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RESEARCH ARTICLE

RUPTURED HEMORRHAGIC OVARIAN CYST IN A PATIENT WITH WILLEBRAND DISEASE: A CASE REPORT AND REVIEW OF LITTERATURE

A. Zerhouni, Y. Boulani, D. Farjia, O. Joutey, A. Rajaa, Y. Hafiani, I. Moussaid and S. Salmi
Department of Obstetrical Anesthesiology and Critical Care, Harouchi Hospital, Casablanca, Morocco.

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Abstract

Willebrand Disease (WD) is a familial haemorrhagic disease described in 1926 by Erik von Willebrand, a Finnish physician (1), and is related to a deficiency of Willebrand Factor (VWF) leading to a disorder of primary haemostasis and secondary coagulation. The clinical and biological expression of WD varies according to the form. The common feature is the existence of a constitutional quantitative and/or qualitative VWF deficiency. A classification of the different forms of the disease exists to guide the diagnosis, treatment and genetic information of patients. While isolated biological abnormalities are relatively common with a prevalence of nearly 1%, symptomatic forms of WD are more rare. The prevalence of WM type 3, the most severe form (autosomal recessive), has been estimated at 1 per million population. While autosomal genetic inheritance predicts that both sexes are affected in the same proportion, symptomatic forms are more common in women because of the haemostatic challenge of menstruation, pregnancy and childbirth. Ovarian cysts are fluid-filled sacs that lie on the surface of or within the ovary(2). Roughly 20% of women develop at least one pelvic mass in their lifetime. Therapeutic management is based on surgical haemostasis by total cystectomy and medical haemostasis by exogenous supply of Willebrand factor and finally resuscitation of any state of haemorrhagic shock starting with vascular filling with crystalloids, the introduction of vasoactive amines and finally massive transfusion.We report here the case of a 24 year old female patient, treated for Willebrand's disease since birth and who presented to the emergency department with haemorrhagic shock due to a ruptured haemorrhagic ovarian cyst with internal bleeding.

Introduction:-

Willebrand disease (WD) is a genetic bleeding disorder caused by a defect in the amount, structure or function of a factor involved in the early phase of the clotting process called Willebrand factor. We report here the case of a 24 year old female patient, treated for Willebrand's disease since birth and who presented to the emergency department with haemorrhagic shock due to a ruptured haemorrhagic ovarian cyst with internal bleeding.

Corresponding Author:- A. Zerhouni
Address:- Department of Obstetrical Anesthesiology and Critical Care, Harouchi Hospital, Casablanca, Morocco.

Case:
A 24 year old female patient, treated for Willebrand disease type 3 since birth, who presented to the emergency room with pelvic abdominal pain in the left iliac fossa, without digestive problems or external bleeding. Clinical examination on admission found

A conscious patient GCS of 15/15,blood pressure of 80/30 mmHg, Heart rate at 130 beats per minute, pulse saturation at 95% on air, polypnoeic at 27 cycles per minute, apyretic with discoloured conjunctiva and skin pallor.Abdominal examination revealed diffuse abdominal tenderness more marked in the left iliac fossa. The gynaecological examination was normal with a date of the last menstrual period 10 days ago. About biological exams Hemoglobin was at 6.3 g/l white blood cells at 9800 e/mm^3, platelets 202 000 e/$mm^3$with a correct hemostasis test. Beta-HCG was negative.

Abdominal ultrasound showed a left latero-uterine echogenic cystic image with peritoneal effusion of great abundance.

The patient was rushed to the operating room for an exploratory laparotomy under general anaesthesia revealing a ruptured haemorrhagic cyst with a large hematic peritoneal effusion. A cystectomy was performed with aspiration of the peritoneal effusion and saline lavage and massive intraoperative transfusion of red blood cells, platelets and fresh frozen plasma.

