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CASE REPORT

IgG4-related sclerosing cholangitis associated with essential thrombocythemia: A case report

Zhi-Nian Wu, Ru JI, Ying Xiao, Ya-Dong Wang, Cai-Yan Zhao

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Abstract

BACKGROUND

The complexity of immunoglobulin G4 (IgG4)-related diseases and their potential connection to hematologic malignancies remains unclear. This article provided a review of the diagnosis and treatment of a patient with IgG4-related sclerosing cholangitis (SC) and essential thrombocythemia (ET), along with an analysis of relevant literature to enhance comprehension of this disease.

CASE SUMMARY

A 56-year-old male was admitted to two hospitals with deteriorating jaundice and pruritus prior to hospitalization. Beyond our expectations, the patient was first diagnosed with IgG4-SC and ET with the Janus kinase 2 V617F mutation. Interestingly, the administration of acetate prednisone significantly resulted in improvements in both IgG4-SC and ET. Clinicians need to pay attention to immune disorders and inflammation as they contribute to the development of various disease phenotypes.

CONCLUSION

When IgG4-SC is suspected without histopathological evidence, diagnostic therapy and long-term regular follow-up can lead to positive treatment outcomes. Clinicians should be mindful of the potential presence of concurrent hematologic diseases in patients with immune disorders.

Key Words: Immunoglobulin G4-related sclerosing cholangitis; Essential thrombocythemia; Autoimmune pancreatitis; Janus kinase 2 mutation; Glucocorticoids; Case report

Core Tip: The case diagnosed with immunoglobulin G4 (IgG4)-related sclerosing cholangitis (SC) and essential thrombocythemia was first reported. In this article, we described the clinical features of this case and reported the diagnosis and treatment process and prognosis. The relationship between IgG4-SC and Janus kinase 2 V617F mutation diseases was analyzed and summarized by retrieving literature.

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INTRODUCTION

Prior research identified a connection between autoimmune disorders and the development of myeloproliferative neoplasms (MPNs)[1]. Immunoglobulin G4 (IgG4)-related sclerosing cholangitis (SC) was first reported in Japan, and it refers to SC of unknown cause. It is characterized by increased serum IgG4 levels and fibrosis associated with marked infiltration of local lesions by lymphocytes and IgG4-positive plasma cells[2]. Precise epidemiological data are currently lacking, and the available data were mainly obtained in Japanese studies. The overall incidence and annual prevalence of IgG4-SC in Japan are 2.1 and 0.63 per 100000 people, respectively[2]. It is challenging to distinguish IgG4-SC from primary sclerosing cholangitis (PSC) and biliary cancer in the absence of histopathologic evidence[2].

Meanwhile, essential thrombocythemia (ET) is an MPN with an annual incidence of approximately 1.2-3.0 per 100000 people[3]. It is rare for patients to experience both ET and IgG4-SC simultaneously. We experienced a patient exhibiting jaundice, pruritus, a high platelet (PLT) count, and bleeding as clinical features. This was the first reported case both domestically and internationally of concurrent IgG4-SC and ET based on a review of the literature.

CASE PRESENTATION

Chief complaints

A 56-year-old male was admitted to our hospital on September 29, 2022 with progressive jaundice and intractable pruritus persisting for 1 mo.

History of present illness

The patient experienced poor appetite, abdominal distension, and intractable pruritus but did not report abdominal pain, diarrhea, rash, hemorrhagic spots, or ecchymosis prior to hospitalization. Prior to coming to our hospital, the patient had visited Municipal hospitals on September 15, 2022 and a tertiary hospital on September 23, 2022. The results of blood tests, as shown in Table 1, revealed significantly elevated serum biomarkers including aspartate aminotransferase (AST), alanine aminotransferase (ALT), alkaline phosphatase (ALP), γ-glutamyl transpeptidase (GGT), and bilirubin. Additionally, the PLT counts in the peripheral blood were significantly higher than normal. A computed tomography scan revealed wall thickening in the cholecystic and common bile duct, along with narrowing in the common bile duct and pancreatic segment, with enlargement of the head of the pancreas, manifestation of pancreatic inflammation. Based on blood tests and imaging, clinicians diagnosed the presence of biliary space-occupying lesions. However, despite treatment, the patient's symptoms, such as jaundice and skin pruritus, did not improve.

