

Right ventricular thrombus in Wegner's granulomatosis

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Abstract

A 14-year-old boy presented with frank haematuria following two weeks of fever, coryzal symptoms and epistaxis. Renal biopsy and serology confirmed Wegner's granulomatosis. This case illustrates the need for vigilance for signs of thrombosis in children with Wegner's granulomatosis, particularly in those with additional risk factors such as heavy proteinuria, reduced mobility and an indwelling central venous catheter.

Case Report

A 14-year-old boy presented with frank haematuria following two weeks of fever, coryzal symptoms and epistaxis. Nephrotic range proteinuria and hypoalbuminaemia were present. Renal function rapidly deteriorated requiring haemodialysis. Renal biopsy showed severe necrotising crescentic glomerulonephritis; serology revealed anti-PR3-ANCA titres >100 units/mL confirming Wegner's granulomatosis.

A right internal jugular central venous line was inserted for haemodialysis and treatment given with serial double filtration plasmapheresis, cyclophosphamide 300 mg/m² and rituximab 750 mg/m². Ten days later the patient

developed unilateral lower limb oedema and persistent tachycardia. Echocardiogram showed a large right ventricular thrombus (Figure 1). Tissue plasminogen activator was administered (2 mg/kg for three consecutive days) and anticoagulation initiated with intravenous heparin and continued subsequently with warfarin. Tachycardia and limb oedema resolved within 24 h of thrombolysis and serial echocardiograms demonstrated progressive reduction in thrombus size.

Thrombosis is a recognised complication of paediatric Wegner's granulomatosis,¹ however to the authors' knowledge this is the first reported case of intracardiac thrombus complicating its presentation.

Thrombotic risk factors in this patient included a nephrotic state, reduced mobility and the presence of a central venous catheter. Nephrotic proteinuria with hypoalbuminaemia is associated with a hypercoagulable state in children.² Intracardiac thrombi have been reported in nephrotic children with differing treatment strategies including thrombolysis and surgical excision.^{3,4} This case illustrates the need for vigilance for signs of thrombosis in children with Wegner's granulomatosis, particularly in those with additional risk factors such as heavy proteinuria, reduced mobility and an indwelling central venous catheter.

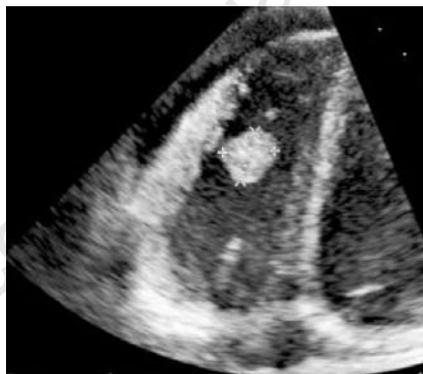


Figure 1. Right ventricular thrombus.

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References

1. Von Scheven E, Lu T, Emery H, et al. Thrombosis and Pediatric Wegener's Granulomatosis: Acquired and Genetic Risk Factors for Hypercoagulability. *Arthritis Rheum* 2003;49:862-5.
2. Zaffanello M, Franchini M. Thromboembolism in childhood nephrotic syndrome: a rare but serious complication. *Hematology* 2007;12:69-73.
3. Weisz W, Kemper MJ, Weil J, Müller-Wiefel DE. Asymptomatic intracardiac thrombus in steroid-sensitive nephrotic syndrome. *Pediatr Nephrol* 2002;17:287-9.
4. Mortazavi F, Samadi M. Asymptomatic intracardiac thrombus in a child with nephrotic syndrome. *Arch Iranian Med* 2006;9:42.