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A STRATEGIC APPROACH TO COMBAT HAEMOPHILIA –A DURING EMERGENCIES

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ABSTRACT :
Background : Haemophilia –A is an X-linked recessive inherited bleeding disorder due to a deficiency of coagulation factor- VIII resulting in increased mortality and morbidity[1].It has an estimated frequency of approximately one in 10,000 live births affecting mostly the males while the females are considered as carriers of haemophilia[2].
Aim : The purpose of the report is to present the evidence of a known haemophilia –A male patient who suffered from the consequences of an emergency (road accident).
Methodology: The familial history of the patient along with clinical manifestations was taken into account. Since he was a known case of haemophilic arthropathy, and a factor-VIII replacement was done as per the guidelines of World Federation of Haemophilia[3]. And the fracture of tibia was fixed with nailing and a good post-operative care was given.
Conclusion : The report concluded that health care professionals combined strategies right from administering medical treatment to rendering surgical support and it underlined the fact that with the timely medical intervention, the complications of haemophilia-A could be successfully managed.
Keywords: Haemophilia –A , factor -VIII , X- linked recessive , carriers , morbidity , mortality

INTRODUCTION:

Haemophilia –A is the most common haemorrhagic disorder due to deficiency of factor -VIII. The frequency of Haemophilia –A is 1 in 10,000 live births[1]. Haemophilia –A is more prevalent than Haemophilia – B. It is an X- linked recessive disorder. In this disorder, 100 % females born from affected fathers will be carriers whereas their male children have 50 % chance of getting affected and 50 % chance of becoming carriers[2]. Genes encoding the factor -VIII are located in the long arm of the chromosome – X and they are mutated in this disorder[3].Haemophilia -A which usually presents with prolonged bleeding time and patients have a family history of this disease[3,4]. It can occur in patients even without significant family history . Such a condition is called acquired haemophilia which is an autoimmune disorder[5] . It is an uncommon bleeding disorder following any injury or surgery. The severity of haemophilia is categorized into mild , moderate and severe as per the plasma factor –VIII levels.

CASE PRESENTATION

A 25 year old male patient, Mr.Perumal , who is a known case of Haemophilia – A on factor- VIII prophylaxis came to the casuality after sustaining a road traffic accident on 18.9.2024. The patient was put on a Thomas

splint .The patient had lost around 200 ml of blood after the injury and was given 2500 units of factor- VIII and bleeding was arrested. However , the patient was unable to move his right lower limb. With C- arm guidance , a fracture of tibia in right leg was found out and an implantation in the tibia was planned for the patient. Blood samples were taken as part of routine investigations from the patient and the bleeding time increased. So the patient was taken for the surgery and tibial nailing was done in right leg. Intra – operative blood loss was around 100 to 150 ml. The patient was again given 2000 units of factor- VIII post-operatively and was kept under observation .