History of past illness

The patient had no history of liver disease or hemopathy. There was also no record of taking drugs that could potentially cause liver injury in the prior 6 mo.

Personal and family history

The patient had no history of alcohol consumption, smoking, or genetic disease in the family.

Physical examination

The patient's temperature was 36.4 °C, heart rate was 72 beats/minute, and blood pressure was 134/71 mmHg. Severe jaundice was observed in the skin, mucous membrane, and sclera along with widespread scratches due to pruritus. No other regions showed any signs.

Laboratory examinations

On September 30, 2022, blood examinations were performed. The results were as follows: Total bilirubin 156.36 μmo1/L; direct bilirubin 126.66 μmo1/L; ALP 722 U/L; GGT 407 U/L; total bile acid 617 μmo1/L; and PLT counts 771 × 10^9/L. These levels were significantly elevated. Blood AST and ALT levels were mildly increased. The levels of serum pancreatic

Table 1 Main outcomes of laboratory tests at hospitalization

Variable	September 15, 2022	September 23, 2022	September 30, 2022	October 04, 2022	October 09, 2022	October 10, 2022	October 14, 2022	October 18, 2022	October 28, 2022
TBIL in μmol/L	151.79	186.78	156.36	135.41	123.79	99.15	83.87	134.00	53.00
ALP in U/L	895.2	856.2	722.0	837.0	1077.0	856.0	715.0	827.0	745.0
GGT in U/L	NA	393.3	407.0	475.0	709.0	580.0	469.0	889.0	1021.0
PLT as $\times 10^9$ /L	998	760	771	813	910	NA	804	1035	813
D-dimer in g/L	NA	NA	0.39	0.39	0.55	NA	1.45	2.33	NA
PT in s	11.4	NA	11.0	11.1	11.0	NA	10.9	9.9	NA
APTT in s	27.9	NA	42.5	35.6	42.5	NA	31.6	30.6	NA
INR	1.00	NA	0.99	1.00	0.99	NA	0.98	0.89	NA
FIB in g/L	3.68	NA	3.99	3.57	3.99	NA	3.72	3.48	NA
COL/EPI-CT in s	NA	NA	NA	NA	NA	NA	> 292	NA	NA
COL/ADP-CT in s	NA	NA	NA	NA	NA	NA	244	NA	NA
P2Y12-CT in s	NA	NA	NA	NA	NA	NA	136	NA	NA

ALP: Alkaline phosphatase; APTT: Activated partial thromboplastin time; COL/ADP-CT (s): Collagen–adenosine diphosphate closure time(s) (normal range: 62-100s); COL/EPI-CT (s): Collagen–epinephrine closure time (s) (normal range: No drugs, 82-150 s; drugs, 193-300 s); FIB: Fibrinogen; GGT: γ-glutamyl transpeptidase; INR: International normalized ratio; NA: Not available; P2Y12-CT(s): Purinergic receptor P2Y, G protein-coupled 12- closure time (s) (normal range: 106-300s); PLT: Platelet; PT: Prothrombin time; TBIL: Total bilirubin. COL/EPI-CT, COL/ADP-CT, P2Y12-CT are indicators of platelet function, and larger values indicate lower platelet function.

enzymes were in the normal range. There was no infectious evidence of hepatitis A-E virus, HIV, Epstein-Barr virus, and cytomegalovirus. The levels of serum tumor markers, including ferritin 354.74 ng/mL (normal range: 23.9-33.6 ng/mL), carbohydrate antigen 199 367.4 U/mL (normal range: 0-25 U/mL), protein induced by vitamin K absence or antagonist-II 80.54 mAU/mL (normal range: 0-40 mAU/mL), and carbohydrate antigen 125 36.5 U/mL (normal range: 0-35 U/mL) were elevated, while alpha-fetoprotein and carcinoembryonic antigen levels were normal. The outcomes of the extractable nuclear antigen polypeptide antibody spectrum, anti-neutrophil cytoplasmic antibodies, and liver-related antibodies such as anti-liver-kidney microsomal, anti-smooth muscle, anti-soluble liver antigen, and anti-mitochondrial antibodies were negative. The serum IgG 20.4 g/L (normal range: 7.51-15.6 g/L) levels were also high.