Discussion:-

There are three main types of Willebrand disease, of varying severity (3):Type 1 (50-75% of patients), the least severe: Von Waillebrand factor (VWF) is produced in lower quantities than normal, which may lead to a deficiency of factor VIII (FVIII).Type 2, also known as molecular variant (1-30% of patients), is classically more severe: VWF is produced but does not fulfil its function. A possible associated FVIII deficiency depends on the type of variant. Type 3, the most severe and rarest: the VWF deficiency is total and is associated with a constant and profound FVIII deficiency. Men and women are equally affected. Causes of the disease Willebrand disease is an inherited genetic disorder[3]. In people with this disease, the gene (located on chromosome 12) that determines the production of VWF has been altered (mutated). Each type of Willebrand disease has a different mutation in the gene. In type 1 or 2 Willebrand disease (except type 2N and some types 2A), which is the majority of cases, the disease is "dominant". This means that the person with the disease has inherited the gene from one of his or her parents (father or mother), who also has the disease.

More rarely, the gene mutation can occur in a person whose parents did not have a mutation in their gene (neo-mutation). A person who carries the mutated gene (and is therefore ill) has a 50% chance of passing the disease on to their children. In type 3 Willebrand disease (or type 2N and some types 2A), the transmission is "recessive": the affected person has received one copy of the mutated gene from each of his or her parents. The parents, themselves carriers of a single mutated gene, have no (or few) signs of the disease, but they can pass on a severe form of the disease to their children (one in four risk). The severity of the bleeding signs of Willebrand disease depends on the type of disease. Type 1 and type 2 are characterised by spontaneous or trauma-induced haemorrhages: skin and mucous membranes: bruising, epistaxis, gingivorrhages, menometrorrhages, digestive bleeding.There is a risk of bleeding, even in moderate de Willebrand's disease, after minor surgery or trauma which can be prevented by therapeutic measures. Type 3 adds to these skin and mucous membrane haemorrhages the risk of more serious internal haemorrhages: haematomas, haemarthrosis and haemorrhagic strokes. There is no cure for Willebrand disease, but regular management and monitoring of the disease can prevent or limit the risk of bleeding. When necessary, VWF deficiency can be corrected with two types of products:A drug (desmopressin or DDAVP), administered intravenously or by intranasal inhalation. It is most effective in type 1, rarely in type 2 and never in type 3. Plasma VWF concentrates (combined, for some types of Willebrand disease, with FVIII in the 1st injection), injected intravenously (4). They are effective in all types of the disease.

The effectiveness of these products is, however, limited in time (their duration of action is limited to a few hours). However, they can prevent or reduce bleeding.

A treatment adapted to each patient is defined by the doctor providing the specialised care. It depends on the type of Willebrand disease and the situations encountered (bleeding accident, childbirth, surgery, etc.): Either curatively to treat an unexpected bleeding event(5). Or, whenever possible, as a preventive measure (before a planned surgical

procedure or childbirth, for example), in order to prevent the occurrence of a bleeding complication. In this patient's case the surgical procedure was a cystectomy with aspiration of the hematic peritoneal effusion.

Apart from accidents and situations where there is a risk of bleeding, in the majority of cases, correction of VWF deficiency is not necessary, particularly in the case of minor bleeding that stops spontaneously.

However, it is important to follow simple preventive measures: compression of a small skin wound or nostrils for a few minutes in case of epistaxis, respect of contraindications of certain drugs (aspirin and non-steroidal anti-inflammatory drugs) and intramuscular injections in particular.

A massive transfusion may be necessary with transfusion of packed red blood cells, platelets and fresh frozen plasma. Injectable tranexamic acid with calcium. Vasoactive amines as in any state of haemorrhagic shock.

Conclusion:-

A deficiency in primary haemostasis, a component involved in the first stage of the coagulation process, results in Willebrand disease, a genetic bleeding illness. In any stage of hemorrhagic shock, blood clotting abnormalities should always be looked into. Desmopressin, an exogenous source of Willebrand factor, is the main component of treatment. A crucial step in the therapeutic therapy is surgical hemostasis to halt the bleeding. As in any state of hemorrhagic shock, massive transfusion may be required in some extreme situations.

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