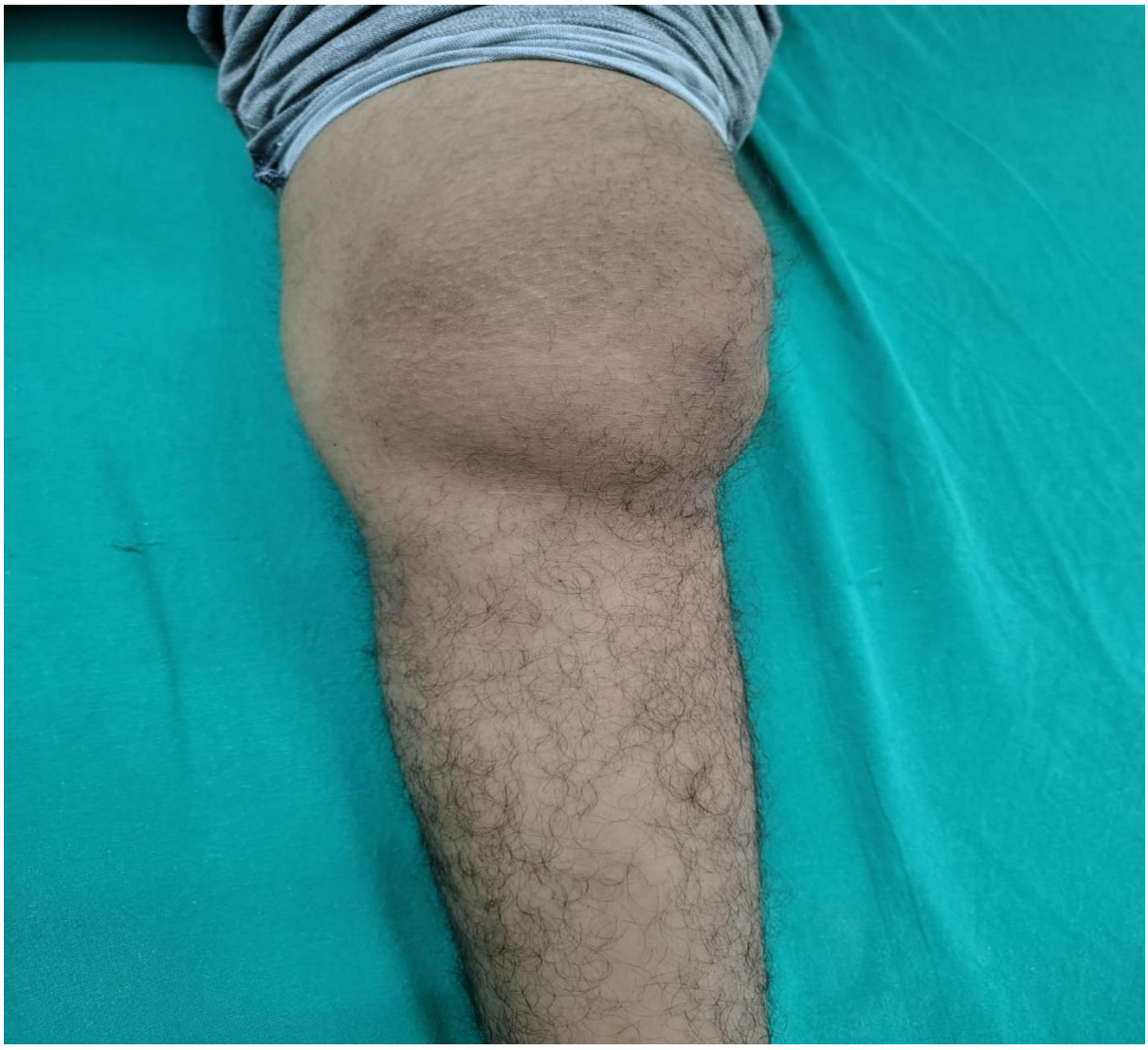

Figure -1 : Patient with swollen knee joint for factor –VIII replacement

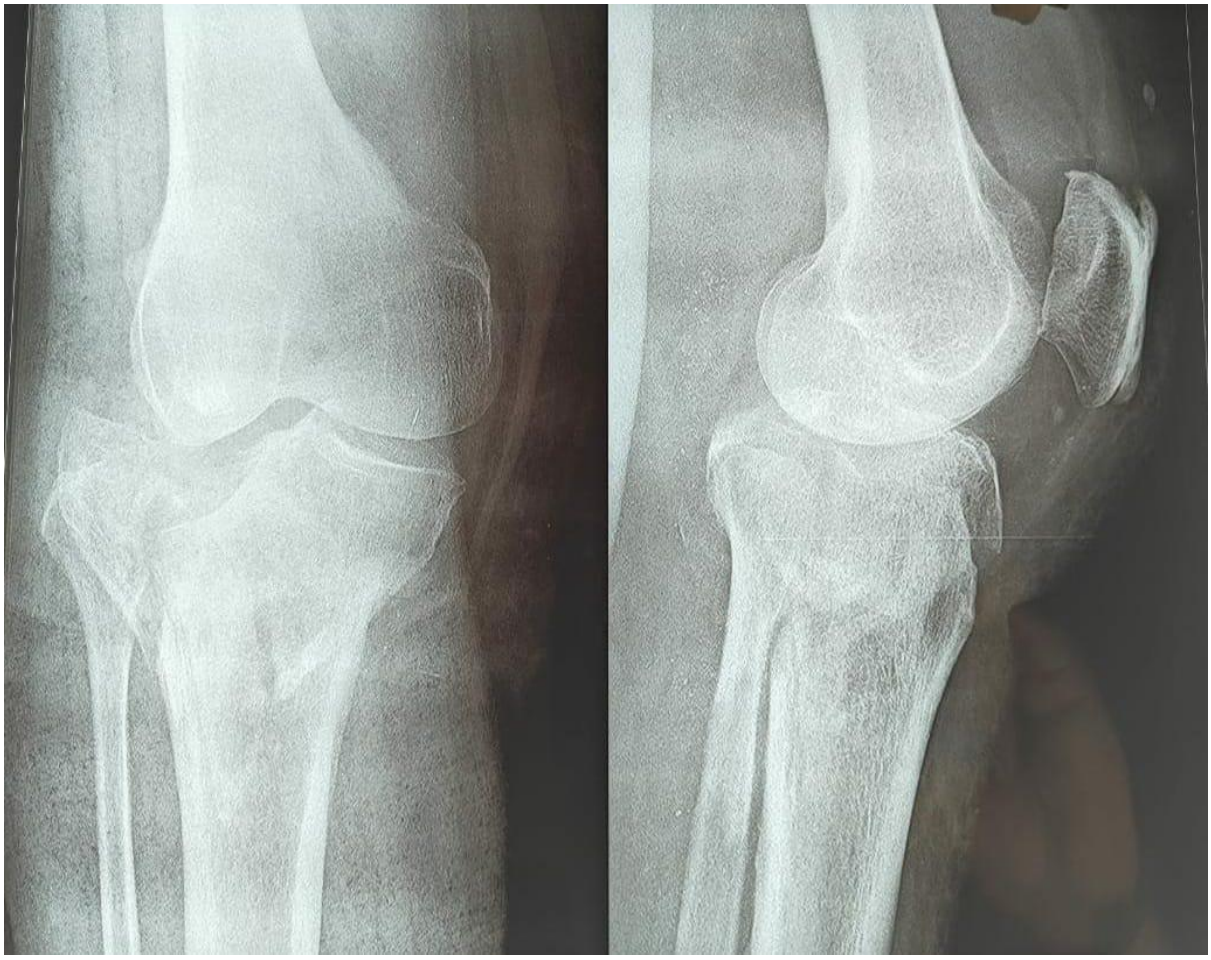

Figure -2 : Proximal tibia fracture after road traffic accident with the swollen knee joint

DISCUSSION:

Haemophilia-A , a congenital X – linked recessive bleeding disorder, results from a deficiency or dysfunction of coagulation factor -VIII[6]. It predominantly affects males and predisposes them to spontaneous and trauma induced bleeding which can be life threatening if not promptly managed. It is quite a challenge to the health care professionals. And it involves a multi-disciplinary approach to overcome this and stabilise the patient.In the present case, the patient had sustained a fracture in his right leg and it was found out that the patient had lost around 150 to 200 ml of blood. So the patient was given first aid using RICE approach i.e. Rest , Ice, Compression and Elevation of limbs[7,8]. An X- ray of the right limb was taken and it showed a fracture in tibia and POP was given for the fractured leg. Since the patient was a known case of haemophilia – A and he was given 2500 units of factor- VIII for attaining haemostasis . All the routine investigations were done and the patient was given a tibial nailing and was transfused intraoperatively 2000 units of factor- VIII to compensate the blood loss. The patient was kept in observation and was discharged with medications .The patient was on a regular followup.

CONCLUSION

The mortality risk of haemophilia-A is high if there is underdiagnosis and the consequent delay in the initiation of treatment[9,10]. Henceforth, a prompt diagnosis and a subsequent treatment are needed for a good prognosis in these patients. The present case report shows the significance of taking up an early intervention to render coagulation support to the patient along with a wholesome perioperative care . The report thus underlined the

fact that a multi-faceted approach is to be taken by the surgical and medical team for the factor- VIII replacement and a quick recovery of the patient.

