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Ovarian Mass with Missed Abortion in Case of Bernard-Soulier Syndrome – An Operative Challenge

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ABSTRACT

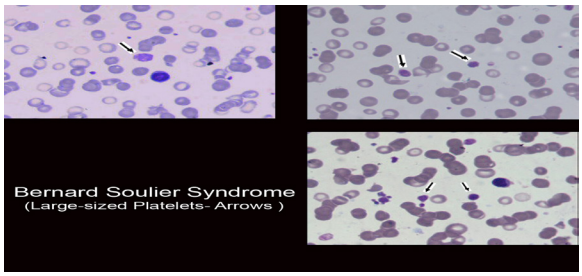
We hereby describe you a case of 22 year old lady with 2 month amenorrhea with spotting per vaginum with known case of Bernard-Soulier Syndrome characterized by a hereditary platelet disorder with thrombocytopenia, giant platelets, and a tendency toward bleeding. She was diagnosed with left adnexal cyst with 6 – 8 week pregnancy with thrombocytopenia. Patient was taken for dilatation & evacuation followed by laparotomy with left sided ovarian cyst removal with left oopherectomy. Pre-operative, intra-operative, post-operative platelet transfusion was given. Patient was discharged on 13th post operative day with normal ultrasound finding and explained about contraception and regular follow up.

KEYWORDS :

CASE REPORT:

A 22 year old woman presented to the OPD with history of 2 months amenorrhea and chief complaints of spotting per vaginum since 1 week and pain abdomen on and off since 1 year dull aching in nature and increasing in severity over the year. Patient was primi-gravida with active married life of four years. Patient was diagnosed as a known case of Bernard-Soulier Syndrome two years back when she was admitted in a private hospital for pain abdomen with fever. She was also diagnosed as having left adnexal cyst. On admission she was vitally stable. Per vaginum examination revealed uterine size of 6 – 8 weeks and left sided 5 X 6 cm adnexal mass.

LAB INVESTIGATIONS	
CBC	Hb - 8.8gm/dl WBC – 4600cells/cumm Platelet – 29,000/cumm
Peripheral Smear	Microcytic Hypochromic RBCs Reduced Platelet Mass with large platelets
RFT	Normal
LFT	Normal
Prothrombin time	Normal
activated Partial thromboplastin time	Normal
Clotting time	Normal
Bleeding time	Slightly prolonged to 4 min
Pelvic ultrasound	7 week intrauterine G-sac with left adnexal mass of size 68 X 46 X 48 mm with small multiple ill defined hypoechoic areas inside mass.



MANAGEMENT

The patient was scheduled for a planned Dilatation & Evacuation for missed abortion followed by laparotomy for ovarian mass in the same sitting. A haematologist was consulted for intra-operative and post–operative management of the patient. The pre, intra and post operative management of the patient was as follows:

Dilatation & evacuation followed by Laparotomy with left side ovarian cyst removal with left oophorectomy was carried out. The patient was vitally stable during intra and post-operative period. An intra-peritonial drain was kept during the surgery which was subsequently removed on 3rd post-operative day. She was given intravenous antibiotics for 3 days followed by oral antibiotics for next 7 days. Stitch removal of the patient was done on 10thpost operative day. Stitch line was healthy.

DAY	PLATELET RICH CONCENTRATE	OTHER BLOOD COMPONENTS (PACKED CELL/ WHOLE BLOOD/FFP	HEAMOGLOBIN(gm/dl)	PLATELET COUNT (/cumm)
PRE – OPERATIVE	10	1 WB	8.8	29,000
INTRA OPERATIVE	10	4 FFP, 1 PCV	7.58	79,000
POD – 1	10	-	5.9	98,800
POD – 2	6	1 WB	-	-
POD – 4	10	-	-	-
POD – 6	10	-	-	-
POD – 8	10	-	10.2	40,000
POD – 9	4	-	-	-

TABLE: PLATELET RICH CONCENTRATE AND OTHER BLOOD COMPONENTS GIVEN

A follow up pelvis ultrasound scan was done was showed normal findings. Patient was discharged on 13thpost operative day with Hb 10.3 gm/dl and PC 83,000/cumm. Histo-pathological evaluation of the specimen showed haemorrhagic luteal cyst of ovary. Patient was explained about contraception and was called for follow up after 6 weeks.

DISCUSSION:

Bernard-Soulier Syndrome is a hereditary platelet disorder characterized by thrombocytopenia, giant platelets, and a tendency toward bleeding. It is inherited in an autosomal recessive manner. The underlying biochemical defect is the absence or decreased expression of the glycoprotein Ib/IX/V complex on the surface of the platelet. This complex is the receptor for von Willebrand factor. The cause of the thrombocytopenia is not definitely known, but it is probably related to a decreased platelet life span. The end result is the lack of formation of the primary platelet plug and increased bleeding tendency in form of nose bleeds, mucosal bleeding or G I bleeding.

In pregnancy, Bernard-Soulier Syndrome may result in severe ante-partum and post partum haemorrhage. If the patient is closely monitored with regular platelet count monitoring and by maintaining

the platelet count above 60,000/cumm[1], even major surgery (laparotomy in this case) can be undertaken without complication. Nevertheless pregnancy with Bernard- Soulier Syndrome is a high risk state which requires vigilant monitoring and combined management from obstetrician, haematologist and blood bank facilities. Hence such patients should be preferably managed in tertiary care centres. Strict and regular monitoring with timely transfusions of blood products can be life saving in such cases. Lastly, patient should be called for follow – up regularly with platelet count monitoring.

CONCLUSION:

In conclusion, women with Bernard-Soulier Syndrome should be observed at an appropriate centre using a multidisciplinary approach. The primary treatment remains platelet transfusion. Appropriate individualised therapy for Bernard-Soulier Syndrome should be defined and new studies are needed on the role of the glycoprotein Ib/IX/V complex in regulating platelet turnover to better depict the clinical course.

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