UDC 617.553-003.215:616.151.5-052

A CASE STUDY ON A RETROPERITONEAL HEMATOMA IN A YOUNG HEMOPHILIC PATIENT

Vivekanantharajah Kesha Shandihashini

Scientific supervisors: Assistant prof. *O. A Yarmolenko;* Dr. *E. M. Rebchenko*

Educational Establishment «Gomel State Medical University» Gomel, Republic of Belarus

Introduction

Hemophilia A, also known as classical hemophilia, is a genetic bleeding disorder caused by insufficient levels of a blood protein called factor VIII. [1].Hemophilia A can be mild, moderate or severe, depending on the baseline level of factor VIII made by that individual. In mild cases, prolonged bleeding episodes may only occur after surgery, dental procedures or trauma. [1] Severe disease presents commonly in children younger than 1 year and accounts for 43–70 % of hemophilia A cases. FVIII level less than 1 % of normal (<0.01 IU/mL) [2], can lead to important life-threatening complications such as intracranial hemorrhage, hemorrhages into the soft tissue around vital areas, such as the airway or internal organs, and develop into hematomas [3]. In addition, chronic debilitating joint disease results from repeated hemarthrosis; synovial membrane inflammation; hypertrophy; and, eventually, destructive arthritis [2]. Here we present a case of a retroperitoneal hematoma in a known case of Hemophilia A, mimicking a tumor.

Case presentation and discussion

A 24 years old male named K, was admitted with complaints such as weakness; dizziness; left sided abdominal pain; lymphostasis of left lower limb; limitation of movements in large joints. He was observed for hemophilia A since childhood and was repeatedly treated in the hematology department, where he received coagulation factor replacement therapy for factor VIII. Further from early pubertal period, he suffered frequent hemarthrosis of large joints and hematuria. With deterioration of his condition, he got hospitalized on 05/25/2017 in the hematological department. On the general physical examination, the patient was pale, weight-68 kg., height-178 cm with a pulse of 80/min, BP 120/80mmHg, Respiratory rate 20/min and febrile 101F. Initial lab reports included common blood test: RBC 3.19 10*12, Hb 73.3.1 g/l, PLT 336 10*9, WBC 9.67 10*9.Eosinophils: 1.42 %, Basophils: 0.62 %, Monocytes 8.58 % Lymphocytes: 11.48 % and Neutrophils 77.87 %. Biochemical blood test: Total protein: 81.6 g/l, Urea: 4 mmol/l, Creatinine: 60 μmol/l, Ferritin: 339.5 μg/l, C-reactive protein: 76.25 mg/l, Glucose: 4.9 mol/l, Aspartate aminotransferase (AST): 21 U/l, Lactate dehydrogenase (LDH): 662 U/l, Calcium: 2.4 mmol/l, Sodium: 145 mmol /l, Potassium: 5.88 mmol/l, Chlorides: 108.1 mmol/l, Rheumaticfactor: 10.3, Haptoglobin: 0.099g/l, cholesterol 3.21mmol/l. Coagulogram: APTT: 78.3 sec, Prothrombin index: 0.92, Fibrinogen: 7.5g/l, Thrombin time: 15.2 sec, INR: 1.05, Factor VIII: 1%, Factor VIII inhibitor: 0.47 (Bu/ml). Determination of blood group and Rh factor (Blood group O(I),Rh positive.

Patient found to be anemic and with increased levels of APTT, fibrinogen and decreased levels of factor VIII. Thus, managed for anemia and conservatively with fresh frozen plasma, clotting factor VIII for Hemophilia A. He showed improvement in APTT level and concentration of factor VIII. Further instrumental diagnostics performed to find the cause of severe pain syndrome and lymphostasis. Radiography of the chest cavity was norm without any focal shadows. ECG showed Moderate sinus tachycardia and heart rate 98 beats per minute. Esophagogastroduodenoscopy showed superficial gastritis and colonoscopy had no any pathology. Ultrasound of kidneys, adrenal glands, liver, pancreas, spleen and thyroid glands made, which

revealed diffuse changes in the liver and pancreas, cysts of the left kidney, and hematoma in the abdominal cavity on the left side. Nevertheless, consultation from surgeon excluded the need of surgical excision and patient was conservatively managed. In the following days patient complaints weren't subsided. Thus, CT and MRI were conducted after a week.

