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Case report

A complicated steroid resistant nephrotic syndrome: a large intracardiac thrombus with bilateral distal embolization

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Abstract

Nephrotic syndrome (NS) is a consequence of the reduced ability of the glomerulus barrier to exclude proteins of intermediate size and other macromolecules from urine. Albumin and proteins that modulate the coagulation cascade are among the substances eliminated in urine. This is responsible of thromboembolic complication. The incidence of this extra renal complication is probably underestimated because of asymptomatic thromboembolic events. We report the case of a 23 years old man followed up for an idiopathic nephrotic syndrome since childhood who presented a large intracardiac thrombus complicated by distal embolisation of his two lower limbs, successfully treated by fogartisation-embolectomy and oral anticoagulation.

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Introduction

Nephrotic syndrome is defined as the presence of heavy proteinuria (protein excretion greater than 3.5g/24h), hypoalbuminemia (less than 3.0g/dl), hyperlipidemia and peripheral oedema. Primary etiologies are minimal glomerular injury, focal segmental glomerulosclerosis and non-membranous glomerulonephritis. Secondary etiologies are diabetes, high blood pressure and amyloidosis [1]. This syndrome is associated with an increased risk for thromboembolic complications. When the Intracardiac location remains very rare with high morbidity and mortality [2].

Patient and observation

We report the case of 23 years old man, with a history of idiopathic nephrotic syndrome since childhood treated by corticosteroids. He was admitted at emergency department for acute pain of his both lower limbs. There was no history of immobilization or recent diuretic therapy. Physical examination revealed a coldness and pulseless of both lower limbs (abolished bilateral popliteal and distal pulses). His blood pressure was 130/80mmHg, cardiac and lung examination were unremarkable. Renal function test, electrolytes, complete blood count and coagulation parameters were normal. Serum albumin and total protein were markedly decreased; total cholesterol and triglycerides levels were elevated. A urine dipstick test was negative for protein. Electrocardiogram showed sinus rhythm. Emergent lower extremity computed tomography angiography (CTA) was performed and revealed an occlusion of right popliteal and left superficial femoral arteries (Figure 1). Transthoracic echocardiography showed a large thrombus in the apico-septal wall of the left ventricle measuring 40x30x20mm (Figure 2). The ejection fraction of left ventricule was preserved without kinetic abnormalities. Urgent revascularization (fogartisation with embolectomy) of acute lower extremities ischemia was performed, secondly completed by medical therapy consisting of thrombolysis in situ with rTPA and intravenous heparin. The result was successful on the right lower leg while the patient underwent a Transmetatarsal Amputation of left foot. Heparinotherapy was relayed by oral anticoagulation. Repeated echocardiography showed a total dissolution of the thrombosis five weeks later.

Discussion

The most common sites of thrombosis in patients with nephrotic syndrom are the deep leg veins and inferior vena cava [3]. Arterial thromboses are less frequent than venous thromboses and can be located in coronary, femoral, pulmonary, cerebral, mesenteric, renal, aortic, subclavian and common iliac arteries [4]. While Intraventricular thrombosis is the least frequent sites of thrombosis [5]. Mahmoodi et Al reported that absolute risks of venous thromboembolism (VTE) (1.02% per year) and arterial thromboembolism (ATE) (1.48% per year) were each approximately 8 times higher in Nephrotic Syndrome patients than in the general population [6]. One of the prevailing hypotheses of thrombosis in NS is an imbalance between antithrombotic and thrombotic factors for a hypercoagulable state, as well as thrombocytosis, platelet hyperactivity, hypercholesterolemia, steroid use, infections and intravascular volume depletion [5]. NS induces the breakdown of permselectivity barrier of the glomerular capillary wall, allowing the passage of The low molecular weight coagulation factors (Factors IX, XI), antithrombin III, plasminogen, protein C and protein S, Which makes their serum concentration reduced. At the same time, there is an increase in high molecular weight coagulation factors (Factors II V, VII, VIII, and X), macroglobulins, fibrinogen and thromboplastin, which precipitates thrombosis [7]. Other studies have highlighted the role of platelet hyperactivity, hyperviscosity of blood, hypoalbuminaemia and hypercholesterolemia. In addition, some iatrogenic factors such as immobilization, multiple vein puncture and treatment with diuretics and corticosteroids diuretics, may increase the risk of thrombosis [5]. Because of the scarcity of this complication, the optimal management of intracardiac thrombosis in patient with nephrotic syndrome is still unclear. Echocardiographic characteristics of thrombosis may help to estimate the risk of embolization; including; mobility, shape, heterogeneity, echo density, layering, central echo lucency, presence within an aneurysm and association with low-density swirling echoes [5].

