

Formation of obstructing blood clot in the ureter in a patient with Glanzmann's thrombasthenia

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Glanzman thrombasthenia is a rare hematologic disorder characterized by qualitative thrombocyte abnormality. Patients present with episodic mucocutaneous bleeding. Thrombosis is a paradox phenomenon observed in patients with Glanzman thrombasthenia and generally considered as a treatment complication. We present a 16-year-old girl referred for severe flank pain beginning after treatment of hematuria due to Glanzman thrombasthenia. The patient underwent endoscopy for further diagnosis and treatment because of the failure of radiologic evaluation. Although the resolution of the large clots was obtained with streptokinase instillation via the ureteral catheter, clot was mobilized with gentle insertion of ureteral catheter in the present case. *Blood Coagul*

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Introduction

Glanzmann's thrombasthenia is a rare hematologic disorder characterized by the platelet aggregation dysfunction. Clinically, Glanzman thrombasthenia manifests itself with a lifelong mucocutaneous bleeding tendency. Epistaxis (73%), gingival bleeding (55%) and menorrhagia (98%) are the most commonly encountered symptoms [1].

Thrombosis is considered as a paradoxical clinical presentation of Glanzman thrombasthenia and generally occurs as a treatment complication. Only a few cases of Glanzman thrombasthenia presented with thrombosis are reported in the literature [2,3]. Presence of a clot obstructing the urinary tract is a very rare complication of percutaneous renal biopsy, treatment of a hematologic disorder or the renal cell carcinoma [4–7]. These clots may lead to pain, infection and renal function deterioration if left untreated. However, the treatment of these patients requires more attention and a multidisciplinary approach due to tendency to bleeding. Here, a case of Glanzman thrombasthenia, referred with acute ureteral colic due to ureteral clot, will be presented and the current literature on therapeutic approaches will be reviewed.

Case report

A 16-year-old girl, previously diagnosed with Glanzman thrombasthenia, was admitted to the pediatric hematology department because of menorrhagia and hematuria. Despite conservative follow-up, the hematuria did not settle spontaneously. The hematuria and menorrhagia was controlled by platelet transfusion. Packed red blood

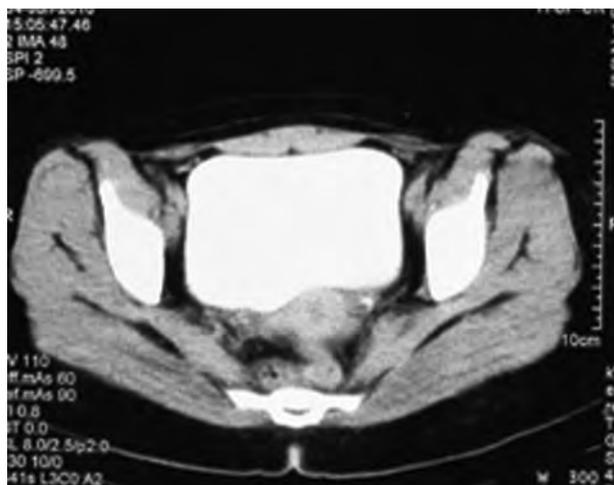
cell transfusion was required because the hemoglobin level fell below 7 g/dl. The patient was discharged due to clinical improvement and normalization of the laboratory measures.

The patient was referred to pediatric surgery department for severe flank pain, nausea and vomiting after 1 week. Physical examination revealed tenderness on the right loin. Ultrasonography showed grade-II hydroureteronephrosis. Intravenous urography revealed a complete obstruction in the distal ureteral level and hydroureteronephrosis (Fig. 1). To rule out any nonopaque ureteral calculi, computed tomography was performed and no reason leading to right hydroureteronephrosis was detected (Fig. 2). Despite analgesic and hydration therapy for two days, the pain of the patient did not resolve. Finally, it was decided to perform endoscopy with the consensus of the clinicians.

Under general anesthesia and lithotomy position, an 8Fr cystoscope was introduced. The bladder cavity and both ureteral orifices were normal. Initially, a 4F ureteral catheter was inserted through the right ureteral orifice to control the passage and facilitate the ureteroscope's ureteral access. An organized blood clot was discharged by the catheter's passage. Ureteral passage was controlled by retrograde ureteropyelography (Fig. 3). Because of bleeding tendency, the procedure was terminated with no further ureteroscopic evaluation.

The patient was asymptomatic after the procedure. No hematuria was observed. She was discharged on post-operative day 1.

Fig. 1



Grade II-hydronephrosis in the intravenous pyelography. The passage of the contrast medium to distal ureter is shown.

Discussion

Glanzman thrombasthenia is an autosomal recessive disorder caused by absence or deficiency of the glycoprotein IIb/IIIa complex. It is more common in countries where

Fig. 2



No passage of contrast-medium to the distal portion of right ureter and no sign of ureteral calculi.

Fig. 3



No obstruction in the retrograde-ureteropyelography.

consanguineous marriages are more frequent [8]. Patients usually present with a wide range of clinical manifestations, from minor bruising to life-threatening hemorrhages. Hematuria is a common clinical presentation of Glanzman thrombasthenia.

The clinical feature of the patients varies according to the type of the disease. Type I is characterized with total absence of glycoprotein IIb/IIIa complex, whereas a partial deficiency is observed in type-II. The variant or third type is defined as the impaired function of the glycoprotein complex [9]. The patients can be easily diagnosed with Glanzman thrombasthenia due to prolonged bleeding time, absent or reduced clot retraction and absent platelet aggregation with physiological agents and normal platelet count [10].

The treatment of the bleeding includes local measures, antifibrinolytic drugs and platelet transfusion. The blood products carry the risk of development of platelet refractoriness and infections [8,9]. Although hematuria is generally controlled with conservative treatment, blood products were needed because of the persistence of bleeding (hemorrhage) in the present case. Bričt *et al.* [11] reported a case with Glanzman thrombasthenia in which hematuria did not respond to platelet transfusions and underwent renal embolization.

Formation of blood clot is a complication of conditions leading to massive hematuria such as percutaneous renal biopsy [4], renal cell carcinoma [6] or hematologic disorders [12,13] and considered as a paradox phenomenon.

In the hematologic disorders, the treatment of hematuria with blood products can initiate the formation of blood clot [12]. In the present case, the blood clot also occurred after treatment of the hematuria with platelet transfusion. In this phase, the patients should be hydrated and the urine output should be monitored.

The blood clot may occur anywhere in/along the urinary tract and cause impaired renal drainage, pain and infection. In the present case, the main symptom was the pain. The physicians should be aware of the symptoms of the patients and suspect about the clot formation in the presence of flank pain and hydronephrosis.

Ureteral catheter placement, irrigation of urinary tract and low-dose streptokinase instillation are commonly used techniques for the resolution of obstructive blood clot [4,7]. The instrumentation of the urinary tract and streptokinase instillation therapy have the risk of recurrent hematuria. Krishnamoorthy *et al.* [14] recently reported a case with Glanzman thrombasthenia who underwent angioembolization because of rupture of an artery supplying the interpole segment of the right kidney by a guidewire during ureteroscopy. So, unnecessary instrumentation and aggressive treatment should be avoided in patients with hematologic disorders. In the present case, the impacted ureteral clot was gently removed by the insertion of a ureteral catheter through the orifice.

In conclusion, a multidisciplinary approach is essential for prompt treatment of hematuria in patients with hematologic disorders. Clot formation in the urinary tract should be kept in mind in presence of renal colic after the treatment of hematuria. Because of the risk of recurrent hematuria, the surgeons should be very careful during the instrumentation.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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