Mesenteric Arterial Thrombosis Revealing Relapse of Nephrotic Syndrome

Maryem Ferjani

department of pediatrics charles nicolle hospital

Yosra Zaimi (✉ yosrazaaimi@hotmail.fr)

department of Gastro Enterology charles nicolle hospital

Nouha Trad

department of Gastro Enterology charles nicolle hospital

Yousra Hammi

department of pediatrics charles nicolle hospital

Myriam Ayari

department of Gastro Enterology charles nicolle hospital

Shema Ayadi

department of Gastro Enterology charles nicolle hospital

Taha Sayari

department of pediatrics charles nicolle hospital

Tahar Gargah

department of pediatrics charles nicolle hospital

Case Report

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Abstract

Background: Nephrotic syndrome (NS) is associated with a hypercoagulable state and may be complicated by thrombotic events. Venous thrombosis is well acknowledged, while arterial thrombosis is rather unusual.

Case presentation: we present the case of a 20-year-old woman with a 12-year history of idiopathic NS revealed by extensive cerebral venous thrombosis with pulmonary embolism treated with anticoagulation therapy and oral corticosteroid therapy followed by mycophenolate mofetil (MMF). The thrombophilia assessment did not show any abnormalities. The evolution was marked by the occurrence of several NS relapses controlled by oral corticosteroid therapy until 2017. Subsequently, the patient had not presented a relapse of her disease, the anticoagulant treatment and the MMF were therefore stopped. One year later, the patient presented with severe diffuse acute abdominal pain associated with postprandial vomiting and bilateral lower limb edema. Laboratory results confirmed a NS relapse. An abdominal CT scan revealed acute thrombosis of the superior mesenteric artery with acute mesenteric ischemia. Intraoperative exploration showed mesenteric ischemia with extensive necrosis of the small intestine making their resections incompatible with life. The patient died after 48 hours.

Conclusions: Mesenteric arterial thrombosis, which is a rare but a life-threatening NS complication, should always be considered, especially in the case of acute non-specific digestive symptoms.

Background:

Nephrotic syndrome(NS) is a well-known condition characterized by a hypercoagulable state due to dysfunction in the hemostatic system. Actually, thrombotic events remain one of the most serious and life-threatening complications in patients with NS. Venous thrombosis during this syndrome has been frequently reported in studies. Nevertheless, NS-related arterial thrombosis is rarer and less well-known [1]. To best to our knowledge, four cases of mesenteric arterial thrombosis as a manifestation of NS have been reported so far in the literature[2–5]. Herein, we report a case mesenteric arterial thrombosis revealing NS relapse and leading to acute mesenteric ischemia and poor outcome.

Case Presentation:

A 20-year-old woman was followed up since the age of eight for idiopathic nephrotic syndrome inaugurated by cerebral venous thrombosis extended to the right jugular vein with a massive pulmonary embolism. The patient had no other medical or surgical history. She was accordingly put on anticoagulant therapy and oral corticosteroid therapy with good evolution. Thereafter, the patient received several cures of high-dose corticosteroids for relapses of NS. She was, hence, put on mycophenolate mofetil (MMF) as a background therapy to avoid corticosteroids and ensure normal growth. An exhaustive assessment of thrombophilia was performed and did not show any abnormality. Homocysteine rate, blood fibrinogen rate, Protein C, protein S, antithrombin III, factor V Leiden mutation,

JAK-2 mutation, cryoglobulins, anticardiolipin antibodies, lupus anticoagulant and beta-1-glycoprotein antibodies were normal. The anticoagulant treatment was stopped after nine years. The evolution was enameled by the occurrence of several relapses of her disease controlled by oral corticosteroid therapy. Remission of NS has been noted since 2017, so MMF was gradually stopped in 2019 and the patient remained asymptomatic and without any relapse.

One year later, the patient came up to our emergency department for acute intense diffuse abdominal pain without any particular irradiation associated with postprandial vomiting and bilateral lower limb edema. The physical examination revealed an intense epigastric tenderness with no fever or any other abnormalities. Laboratory findings confirmed relapse of nephrotic syndrome: nephrotic range proteinuria and hypoalbuminemia. Renal and liver functions were normal. An abdominal ultrasound was performed urgently showing an intra-abdominal effusion of low to moderate abundance. An abdominal CT scan revealed acute thrombosis of the superior mesenteric artery with acute mesenteric ischemia [figure 1-2]. The patient was immediately routed to the operating room. Intraoperative exploration confirmed mesenteric ischemia with extensive necrosis of almost entirely of the small bowel making their resections incompatible with life [figure 3]. The patient died after 48 hours.