The enhanced CT of abdominal cavity revealed, a significant size (316×184×230 mm) liquid heterogeneous structure, with destruction of left iliac crest. According to the obtained MRI images, in the abdominal cavity, pelvis, a gigantic formation was visualized. It was compressing the intestines and bladder, through a defect (approximately 8×4 cm) from the crest of the left ilium, and the formation spreads to the left gluteal region, pushing the muscles apart. The formation is delimited, has a capsule 1–2 mm thick, an inhomogeneous structure, with areas of liquid and soft tissue intensity. Thus concluded as a volumetric formation of the abdominal cavity, small pelvis with left iliac crest destruction. A tumor was suspected. Further tests excluded the suspected tumor of other locations. Patient was managed in a multidisciplinary team consisting of oncologists, hematologists and surgeons. Moreover, suspected formation was excised in a 12-hour surgery with exclusion of malignancy. Post operatively patient was managed with Ferronal Belmed 35 mg 2 tablets 3 times a day. Folic acid 1 mg 5 tabs 2 times a day. And correction of congenital deficiency of factor VIII of blood coagulation by Emoclot 500 IU, 2000 IU intravenously 1 time per day. Discharged with improvement in a satisfactory condition.

Severe hemophilia usually manifests in the first few months of life. Prior to the availability of treatment with factor VIII preparations, young males died from uncontrolled bleeding, either spontaneous or a bleed after injury, before reaching 20 years of age. Those who survived suffered from the complications of frequent bleeding, primarily severe and incapacitating damage involving their weight-bearing joints. [4]. Bleeding in muscle includes 10–25 % of hemorrhagic conditions in severe Hemophilia [5]. Rare bleeds include central nervous system bleeds such as intracranial and spinal hematomas, urogenital bleeds, intra-abdominal bleeds like mesenteric and gastrointestinal wall hematomas and pseudo tumors in unusual locations.

Hemophilic pseudotumour arises in 1–2 % of patients with severe forms of hemophilia. Pseudotumours are slow-growing lesions comprising of a chronic hematoma delimited within a fibrous capsule that prevents resorption and promotes neovascularization and recurrent intracapsular bleeding. They can be of soft-tissue origin or osseous pseudotumors (intraosseous or sub-periosteal) [6]. Rare locations include the lungs,abdomen and retroperitoneal spaces [7]/ Pseudotumor is diagnosed based on clinical and imaging findings. A soft tissue pseudotumor may contain calcifications or ossifications. Adjacent bony structures may be normal or show involvement ranging from periosteal reaction to severe bone destruction [6]. CT and MRI can be used in visualizing the margins of hemophilic pseudotumours. In patients with severe hemophilia, the presence of typical imaging features confirms the diagnosis of pseudotumor. Osseous pseudotumors can mimic Ewing's sarcoma, metastasis or infection [8]. It is essential to remember that when there is a suspicion of a pseudotumor in a patient with congenital coagulopathy, we must always rule out the existence of a true tumor, which can be achieved through exhaustive study of the case by diagnostic imaging [9]. Ultimately, if there is any doubt, biopsy should be discussed, with special protocols for hemophilia patients.

When pseudotumors are large, they can compress neighboring anatomical structures [10]. The most frequent is the pelvic pseudotumor as the case described here, secondary to a hematoma of the iliopsoas muscle. The best treatment for pseudotumor in patients with congenital coagulopathies is prevention, that is, proper long-term hematological treatment of muscle hematomas, until total resolution (reabsorption). Imaging tests (US, CT scan, MRI) are fundamental for monitoring the evolution of hematomas in patients with congenital coagulopathies. In most cases the best solution is radical surgical excision [10]. Preoperative arterial emboli-

zation (ideally 2 weeks before surgery) may be helpful in giant pelvic pseudotumors. Surgical resection is considered only for symptomatic pseudotumors, and microinvasive endoscopic surgical approaches are preferred.

