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Brief Report

A large intracardiac thrombus in a child with steroid-resistant nephrotic syndrome

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Abstract Intracardiac thrombus is a rare complication of nephrotic syndrome in children, and only a few cases have been reported. We report the case of a 3.5-year-old child diagnosed with Henoch–Schönlein Purpura and steroid-resistant nephrotic syndrome for 2 months. The patient had massive oedema but no cardiovascular instability. Echocardiography displayed a large thrombus within the right atrium and the patient was successfully managed with enoxoparine.

Keywords: Thromboembolism; children; nephrotic syndrome

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Children with nephrotic syndrome are at increased risk for thromboembolic events.[1–6] The incidence of this complication in children is 2%, which represents a much lower risk than that of adults with nephrotic syndrome.[6] Both arterial and venous thromboses may be seen, including renal vein thrombosis, pulmonary embolus, sagittal sinus thrombosis, and thrombosis of indwelling arterial and venous catheters.[1–6] The mechanism and optimal treatment strategies of intracardiac thrombus are still unclear. Here, we report the case of a 3.5-year-old child with steroid-resistant nephrotic syndrome and intracardiac thrombus.

Case

A 3.5-year-old girl was referred to our hospital for treatment of intracardiac thrombosis. She had been diagnosed with Henoch–Schönlein Purpura and nephrotic syndrome 2 months ago. Despite standard treatment with oral prednisolone, cyclosporin A, enalapril, and dipyridamole for 2 months, nephrotic syndrome persisted as observed by clinical and laboratory examinations. Renal biopsy was compatible with Henoch–Schönlein Purpura and Immunoglobulin A nephropathy. Meanwhile, searching for pleural effusion, a large intracardiac thrombus was detected by echocardiography and the patient was admitted to our hospital 3 days later. Physical examination revealed generalised oedema and first-degree systolic murmur but no cardiovascular instability. Complete blood count, renal function test, and electrolytes were normal. Total cholesterol and triglycerides levels were elevated. Serum albumin and total protein were markedly decreased. Anti-nuclear antibody, antideoxyribonucleic acid, complement 3 and 4, and immunoglobulins were normal. On admission, echocardiography showed a large right atrial non-organised thrombus. The thrombus was 19×13 millimetres in size and heterogeneous in appearance (Fig 1). She also had a small patent foramen ovale. The sizes and functions of both ventricles were normal. Doppler ultrasound of both kidneys was normal; there was no evidence of renal thrombosis. Coagulation parameters were normal. Apoprotein A, protein S, Factors II, VIII, IX, and X, von-Willebrand factor antigen, Protein C, and Homocysteine were normal. Antiphospholipids and anticardiolipin antibodies, Factor V Leiden, Factor II, and methylenetetrahydrofolate reductase

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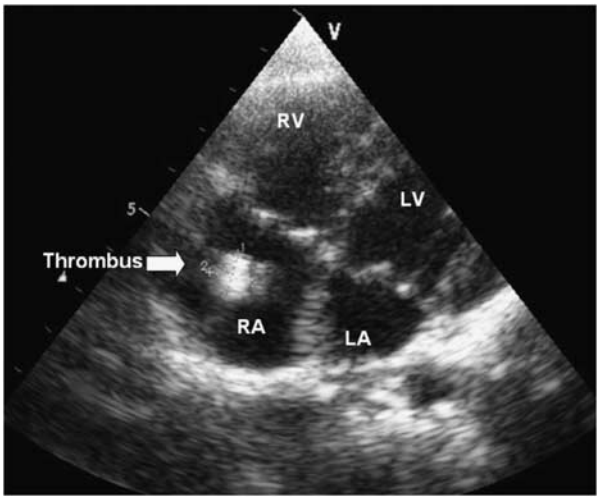

Figure 1.
Echocardiography showing a large right atrial thrombosis (19 × 13 millimetres) in an apical four-chamber view (LA = left ventricle, LV = left ventricle, RA = right atrium, RV = right ventricle).

gene mutations were negative. We started anticoagulation with enoxoparine and intavenous pulse methyl prednisolone (30 milligrams per kilogram for 3 days). After 11 days of treatment with enoxoparine, a repeated echocardiography displayed the regression of thrombus. For treatment of nephrotic syndrome, she received alternate-day therapy with oral prednisolone and cyclophosphamide. After 2 months, her general condition improved and she had no oedema. The thrombus was not detected by echocardiography and cardiac structures and functions were normal. Remission was not achieved. We planned to continue cyclophosphamide for 12 weeks and enoxoparine for prophylaxis of thrombosis.

Discussion

The presented case had secondary-form and steroid-resistant nephrotic syndrome. Children with a secondary form of nephrotic syndrome have a higher incidence of thromboembolism than those cases with minimal change disease.[6] In addition, the incidence of clinically apparent thromboembolism in children with steroid-resistant nephrotic syndrome (3.8%) is higher than those with steroid-sensitive nephrotic syndrome (1.5%).[7] There were no reports of intracardiac thrombus in children with Henoch–Schönlein Purpura, but many in adults with secondary-form nephrotic syndrome.[8,9]

The possible mechanism of thrombosis in cases with nephrotic syndrome is multifactorial. Intravascular volume depletion, thrombocytosis, platelet hyperactivity, hypercholesterolaemia, and infections play a role for pathogenesis of thromboembolism.[10] Some iatrogenic factors such as immobilisation, multiple venous puncture, and treatment with diuretics and steroids may increase the risk of thrombosis. Hypercoagulability state in some inherited disease may be responsible for development of thromboembolism.[5] The risk factors in our patient included a prolonged course of oedema, hypoalbuminaemia, hyperlipidaemia, and secondary form of nephrotic syndrome. Overaggressive diuresis should be avoided and the use of indwelling catheters limited because these factors may increase the likelihood of clotting complication.

In contrast to adult cases, children with thromboembolism usually have no symptom or subclinical course.[1–7] Similar to previously reported cases,[1–6] our patient was also asymptomatic at initial presentation and during the treatment period. She had no clinical findings or radiographic evidence related to pulmonary embolism, and thus we did not perform pulmonary scintigraphic examination. However, Hoyer et al[7] evaluated 26 nephrotic children in the remission period with pulmonary ventilation/perfusion test and found that 28% had evidence of pulmonary embolism. Prophylactic anticoagulation is not recommended in children unless they have had a previous thromboembolic event. However, some authors recommend prophylactic use of warfarin or aspirin in high-risk patients with plasma albumin less than 2 grams per decilitre, fibrinogen level over 600 milligrams per decilitre, or an antithrombin III level 70% below normal.[10]

The optimal management of intracardiac thrombosis is still unclear. In previous literature, anticoagulation is the first choice of treatment in many cases, but surgery should be considered in cases in which the risk of embolisation is high or the patient is not a candidate for anticoagulation therapy.[1–7] Echocardiographic characteristics of thrombus may help to estimate the risk of embolisation. Owing to the absence of cardiovascular instability, no evidence of peripheric embolisation, and non-organised appearance of the thrombosis, our case could not be managed with surgical resection of the thrombus. We thought that our patient had chronic thrombosis and thus preferred to use enoxoparine.

Conclusion

Paediatricians should be aware of thromboembolic complications in children with nephrotic syndrome. We recommend echocardiographic examination in cases with prolonged course of nephrotic syndrome.

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