

The Context of Bernard – Soulier Syndrome & It's Culmination in the Demense of Prosthodontics.

Abstract:

Bernard : Soulier syndrome is a rare hereditary disorder with giant cell platelets, thrombocytopenia and extended bleeding time. In thrombosis and hemostasis, platelets play a main role. The first stage in the creation of hemostatic plugs is the accumulation of platelets at the site of vascular injury that plays a key role in stopping the blood loss following the injury. The genetic error is induced by the glycoprotein complex Ib/IX/V, which is the Von Wille brand factor on the platelet surface. Here we are reporting a 78 year old male patient which comes with chief complaint of missing teeth, root stumps & worn out teeth in upper and lower arches, inability to chew properly and wants replacement for the same. The primary treatment remains platelet transfusion pre & post extraction. Root canal treatment was performed on remaining natural teeth. Conventional over denture prosthesis for upper arch and dolder bar attachment overlay denture was fabricated for the lower arch.

Keywords: Bernard – Soulier syndrome; Prosthodontic management; Dolder bar; Overdenture

Introduction:

The Bernard – Soulier syndrome (BSS) is an autosomal recessive disease associated with bleeding tendency, giant blood platelets and low platelet count. The defect is restricted to the megakaryocyte/platelet lineage.[1] In 1948 , Jean Bernard and Jean – Pierre Soulier, two French hematologist, described a young male patient who had a severe bleeding defect with a prolonged bleeding time, a low platelet count with very large platelets (macrothrombocytopenia), It is also known as “Dystrophie thromobocyttaire – hemorogipare congenitale” (Hemorrhagiparous thromobocytic dystrophy).[1] This is a very uncommon syndrome because in published papers, mostly in Japan, Europe, and North America, only 100 instances were recorded. The prevalence of < 1/1,000,000 was estimated.[2] Reported cases in India are 27 till date.3The inheritance mode is generally autosomal recessive with an autosomal dominant pattern seen in individual instances.

Defect in three genes give rise to the typical clinical features and platelet anomalies associated with BSS. This is due to the multisubunit nature of the affected GPIb-V-IX receptor,

whose structure is shown in (Fig.1). GPIb-V-IX complex main role is to guarantee ordinary primary hemostasis by initiating platelet adhesion at vascular injury locations.[4] Adhesion is carried to Von willebrand by its binding, which is caught by sub endothelial collagen from plasma.[5] Four distinct transmembrane proteins, GPIba (MW 135kDa), GPIb, (MW26kDa), GPIX (MW 20 kDa) and GPV (MWkDa) assemble to form the functional receptor at the surface of bone marrow megakaryocytes, the precursors of mature circulating platelets.[6] The related GPIba, GPIb, and GPIX are all needed for effective receptor biosynthesis.[7] The absence of a single subunit reduces the surface expression of the entire complex dramatically. Most instances are caused by defects in the sub – unit GPIb – alpha. The deficiency can be combined in qualitative or quantitative GPIb – alpha units.[8] The definitive diagnosis of BSS is done when inserted in an aggregometer by means of an isolated ristocetin –

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induced agglutination. Biochemically or by genotyping, diagnosis can be verified. In peripheral BSS smear, neutrophil inclusions are missing.[9] Thrombocytopenia, abnormal vWF platelet interaction, abnormal platelet interaction with thrombin and abnormal platelet coagulant activity may be contributors to BSS' hemorrhagic diathesis.

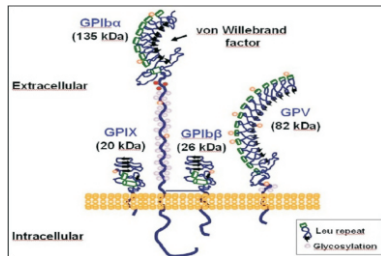


Fig. 1 .Platelet glycoprotein Ib-V-IX complex (courtesy of François Lanza Université Louis Pasteur, Strasbourg, France)

For any tooth and tissue supported removable prostheses the support from existing natural abutment is very important. Maligned and worn out present natural abutment put a question mark on the prognosis of the removable prostheses. Pre prosthetic surgical procedure is mandatory before starting any prosthetic rehabilitation to improve the foundation for the removable prostheses. Managing a patient with bleeding disorder requires meticulous planning and a multidisciplinary approach with special care. There is no case report till date documented in the literature for the prosthetic rehabilitation of the patient suffering from BSS.