Discussion:

We report a case of a mesenteric ischemia caused by an acute thrombosis of the superior mesenteric artery, occurring in a young patient with a history of a NS evolving for twelve years and in remission since three years. Nephrotic syndrome is caused by kidney damage resulting in loss of urine protein through small pores in podocytes. This syndrome is characterized by significant proteinuria, hypoalbuminemia, and hypercholesterolemia. Severe complications may occur during the course of NS, including acute kidney injury, infection, bone mineral loss, and thromboembolic events. These thromboembolic complications might be the first manifestation of NS or may occur over time[6]. Our patient presented two episodes of thromboembolic complications during the follow-up, one of which was inaugural of NS. NS was incriminated with no abnormality of the thrombophilia assessment.

The predisposition to thromboembolic complications in NS could be due to a hypercoagulable state. An imbalance of antithrombotic and prothrombotic factors should explain this hypercoagulability[6, 7]. Moreover, steroids, by shortening prothrombin and speeding up partial thromboplastin times, can worsen the hypercoagulable condition[8].

Furthermore, several studies have shown that the risk of thrombosis is often impacted by the severity and duration of NS[1, 9]. In our case, the patient was followed up for an NS for 12 years and she received several courses of steroids but she had been in remission for three years.

Venous thrombosis was predominant and well acknowledged during NS while arterial thrombosis was less frequent, involving coronary, cerebral, renal and femoral arteries more than any other vessels. As far as we know, four cases of mesenteric arterial thrombosis as a manifestation of NS have been reported so far in the literature (Table 1) [2–5]. The mesenteric arterial thrombosis causes acute and severe

abdominal pain in most patients, usually associated with nausea and vomiting. In the early phase of thrombosis, there is a discrepancy between the intensity of the abdominal pain and the minimal results of the physical examination. This non specific symptomatology with poor physical examination makes the diagnosis of mesenteric arterial thrombosis difficult. Untreated mesenteric arterial thrombosis often moves towards acute mesenteric ischemia with intestinal necrosis. At this point, the abdominal pain becomes more and more diffuse as the ischemia progresses. Mesenteric infarction can also be signaled by fever, bloody diarrhea, and shock. The evolution at this stage, as in the case of our patient, is often towards death[10].

Conclusion:

Our observation highlights that mesenteric arterial thrombosis is a rare and life-threatening complication of nephrotic syndrome even in remission, which should always be suspected in any patient with any acute non-specific digestive symptoms.

Declarations:

- Ethical Approval and Consent to participate:

Not applicable

- Consent for publication:

Written informed consent for publication of clinical details and accompanying images was obtained from the family of the patient (parents)

- Availability of data and materials:

All data are available as part of the article and no additional source data are required.

- Competing interests:

No competing interest to declare .

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- Authors' contributions:

Maryem Ferjani ¹: Writing-review & editing

Yosra Zaimi ²: Conceptualization, Writing-original draft, Writing-review & editing , corresponding author

Nouha Trad ²: Writing-original draft

Yousra Hammi ¹ : Writing-review & editing

Myriam Ayari ² : Writing-review & editing , preparing figures

Shema Ayadi ² : Writing-review & editing

Taha Sayari ¹ : Writing-review & editing

Tahar Gargah ¹ : Data curation , Supervision, Validation

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Table:

Table 1: Summary of reported cases of mesenteric thrombosis complicating nephrotic syndrome

Case characteristics:	Year of publication	Journal	Authors
Congenital nephrotic syndrome of Finnish type in a term newborn complicated by Mesenteric arterial thrombosis and with poor outcome	1995	<i>Zhonghua Min Guo Xiao Er Ke Yi Xue Hui Za Zhi.</i>	Tsai MJ et al
Mesenteric arterial thrombosis complicating a nephrotic syndrome	1997	The New Zealand Medical Journal	Do M et al
Mesenteric and cerebral arterial thrombosis in an 8-year-old child with nephrotic syndrome	2002	The Indian Society of Nephrology	Sandhu JS et al
Mesenteric arterial thrombosis causing short bowel syndrome in an 8-year-old child with a steroid-dependent nephrotic syndrome and a homozygous mutation of methylenetetrahydrofolate reductase	2008	Pediatric Nephrology	Ulinski T et al
Our case: mesenteric arterial thrombosis revealing nephrotic syndrome relapse and leading to acute mesenteric ischemia and poor outcome in 20-year-women.	2021	-	Zaimi Y et al

Figures

Figure 1

Axial CT-scan

A : Pneumoperitoneum of large abundance extending to the retroperitoneum, the retrocrural space and the mediastinum

B : Diffuse hypoenhancement of the small bowel, with marked dilatation of the small and large bowel loops

Figure 2

Thrombosis of the superior mesenteric artery

C : Axial CT- scan

D : Sagittal reconstruction

Figure 3

An intraoperative aspect of mesenteric ischemia with extensive necrosis of almost all of the small bowel.