



## Pulmonary Thromboembolism In Case Of Frequent Relapse Steroid Sensitive Nephrotic Syndrome

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Type of Publication: Original Research Paper

Conflicts of Interest: Nil

### Abstract

Nephrotic syndrome is common renal disorder. Children with nephrotic syndrome may develop a number of life-threatening complications amongst them thromboembolism is known complication. We present a case of 7-year-old female child case of frequent relapse steroid sensitive nephrotic syndrome diagnosed at age of 5 years with complaints oedema, abdominal distension and respiratory distress. On further evaluation CT Pulmonary angiography was done suggestive of pulmonary thromboembolism (saddle type) with 70-80% luminal compromise in the right main pulmonary artery and complete luminal compromise of segmental and subsegmental branches. Child was started on low molecular weight heparin and heparin infusion. Currently child is doing well and had gone in to the remission and started on steroid sparing agent.

**Keywords:** Nephrotic syndrome, thromboembolism, low molecular weight heparin, steroids

### Introduction

Nephrotic syndrome is common renal disorder with incidence of 20 to 40 per million population in the developed country.<sup>1</sup> Specifically in India it is estimated to be at 90 to 100 per million population. It is characterised by massive proteinuria (more than 40mg/m<sup>2</sup>/hr; 1g/m<sup>2</sup>/24hrs), hypoalbuminemia and oedema. Hypercholesterolemia is almost always present with occasional haematuria, hypertension and raised blood urea levels are observed. In about 95% cases of nephrotic syndrome in children it is primary glomerular abnormality. Remaining may be secondary to a systemic disorder. The onset of the disease specifically minimal change disease is between 2-5 years and in more common in boys. Children with nephrotic syndrome may develop a number of life-threatening complications during their disease course including infections, cellulitis, peritonitis, hypovolemia, thromboembolism,

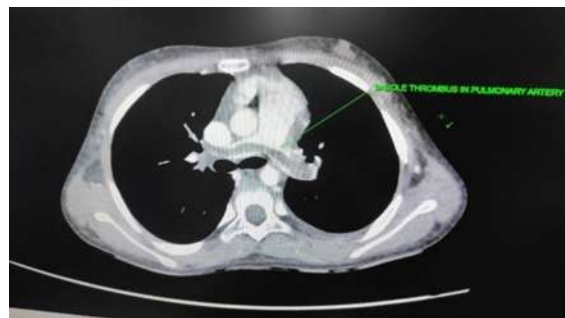
hyperlipidaemia. These infections are more common with frequent relapses and high dose steroid dependence. With advent of newer steroid sparing agents, relapses are better controlled and complications are uncommon. It requires a high degree of suspicion to identify a complication as many of its symptoms are masked due to presence of oedema and underlying hypovolemia. We present a case of steroid dependent nephrotic syndrome with timely diagnosed pulmonary embolism proved lifesaving.

### Clinical Presentation

We present 7-year-old female child born of non-consanguineous marriage was a known case of frequent relapse steroid sensitive nephrotic syndrome. She was diagnosed at age of 5 years and since then had frequent relapses and had received

short courses of high dose steroids. Steroid sparing agents were not considered by treating physician. She presented with oedema with abdominal distension, breathlessness and pain in abdomen since 4 days. On presentation child was in shock with blood pressure of 88/64 with tachycardia (heart rate of 140 beats/minute) with tachypnoea. Initial fluid resuscitation was done with IV crystalloid bolus of 20ml/kg for shock and oxygen support. Child was also started on IV Albumin (1gm/kg over 4 hours) and inotropic support due to fluid unresponsive shock. At this point child was noticed to be having raised jugular venous pulse with tender hepatomegaly. 2D echo was done which was suggestive of moderate Tricuspid Regurgitation with pulmonary artery hypertension with mean systolic pulmonary artery pressure of 55mmhg. CT Pulmonary angiography was done suggestive of

pulmonary thromboembolism (saddle type) with 70-80% luminal compromise in the right main pulmonary artery and complete luminal compromise of segmental and subsegmental branches. (Figure 1) Child fluid was tapered and she was started on Subcutaneous Low molecular weight heparin with heparin infusion. Serial APTT monitoring was done. She was started on prednisolone at 60mg/m<sup>2</sup>. Gradually shock improved and inotropes tapered. Review 2d echo suggested resolution of pulmonary hypertension. Low molecular weight heparin dose was reduced and she was started on oral acetylsalicylic acid as per paediatric cardiologist. On follow up child is doing well, has gone into remission and thrombus has resolved. Child was started on Mycophenolate as a steroid sparing agent. Low molecular weight heparin was continued for 3 months along with acetylsalicylate acid.



## Discussion

Nephrotic syndrome is common renal disorder in paediatric age group. The reported incidence of thromboembolic complications in adults with nephrotic syndrome ranges from 9 to 70% while in paediatric age group clinically evident thromboembolic complications as reported in various studies is only between 1.8 and 4.4 %.<sup>2</sup> Thromboembolic event is associated with many factors mainly attributed to haemoconcentration from hypovolemia, increase in blood viscosity, low levels of antithrombin III and protein S, fibrinogen concentration.<sup>3</sup> The commonest types of thrombosis in paediatric age group with nephrotic syndrome are renal vein thrombosis and pulmonary embolism.<sup>4</sup> It is diagnosed on the basis of radiological modality such as CT scan and gives idea about the severity of disease. Anticoagulation is recommended for a first venous thromboembolism is at least 3–6 months and

nephrotic syndrome has resolved or is in remission. There is limited role of thrombolysis.<sup>5</sup> There is no proven role of prophylactic anticoagulation in diagnosed case of Nephrotic syndrome.<sup>6</sup> In cases of frequent relapses with complications or steroid dependent nephrotic syndrome, signs of steroid toxicity consider use of steroid sparing agent to reduce the further complications and to improve the prognosis.

## Conclusion

We are presenting this case to highlight the importance of early recognition of complication and initiation of treatment. It also highlights the need to start early steroid sparing agents in children with FRNS and SDNS to improve the prognosis and to prevent dreaded complications of nephrotic syndrome

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