Thrombocytapheresis in patient of essential Thrombocytosis with acquired von willebrand syndrome : A case report

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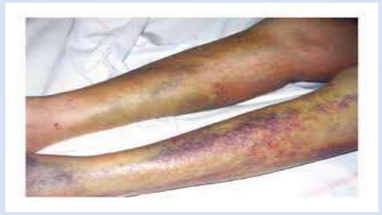
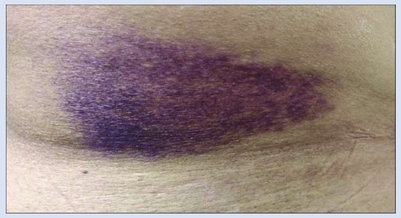
INTRODUCTION

- Essential thrombocythemia is one of the chronic myeloproliferative neoplasms characterized by clonal proliferation of myeloid cells with variable morphological maturation and hematopoietic activity.
- It is characterized by excessive clonal platelet production with a tendency to thrombosis and bleeding.
- Thrombocytapheresıs is the removal of platelets by apheresis techniques.
- Thrombocytapheresıs is generally recommended in patients with essential thrombocythemia with acute, severe thrombotic or hemorrhagic events.
- Therapeutic thrombocytapheresis using an automatic cell separator can help to achieve prompt platelet count reduction to decrease the rate of thrombotic events.
- In cases of extreme thrombocytosis, therapeutic thrombocytapheresis can be a useful procedure. The present case report is of a 76-year-old-man previously diagnosed with leukocytosis with acquired von willebrand syndrome ?myeloproliferative neoplasm.

CASE DETAIL

76 year old male patient presented to haematology at BMT center with complaints of ecchymosis over right forearm, generalised weakness and bodyache. He also complained of inability to walk due to severe pain along left lower limb. On examination there was swelling with redness noted in left gluteal region. Swelling was tender suggestive of underlying hematoma? Patient also had 4 cm splenomegaly on examination.





❖ **LABORATORY INVESTIGATIONS :**

1. **Bone marrow report :** The bone marrow biopsy findings suggestive of Myeloproliferative disorders suspicious of essential thrombocythemia.

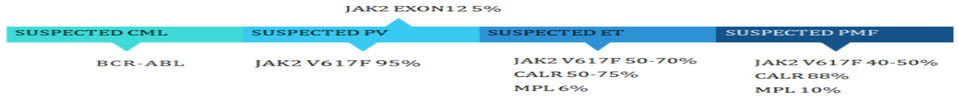
2. **MPN Panel report :** V617F mutation detected in exon 14 of JAK2 gene.

POSITIVE
(JAK2 V617F DETECTED IN EXON 14)

Gene	Variant	Location	Classification
JAK2 (NM_004972.4)	JAK2:c.1849G>T:p.V617F	EXON 14	Pathogenic

Interpretation:
- V617F mutation detected in exon 14 of JAK2 gene.

Myeloproliferative Neoplasm:



MATERIALS AND METHODS

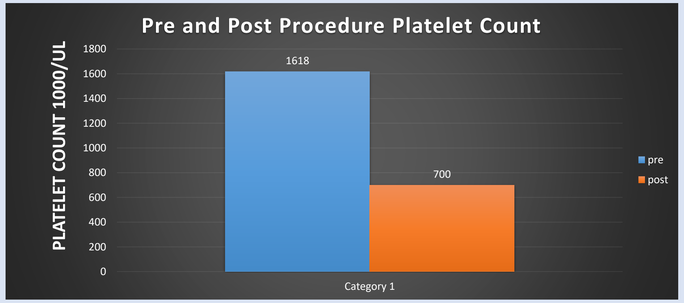
- This a case report presented in Department of Transfusion medicine at Pramukhswami Medical College, Karamsad, Gujarat,India.
- Thrombocytapheresis was performed on a patient with essential thrombocythemia complicated by acquired von Willebrand syndrome.Informed consent for the procedure was obtained . The procedure was carried out using the Terumo BCT Spectra Optia Apheresis System in the Bone Marrow Transplant (BMT) unit. Prior to the procedure, the patient received pre-transfusion therapy with four units of cryoprecipitate due to the acquired von Willebrand syndrome.

RESULT

The thrombocytapheresis procedure was well tolerated by the patient with no adverse reactions. The platelet count decreased from 17.80 lakh/µL to 7 lakh/µL, representing a 58% reduction. Clinically, the patient showed resolution of the hematoma and bleeding episodes. The patient regained mobility, and was discharged four days after the apheresis procedure.

	PRE PROCEDURE INVESTIGATION	POST PROCEDURE INVESTIGATION
Total Leukocyte Count	80.2 x 1000/ul	61.9 x 1000/ul
Red Blood Cell Count	6.50 million/cmm	6.24 million/cmm
Haemoglobin	10.6 g/dl	10.4 g/dl
Haematocrit	40.2 %	38.5 %
Platelet Count	1780 x 1000/ul	700 x 1000/ul
PT	14.60 sec	13.30 sec
APTT	37.50 sec	35.20 sec
S. Creatine	0.96 mg/dl	0.96 mg/dl

Run values	Final
Collection preference	73
Packing Factor	20
Whole Blood Processed (ml)	7160
Run Time (min)	177
TBV Processed	2
Target Fluid Balance	106
AC (Anti Coagulant)	796
Inlet (ml)	7956
Collect (ml)	701



CONCLUSION

Thrombocytapheresis is an effective treatment for acute uncontrolled thrombocytosis. This case report shows rapid improvement in symptoms and resolution of thrombotic/hemorrhagic complications following apheresis procedure in timely manner.

REFERANCE

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