REFERENCES

1.Mannucci PM, Tuddenham EG. The hemophilias--from royal genes to gene therapy. N Engl J Med. 2001;344(23):1773–9. https://doi.org/10.1056/NEJM200106073442307.
2.Clausen N, Petrini P, Claeyssens-Donadel S, Gouw SC, Liesner R, PedNet, et al. Similar bleeding phenotype in young children with haemophilia a or B: a cohort study. Haemophilia. 2014;20(6):747–55. https://doi.org/10.1111/hae.12470.
3.VandenDriessche T, Chuah MK. Hemophilia gene therapy: ready for prime time? Hum Gene Ther. 2017;28(11):1013–23. https://doi.org/10.1089/hum.2017.116.
4.Kurnik K, Bidlingmaier C, Olivieri M. How do I counsel parents of a newly diagnosed boy with Haemophilia a? Hamostaseologie. 2020;40(1):88–96. https://doi.org/10.1055/s-0039-3402805.
5. Mehta, P. and Reddivari, A.K.R. (2022) Hemophilia. StatPearls Publishing, Treasure Island, FL. https://www.ncbi.nlm.nih.gov/books/NBK551607/
6.Nagae C, Yamashita A, Ashikaga T, Mori M, Akita M, Kitsukawa K, et al. A cohort study of the usefulness of primary prophylaxis in patients with severe haemophilia a. Int J Hematol. 2016;104(2):208–15. https://doi.org/10.1007/s12185-016-2005-3.
7.Blanchette VS, Key NS, Ljung LR, Manco-Johnson MJ, van den Berg HM, Srivastava A, et al. Definitions in hemophilia: communication from the SSC of the ISTH. J Thromb Haemost. 2014;12(11):1935–9. https://doi.org/10.1111/jth.12672.
8.Lovgren KM, Sondergaard H, Skov S, Wiinberg B. Non-genetic risk factors in haemophilia a inhibitor management - the danger theory and the use of animal models. Haemophilia. 2016;22(5):657–66. https://doi.org/10.1111/hae.13075
9.van Velzen AS, Eckhardt CL, Streefkerk N, Peters M, Hart DP, Hamulyak K, et al. The incidence and treatment of bleeding episodes in non-severe haemophilia a patients with inhibitors. Thromb Haemost. 2016;115(03):543–50. https://doi.org/10.1160/th15-03-0212.
10.Srivastava A, Brewer AK, Mauser-Bunschoten EP, Key NS, Kitchen S, Llinas A, et al. Guidelines for the management of hemophilia. Haemophilia. 2013;19(1):e1–47. https://doi.org/10.1111/j.1365-2516.2012.02909.x.