vein thrombosis
AVTE	Arterial and venous thromboembolic events
IL	Interleukin
ASH	American Society of Hematology
VTE	Venous thromboembolism
CYC	Cyclophosphamide
FFS	Five Factor Score
PE	Pulmonary embolism

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Authors' contributions

Conceptualization: BK, MY; Investigation: BK, BA, ÇY, NB, MK; Methodology: BK, PEŞ, NK; Visualization: MY, ÇY; Supervision: DGY, SAB; Writing – original draft preparation: BK, ÇY; Writing – review & editing: ZK, DGY, SAB.

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Data availability

The data of the clinical case will be shared on reasonable request to the corresponding author.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written consent was obtained from the patient and the patient's family.

Competing interests

All authors declare that they have no conflict of interest.

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