Anticoagulation is treatment of choice. However surgery should be considered if the risk of embolization is high or whenever the anticoagulation therapy is contraindicated [5]. According to Joo Myung Lee et and Al, the rate of post-treatment thromboembolism was not significantly different whatever the chosen treatment (anticoagulation, surgery or antiplatelet agents) in patients with left ventricular thrombus; however, operative treatment tended towards less post-treatment thromboembolism than other treatment [8]. Soren C et al. reported that high dose intravenous heparin is a highly effective and safe treatment for left ventricular thrombosis at high risk of embolization. In most cases, thromboses disappear completely within 1 to 3 weeks of anticoagulation without embolic or hemorrhagic complications [9]. In our case, the coexistence of thrombosis in left ventricle and in the lower extremities arteries in a context of hypercoagbility seems unlikely considering the aspect of intracardiac thrombus and arterial lesions. We leans more towards an intracardiac thrombus complicated by multiple arterial emboli. Considering the rapid regression of intra-cardiac thrombosis on the association a Low-molecular-weight heparin, warfarin and acetylsalicylic acid, We opted for medical therapy alone. Local thrombolysis allowed the rescue of the right leg and the limitation of the amputation of left leg to the knee level. The current scientific literature does not provide a solid answer on the role of primary thromboprophylaxis among patients with NS. One possible exception is patients with membranous nephropathy and hypoalbuminemia who are at low risk for bleeding for whom primary thromboprophylaxis can be proposed. Primary thromboprophylaxis can be indicated in patient with known nephrotic syndrome and a history of thromboembolic complications. As our patient was admitted with diffuse thromboembolic episode we decided to put him on oral anticoagulation during a long term, with careful monitoring of all the factors favoring an hypercoagulability state in particular the degree of hypoalbuminemia [10].

Conclusion

Patients with NS are at high risk of thrombotic events. The current scientific literature does not provide a solid answer on the role of primary thromboprophylaxis among patients with NS, neither on the manner in which we have to manage intra cardiac thrombotic complications. A thorough approach which includes accounting for all additional thrombotic risk factors is therefore essential.

Competing interests

The authors declare no competing interest.

Authors' contributions

All the authors have read and agreed to the final manuscript.

Figures

Figure 1: Computed tomography angiogram (CTA) of the abdominal aorta and bilateral lower limb showing occlusion of right popliteal and left superficial femoral arteries
Figure 2: Transthoracic echocardiography: apical four chamber view showing apico-septal thrombus of the left ventricle

References

1. Chaudesaygues E, Grasse M, Marchand L, Villar E, Aupetit J-F. Embolie pulmonaire révélant un syndrome néphrotique: à propos de quatre cas. Annales de cardiologie et d'Angéiologie. 2014 Nov 1; 63(5): 385-8.**Google Scholar**

2. Yang Y, Lv J, Zhou F, Chen M, Wang R, Zhao M *et al*. Risk factors of pulmonary thrombosis/embolism in nephrotic syndrome. Am J Med Sci. 2014 Nov; 348(5): 394-8. **PubMed** | **Google Scholar**

3. Weisz W, Kemper MJ, Weil J, Müller-Wiefel DE. Asymptomatic intracardiac thrombus in steroid-sensitive nephrotic syndrome. Pediatr Nephrol. 2002 Apr; 17(4): 287-9. **PubMed** | **Google Scholar**