Imaging examinations

Magnetic resonance imaging revealed dilation of the intrahepatic bile duct, local narrowing, and uneven thickness of the bilateral hepatic duct, common hepatic duct, and lower segment of the common bile duct. The head of the pancreas was enlarged, and the imaging physician considered possible localized pancreatitis (Figure 1).

Further diagnostic work-up

Unfortunately, jaundice did not vanish gradually. At the same time, PLT levels progressively increased (813 × 10^9/L) starting on October 4, 2022. To prevent thrombosis development and embolism because of significantly high PLT counts, aspirin (0.1 g per day) and low-molecular-weight heparin (4250 IU q12h ih) were administered. After 4-d anticoagulation and anti-PLT therapy, ecchymosis and a lump with tenderness were found at the needle sites and the back of the left lower leg, respectively. Color doppler ultrasound showed a liquid mass in the muscle layer of the left lower leg, and there were no signs of clot or deep vein thrombosis in the legs. We considered the lump a hematoma. Therefore, aspirin and low-molecular-weight heparin were discontinued immediately.

According to the above clues, the cause of jaundice in patients may be any of the following: PSC; IgG4-SC; and tumor. Due to the patient's high risk of bleeding, a liver biopsy was not conducted. This decision complicated the process of distinguishing between PSC and a bile duct tumor, but quantitative analysis of immunoglobulin subclasses was carried out. The coagulation function examination suggested a normal or hypercoagulable state and decreased PLT function (Table 1). To determine the cause of hyperthrombocytosis and bleeding, we conducted bone marrow and genetic testing.

The outcomes of the examinations were gradually reported. Quantitative analysis of immunoglobulin subclasses: Serum IgG4 (19 g/L, normal range: 0.03-2.01 g/L) level was significantly elevated. Bone marrow cytology examination and bone marrow histopathology are depicted in Figure 2. Gene testing found that the Janus kinase 2 (*JAK2*) V617F mutation was positive.

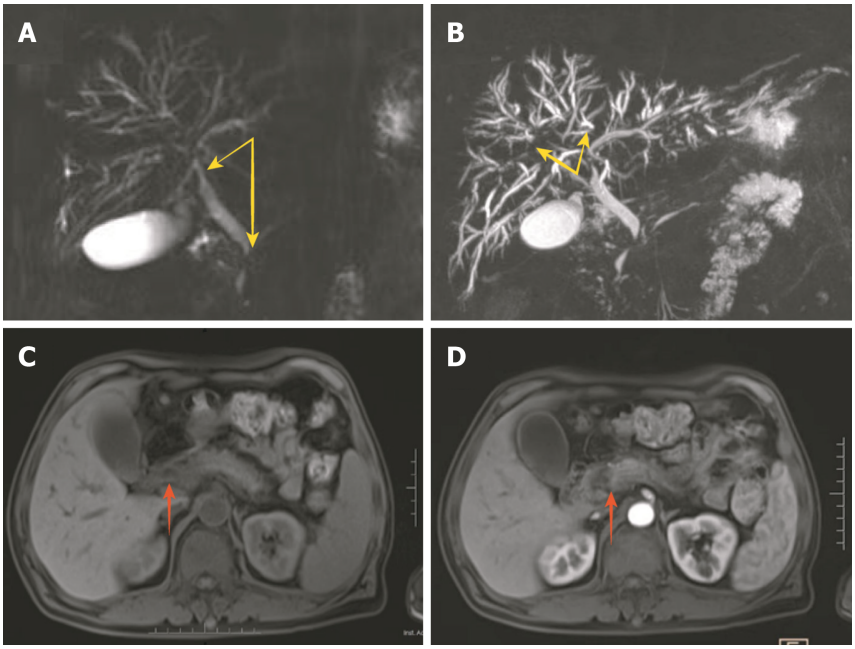

Figure 1 Magnetic resonance imaging of the bile ducts. A and B: Magnetic resonance cholangiopancreatography revealed dilation of the intrahepatic bile duct and local narrowing in the bilateral hepatic duct, common hepatic duct, and lower segment of the common bile duct (yellow arrows); C and D: Plain and enhanced magnetic resonance imaging showed an enlarged pancreatic head. Possible local inflammation was, but there was no delayed enhancement (red arrows).

