

Case Report

Corticosteroid-dependent chronic immune thrombocytopenic purpura in an 11-year-old child responsive to low-dose rituximab: a case report

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Abstract: Immune thrombocytopenia (ITP) is the most common platelet disorder in children. The peak incidence is observed from 1 to 7 years of age. We report the case of an 11-year-old boy who was diagnosed with ITP four years before visiting our clinic following a viral infection. He was corticosteroid-dependent and refractory to treatment after intravenous immunoglobulin (IVIG), splenectomy and vincristine administration. Thrombopoietin receptor agonists (TPO-RA) were not an option due to family socioeconomic conditions. He exhibited an excellent response to an unusual treatment modality with a low fixed dose of rituximab (100 mg weekly) over the period of 4 consecutive weeks.

Keywords: chronic immune thrombocytopenia purpura, splenectomy, corticosteroid, rituximab, intravenous immunoglobulin (IVIG), thrombopoietin.

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Introduction

Immune thrombocytopenia (ITP) is an acquired immune-mediated disorder that manifests itself as a temporary or permanent reduction in platelet count and bleeding tendency [1]. Patients have isolated thrombocytopenia (less than 100×10^9 cells/L) without any other predisposing conditions known to decrease platelet count. The condition is usually self-limited and resolves with time, although in some cases it may persist for over 12 months, becoming chronic ITP, which affects 10–20% of children: approximately 1 out of 10,000 children aged 2 to 6 years, with equal incidence in boys and girls [2]. The exact pathogenesis of ITP remains unclear. It is thought that platelets become coated with autoantibodies against platelet membrane antigens; destruction of platelets occurs mediated by T cells, while megakaryocyte function is impaired. This significantly reduces survival to a few hours [3]. Children with ITP usually have a history of a previous viral infection, which may be a possible cause of the formation of these autoantibodies. ITP manifests itself as easy bruising (purpura) or extravasation of blood from capillaries into the skin and mucous membranes (petechiae). Although most cases of acute ITP, especially in children, are mild and self-limited, intracranial hemorrhage may occur when the platelet count falls below $10^{10}/L$ ($<10^4/\mu L$) [4]. The main goal of treatment is to prevent severe bleeding and to increase platelet counts. High-dose corticosteroids, intravenous immunoglobulin (IVIG), rituximab, splenectomy, and thrombopoietin receptor agonists (TPO-RA) are treatment options that may be used as needed. In some cases, immunosuppressants are prescribed [5]. In this case report, we

present a case of chronic ITP in an 11-year-old boy who was corticosteroid-dependent for the past 4 years and was not cured by IVIG, splenectomy and vincristine administration. He exhibited an excellent response after a low fixed dose of rituximab over 4 consecutive weeks and finally stopped taking corticosteroids after long-term intake.

Case report

An 11-year-old boy was examined by us for the first time in May 2023. Following a viral infection, he was diagnosed with ITP at the age of 8 years at a peripheral medical center. The child was corticosteroid-dependent over the past 4 years. He had mild epistaxis in anamnesis but no history of severe bleeding, trauma, or joint disease. He was born by non-consanguineous parents and had two siblings, none of whom had a significant medical history. He had no family history of ITP, bleeding disorders, malignancy, or sudden death. Physical examination results were normal, and he had no lymphadenopathy or organomegaly. A complete blood count (CBC) showed a normal hemoglobin level and white blood cell count. A peripheral blood smear demonstrated normal morphology and no abnormal cells. His platelet count dropped further to less than $10 \times 10^3/dL$ when we tried to taper the prednisolone dose, and he started having bleeding symptoms of epistaxis and gingival bleeding in addition to ecchymosis and bruising on both legs. Consequently, IVIG was started and there was a rapid response; platelet count reached $800 \times 10^3/dL$ but dropped again to less than $10^4/dL$ after a week of treatment. Splenectomy was performed in June 2023 as second-line therapy