4. Hull RP, Goldsmith DJA. Nephrotic syndrome in adults. BMJ. 2008 May 24; 336(7654): 1185-9. **PubMed**| **Google Scholar**

5. Gurbanna BA, Smer AM, Abdulmajid MR, Alhasadi MM, Almosbahi HK, Derbi HA *et al*. Nephrotic syndrome with intracardiac thrombosis: a case report. Ibnosina Journal of Medicine and Biomedical Sciences. 2009 Jan 9; 1(2): 67. **Google Scholar**

6. Mahmoodi BK, ten Kate MK, Waanders F, Veeger NJGM, Brouwer J-LP, Vogt L *et al*. High absolute risks and predictors of venous and arterial thromboembolic events in patients with nephrotic syndrome: results from a large retrospective cohort study. Circulation. 2008 Jan 15; 117(2): 224-30. **PubMed** | **Google Scholar**

7. Iwaki H, Kuriyama M, Neshige S, Takeshima S, Himeno T, Takamatsu K *et al*. Acute ischemic stroke associated with nephrotic syndrome: incidence and significance-retrospective cohort study. eNeurologicalSci. 2015 Sep 1; 1(3-4): 47-50. **PubMed** | **Google Scholar**

8. Lee JM, Park JJ, Jung HW, Cho Y-S, Oh I-Y, Yoon C-H *et al*. Left ventricular thrombus and subsequent thromboembolism, comparison of anticoagulation, surgical removal and antiplatelet agents. J Atheroscler Thromb. 2013; 20(1): 73-93. **PubMed** | **Google Scholar**

9. Heik SC, Kupper W, Hamm C, Bleifeld W, Koschyk DH, Waters D *et al*. Efficacy of high dose intravenous heparin for treatment of left ventricular thrombi with high embolic risk. J Am Coll Cardiol. 1994 Nov 1; 24(5): 1305-9. **PubMed** | **Google Scholar**

10. Mirrakhimov AE, Ali AM, Barbaryan A, Prueksaritanond S, Hussain N. Primary Nephrotic Syndrome in Adults as a Risk Factor for Pulmonary Embolism: an up-to-date review of the literature. Int J Nephrol. 2014; 2014: 916760. **PubMed** | **Google Scholar**