Conclusion

This case highlights the diagnostic and therapeutic challenges associated with hemophilic pseudotumor, especially in patients with large masses and with extensive bone destruction. Incidence of case like this are increased with increased longevity. Specially currence of complications affecting the osseous system. Here we studied one of the more serious bone complications where hemophilic hemotoma mimicked like a tumor. Sometimes we can find true tumors that mimic hemophilic pseudo tumors too. Diagnosing such pseudo tumor differs from diagnosing a cancerous tumor, and there is high potential for misdiagnosis thus resulting in severe bleeding and fatal consequences. Instead, diagnosing a pseudotumor usually involves taking a history, conducting a physical exam and performing an imaging study, such as a CT scan or magnetic resonance imaging (MRI). Early diagnosis is crucial for evaluation and for proper surgical planning. Replacement therapy is often the first therapeutic approach; however, surgery is the most effective and the only definitive treatment even though it may be associated with higher rates of complications. The main limitation of hemophilic pseudotumors is we have a low degree of evidence (case reports, case series etc). However, it cannot be forgotten that this is a very infrequent and potentially serious injury.

LITERATURE

1. https://rarediseases.org/rare-diseases/hemophilia-a/.
2. https://emedicine.medscape.com/article/779322-overview#a2.
3. Clinical findings in a patient with hemophilia A affected by COVID-19 / D. Cui [et al.] // Haemophilia. medscape. [Online] 2020. https://www.medscape.com/answers/779322-78130/what-are-possible-life-threatening-complications-of-hemophilia-a.
4. Revisiting hemophilia management in acute medicine / S. Sahu [et al.] // J Trauma Shock. 2011. doi:10.4103/0974-2700.82225, 4(2):292-298.
5. Haemophilia / H. Mansouritorghabeh [et al.]. Iran: Mashhad University of Medical Sciences and Scholar.
6. Imaging features of atypical bleeds in young patients with hemophilia. Diagn Interv Imaging / M. Gavrel [et al.] // Adamsbaum. 2019 Mar. Vol. 100(3). P. 135–145. doi: 10.1016/j.diii.2018.11.010. Epub 2018 Dec 14. PMID: 30559038
7. Imaging features of atypical bleeds in young patients with hemophilia / M. Gavrel [et al.] // Adamsbaum. 2019. Vol. 100. P. 135–145.
8. Intraabdominal hemophilic pseudotumor: case report / R. García-Pérez [et al.] // Rev Esp Enfermedades Dig. 2010. Vol. 102. P. 275–280.
9. Haemophilia imaging: a review / J. Maclachlan [et al.] // Skeletal Radiol. 2010. Vol. 38. P. 949–957.
10. Abjs. Mums. Ac. Ir. Hemophilic Pseudotumors // The Archives Of Bone And Joint Surgery. 2020. Vol. 8, № 2.
11. Hemophilic Pseudotumors: Diagnosis and Management / C. Rodriguez-Merchan [et al. // The archives of bone and joint surgery. 2020. Vol. 8. P. 121–130.
12. Hemophilic pseudotumor: radiologic-pathologic correlation / J. M. Stafford [et al.] // Radiographics. 2003. Vol. 23. P. 852–856.

UDC [616.98:578.834.1]:616.2-036.11

COVID 19-EFFECT ON PULMONARY SYSTEM. ACUTE RESPIRATORY DISTRESS SYNDROME. COMPARISON BETWEEN H1N1 AND SARS CoV19

Zeeshan Ahmad

Scientific supervisor*: Dr. Pervez Ansari MBBS MD* (internal medicine)
Kota, Rajasthan, India
Senior lecturer of the department *A. N. Kovalchuk*

Educational Establishment
«Gomel State Medical University»
Gomel, Republic of Belarus

Introduction

The research was conducted on 406 patients in total, during their admission in covid 19 ward and record data of H1N1 patients in the department of internal medicine at Govt. Medical Hospital KOTA Rajasthan India.