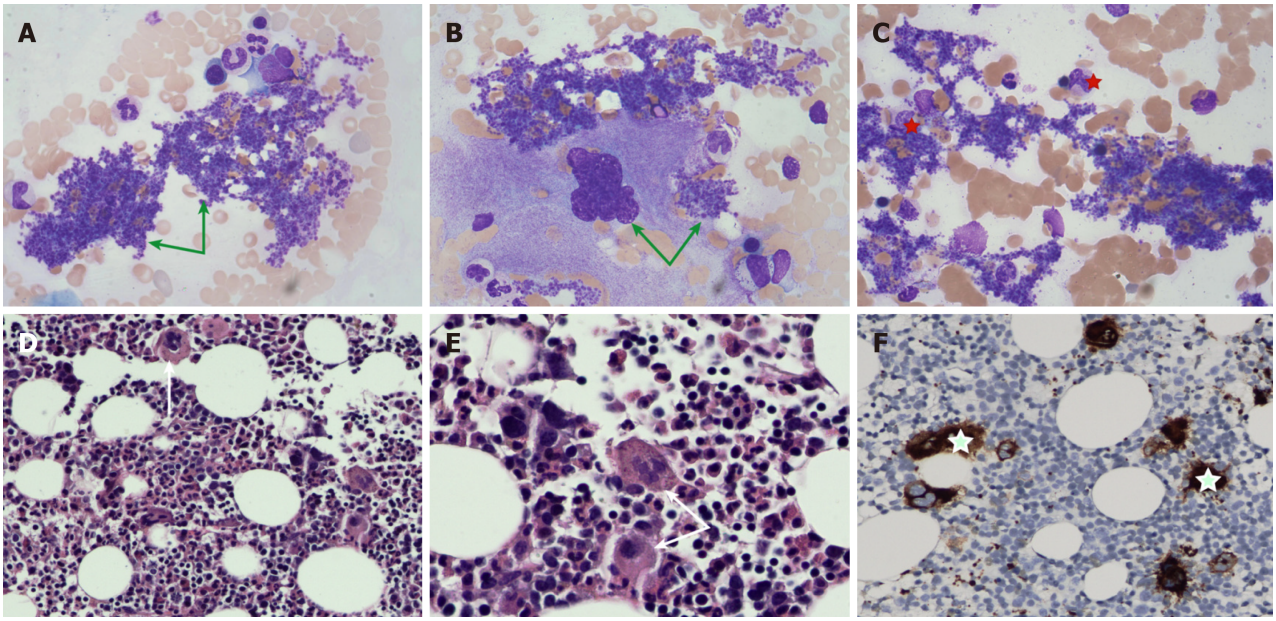

Figure 2 Bone marrow examination. A and B: Platelets existed in the form of piles and pieces in the peripheral blood smear and bone marrow smear (green arrows); C: Proplatelet-producing megakaryocytes were markedly increased in the bone marrow smear (red five-pointed star); D and E: The number of megakaryocytes was significantly elevated, especially megakaryocytes with large cell bodies and hyperlobated nuclei, and they were isolated or arranged in dense clusters (reticular fibrosis grade of MF-1) (Hematoxylin-eosin staining: 200 times, 400 times) (white arrows); F: CD61 immunohistochemistry was also positive on megakaryocytes (green five-pointed star).

FINAL DIAGNOSIS

Although pancreatic enzymes did not show significant abnormalities, localized pancreatitis was still be considered in conjunction with imaging studies conducted before and after hospital admission. Based on the significantly elevated IgG4 levels, we concluded that the patient's diagnosis was autoimmune pancreatitis. Additionally, the clinical presentation of this patient was mainly jaundice, and imaging findings showed significant bile duct abnormalities, therefore the final diagnosis included IgG4-SC[4]. ET was diagnosed in the patient in accordance with relevant guidelines (Table 2)[5].

Table 2 Criteria for diagnosis of essential thrombocythemia

Major criteria	Minor criteria
Platelet count ≥ 450000 per mm^3	Presence of clonal marker or of evidence of reactive thrombocytosis
Bone marrow biopsy showing proliferation mainly of the megakaryocytic lineage, with increased numbers of enlarged, mature megakaryocytes with hyperlobated nuclei; no substantial increase or left shift in neutrophil granulopoiesis or erythropoiesis; in rare instances, minor (grade 1) increase in reticulin fibers	
Criteria for BCR-ABL1-positive chronic myeloid leukemia, polycythemia vera, primary myelofibrosis, or other myeloid neoplasm not met; *JAK2* V617F, *CALR*, or *MPL* mutation.	