and platelet count rose to $1,000 \times 10^3/\text{dL}$. The duration of response after splenectomy was only two months. Platelet count dropped to $5 \times 10^3/\text{dL}$ in August 2023. As a third line, we decided to administer 1 mg of vincristine weekly for 4 weeks. The response was impermanent. The patient has been hospitalized several times due to recurrent epistaxis and ecchymosis. TPO-RA and rituximab were not second-line treatment options due to family socioeconomic conditions. Bone marrow aspiration has already been performed and showed elevated megakaryocyte count, which led to the diagnosis. Therefore, we decided to administer a low fixed dose of rituximab (100 mg) weekly for 4 consecutive weeks. Fortunately, the platelet counts gradually increased and remained within the normal range. When the patient was last seen in August 2024, six months after the last dose of rituximab, he was asymptomatic and was provided with free corticosteroid-containing medications for the first time since diagnosis. The platelet count was $600 \times 10^3/\text{dL}$. The boy is still seen by the doctor and undergoes monthly follow-up (Table 1).

Discussion

Immune thrombocytopenic purpura (ITP) is one of the most common platelet disorders in children. It usually occurs from 1 to 7 years of age. Its incidence in children is approximately 4.0-5.3 per 100,000, which is approximately twice as much as in adults [2, 6]. The disease is usually preceded by a viral infection. It can also occur after vaccination, especially the measles, mumps, and rubella (MMR) vaccine [7]. For children who require therapy without life-threatening bleeding, corticosteroids are the recommended first-line therapy compared with IVIG or anti-D [8]. Our patient was diagnosed at the age of 8 years after a viral infection. The treatment started with prednisolone (1 mg/kg), and the patient exhibited a good response. However, he had been corticosteroid-dependent for the past four years. IVIG, splenectomy, and vincristine did not provide a robust response. Rituximab is a chimeric anti-CD20 monoclonal antibody that causes rapid but reversible depletion of CD20-positive B lymphocytes and modulation of T cells. It has a response rate of up to 50%. Various regimens have been proposed, including a standard dose (375 mg weekly for four weeks) and a low fixed dose regimen (100 mg weekly for four weeks), with the advantages of low cost and potentially fewer side effects. However, the gold standard dose remains unknown [9, 10]. In our case, we administered 100 mg weekly for four weeks and achieved a very good response after the second week of administration. Six months later, the patient remained asymptomatic and was free of corticosteroid-containing medications for the first time since initial diagnosis.

Conclusion

We succeeded to manage a case of chronic ITP in a corticosteroid-dependent child who exhibited a temporary response to IVIG, splenectomy, and vincristine. We were able to control symptoms in the long-term by using a low fixed dose of rituximab.

Conflict of Interest

None declared by the authors.

Table 1. Platelet counts during the treatment and follow-up period

Date	Platelet count	Comments
June 10, 2023	$641 \times 10^3/\text{dL}$	After splenectomy
July 07, 2023	$1200 \times 10^3/\text{dL}$	
July 18, 2023	$1023 \times 10^3/\text{dL}$	
August 28, 2023	$5 \times 10^3/\text{dL}$	There was a relapse with bruises
September 16, 2023	$800 \times 10^3/\text{dL}$	The patient improved on IVIG, but there was a relapse after 7 days and the platelet count became 6,000
23.09.2023	$1100 \times 10^3/\text{dL}$	The patient showed improvement with the use of vincristine+1mg/kg prednisolone
May 02, 2024	$8 \times 10^3/\text{dL}$	Platelet count declined
May 30, 2024	The first dose of rituximab was administered (100 mg weekly for 4 weeks)	
June 06, 2024	$290 \times 10^3/\text{dL}$	
June 13, 2024	$650 \times 10^3/\text{dL}$	
June 20, 2024	$373 \times 10^3/\text{dL}$	
June 27, 2024	$1240 \times 10^3/\text{dL}$	
July 04, 2024	$500 \times 10^3/\text{dL}$	
July 11, 2024	$477 \times 10^3/\text{dL}$	
July 18, 2024	$460 \times 10^3/\text{dL}$	
July 29, 2024	$635 \times 10^3/\text{dL}$	
August 13, 2024	$600 \times 10^3/\text{dL}$	
September 13, 2024	$688 \times 10^3/\text{dL}$	
October 13, 2024	$998 \times 10^3/\text{dL}$	
November 01, 2024	$540 \times 10^3/\text{dL}$	

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Ethical approval

Written consent was obtained from the parents of the patient.

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