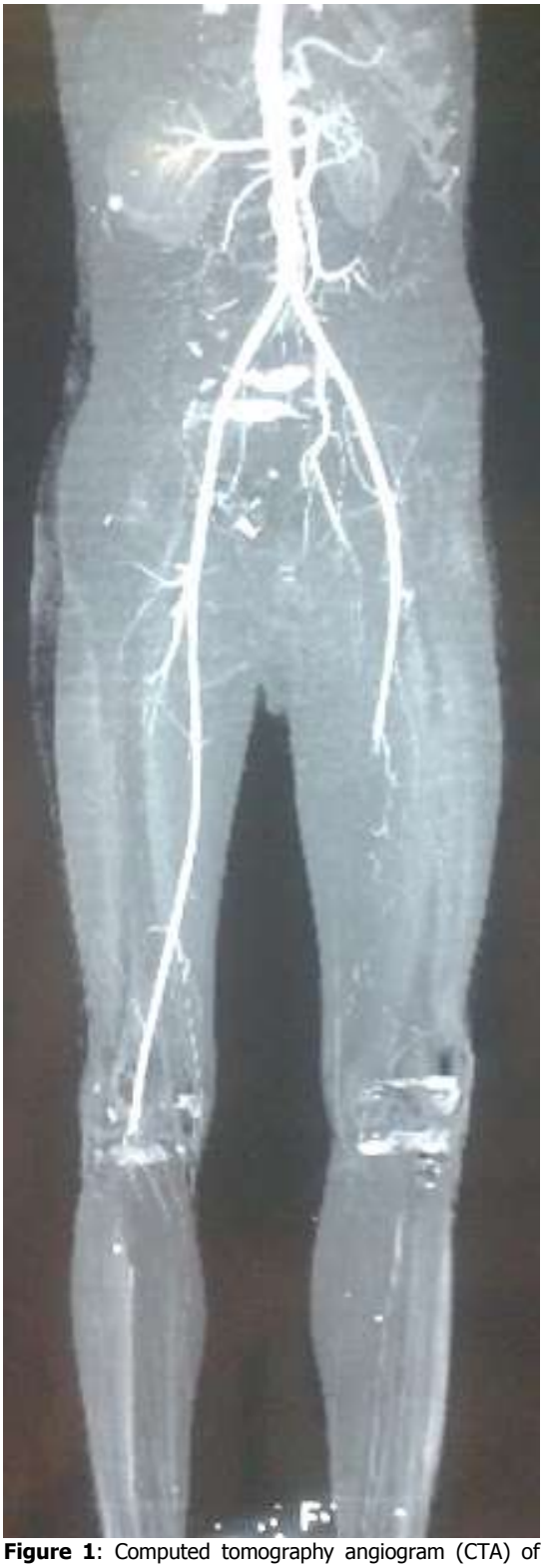

Figure 1: Computed tomography angiogram (CTA) of the abdominal aorta and bilateral lower limb showing occlusion of right popliteal and left superficial femoral arteries

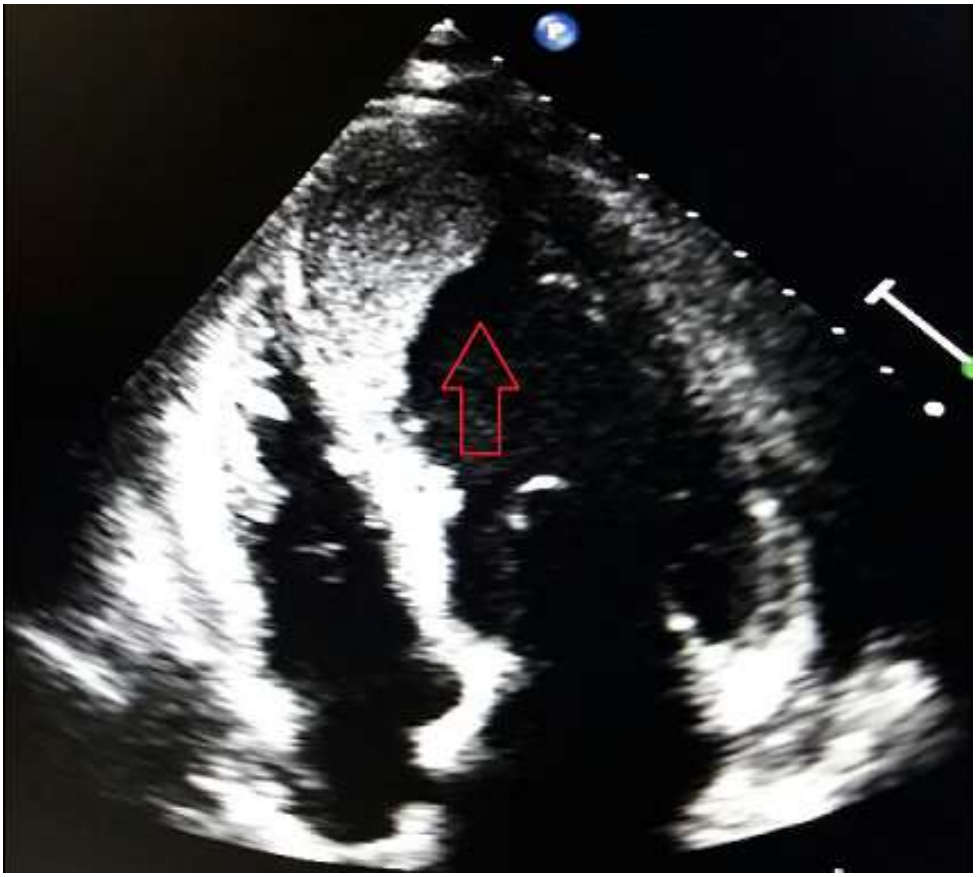

Figure 2: Transthoracic echocardiography: apical four chamber view showing apico-septal thrombus of the left ventricle