Diagnosis requires all major criteria or the first three major criteria plus a minor criterion. *CALR*: Calreticulin; *JAK2*: Janus kinase 2; *MPL*: Myeloproliferative leukemia.

TREATMENT

At admission, the patient was given symptomatic treatments, including liver protection and choleretic treatment. The patient (body weight: 65 kg) was treated with oral glucocorticoids (acetate prednisone tablets, 30 mg, once daily) combined with hydroxyurea tablets (500 mg, twice daily).

OUTCOME AND FOLLOW-UP

The patient's jaundice resolved, and he was discharged on October 20, 2022. One month after discharge, the patient stopped taking hydroxyurea tablets by himself. The dosage of corticosteroids was gradually reduced. Follow-up was conducted 7 mo later by phone. Jaundice and hematoma had subsided, and the bleeding tendency had disappeared. Bilirubin levels and PLT counts gradually return to normal (Figure 3).

DISCUSSION

IgG4-SC is an autoimmune disease closely associated with IgG4 and falls under the umbrella of IgG4-related diseases. These conditions are known for causing ongoing chronic inflammation in individuals who have not received treatment. Glucocorticoids are recommended as the first-line treatment therapy for IgG4-SC[4].

ET is a kind of MPN characterized by the proliferation of bone marrow megakaryocytes and increased PLT counts in peripheral blood[5]. The diagnosis of ET is mainly based on the diagnostic criteria proposed by Tefferi and Pardanani[5] in 2019, and this case fulfills all the diagnostic criteria. Low-dose aspirin (81-100 mg, once daily), hydroxyurea (500 mg, twice daily), or interferon alpha-2α (45 μg once a week, the maximum dose of 180 μg once a week) should be given for patients diagnosed with ET until normal PLT counts[5,6]. It is well-known that glucocorticoids promote PLT production, which will not be conducive to the treatment of ET. Therefore, whether glucocorticoids should be used in this patient became a topic to be discussed by clinicians.

To enhance our understanding of the pathogenesis of ET, we conducted an extensive literature review. Chronic inflammation is considered a prerequisite for defending against clonal evolution and cancer development due to its effective DNA repair mechanism in response to sustained oxidative stress caused by chronic inflammation[7]. It is important to note that mutations resulting from DNA repair mechanisms may also increase the risk of clonal evolution. Early studies have shown that sustained inflammation can activate JAK2 (including the V617F mutation) and lead to genome instability, increasing the risk of mutation[8]. Further, chronic inflammation has been demonstrated to be an important driver of ET[7,9]. Therefore, it was not unexpected that this patient suffered from IgG4-SC and ET.

IgG4-SC is an autoimmune disease closely related to IgG4 and is classified as one of the IgG4-related diseases. These diseases are known for causing persistent chronic inflammation in untreated individuals. Emerging evidence indicates that the pathophysiological mechanisms linked to immune inflammation could potentially affect PLT production. Kristinsson *et al*[1] confirmed that autoimmune diseases (such as immune thrombocytopenic purpura, Crohn's disease, polymyalgia rheumatica, giant cell arteritis, Reiter's syndrome, and aplastic anemia) increase the risk of MPN by 20%, especially ET[1]. PLT counts have significant variability in immune-mediated conditions, specifically, ET and immune thrombocytopenic purpura can interconvert in different immune states[10].

During follow-up of this case, the patient came to our clinic again due to weakness, and examination showed ALT 637 U/L, AST 394 U/L, total bilirubin 38.8 μmo1/L, direct bilirubin 27.6 μmo1/L, GGT 1141 U/L, ALP 1285 U/L, WBC 14.46 × 10^9/L, N% 75.4%, eosinophil 0.88 × 10^9/L, and PLT 1461 × 10^9/L (because of poverty, the patient did not receive inpatient treatment). Multiple simultaneous increases in biliary enzymes and PLT counts further supported a potential link between IgG4-SC and ET.

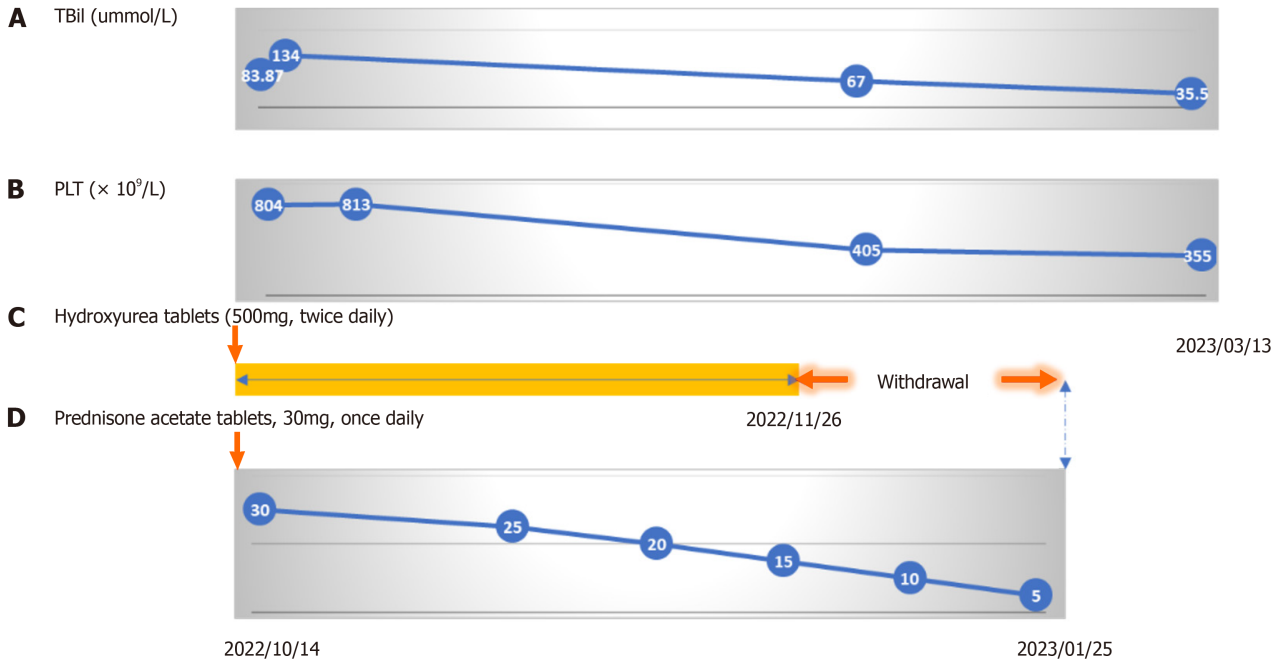

Figure 3 Follow-up procedure. A and B: Changes in total bilirubin (TBIL) and platelets (PLT) in the patient during the treatment process; C: Hydroxyurea (500 mg 2/d) was self-discontinued after 1 mo; D: Acetate prednisone tablets were reduced by 5 mg after the first 2 wk, followed by a weekly reduction of 5 mg. The patient self-discontinued the medication after 3 mo of use.

Mechanistically, aberrant B cell activation is present in both diseases[11,12]. This suggests that the presence of IgG4-SC and ET in this case is not merely a combination of the two diseases but rather distinct manifestations triggered by inflammatory and immune-related factors. Based on these theories, JAK inhibitors hold promise as alternative treatment options for IgG4-related diseases and MPNs[13,14]. Importantly, this case study can aid clinicians in expanding their diagnostic and treatment approaches.

CONCLUSION

IgG4-SC can be challenging to diagnose as it lacks specific clinical manifestations and can be mistaken for biliary malignancies, particularly when obtaining pathological examination results is difficult. Long-term follow-up and glucocorticoid treatment play a crucial role in establishing a definitive diagnosis. Furthermore, this manuscript explored the potential mechanistic connection between IgG4-SC and ET, expanding clinicians' perspectives on diagnosis and treatment strategies.

FOOTNOTES

Author contributions: Wu ZN and Xiao Y accessed the literature and wrote the case; Ji R and Zhao CY provided material support for the study; Wang YD served as the corresponding author and critically reviewed and revised the manuscript; All authors wrote the manuscript and approved this version to be submitted.

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