Clinical Report:

A -78- year old patient reported to the department of prosthodontics with a chief complaint of inability to chew properly with his remaining natural teeth. The patient also complains of pain in upper and lower front & back region of mouth for two months. Intraoral examination shows mutilated and worn off existing natural teeth with missing 1[6,17,26,27,] root stumps with [13,14,17,23,26,34,37,46] region, caries with [11,12,15,24,28,31,32,34,41,42,43] region, Grade II mobility reported with 15,24,35,31,32,36 and Grade III mobility reported with 28,36 and 47 region which can also be seen in a OPG (Fig. 2).The patient was diagnosed with BSS twelve years back. Treatment plan include extraction of existing root stumps and grossly

decayed teeth, root canal of [11,12,21,22,33,43,44] regions. Prosthetic rehabilitation involves fabrication of maxillary coping supported overlay denture and mandibular Dolder bar supported overlay denture with soft reline.

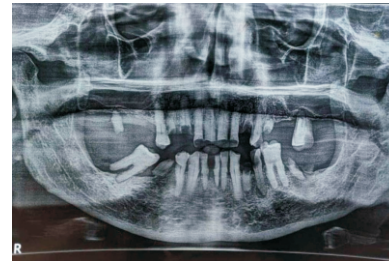


Fig.2. Orthopantomogram (OPG)

Medical management before dental treatment:

BSS patient generally needs a platelet concentrate supplement to regulate post-operative bleeding before dental invasive processes and tranexamic acid. It was instructed from the hematologist to give two units of platelet- rich plasma before and after extraction for post -operative healing. The hematology department's referring physician also recommends that iron dextran or iron sucrose (4 ampules) be given for thrombocytopenia and anemia. There was an acceptable increase in platelet and hemoglobin level which is illustrated in Table 1.



Fig. 3.Extraction of grossly decayed maxillary teeth



Fig. 4. Extraction of grossly decayed mandibular teeth

Hematology	Before Transfusion	After transfusion
Hemoglobin (g/dl)	10.0	11.1
Total leucocyte count (cells/cumm)	7400	6800
Mean corpuscular volume (fl)	87	86
Mean corpuscular hemoglobin (pg)	27.2	25
Platelet count(lakhs/cumm)	0.20 thousand/cumm	0.88 thousand/cumm
RBC(Millions/cumm)	4.33	4.5
PCV / Hematocrit (%)	37.60	30.3

Table 1. Hematology report of the patient before & after platelet rich plasma transfusion

After all investigations & consent from the patient, he was referred to the department of oral surgery for extraction of grossly decayed maxillary (Fig. 3) & mandibular teeth (Fig. 4) under medical supervision. Post healing patient was reevaluated and advice maxillary conventional and mandibular bar supported overdenture on existing upper and lower natural teeth [11,12,21,22,33,43,44] after root canal treatment. As there are bony irregularities found at [16,17,26,27,35,36,46] regions, the patient has been advised for application of the soft-liner in both upper and lower dentures to avoid any further surgical procedure.

After root canal treatment of the present teeth, crown preparation was done and the final impression was made using addition silicone putty & light body consistency material. Dolder bar pattern attachment to the wax pattern of 33 & 43 regions were done with the help of the surveyor. Coping trial and cementation of the same was done with glass ionomer luting cement in upper (Fig. 5) and lower jaw (Fig. 6). Preliminary impressions were made with irreversible hydrocolloid and models were poured with dental plaster. Special trays were fabricated using pattern resin. Greenstick compound was used to complete the peripheral tracing (Fig. 7) and final impressions were made using addition silicone light body consistency (Fig. 8), the master cast was poured using Type IV gypsum product. After the fabrication of temporary denture bases and occlusal rim, the jaw relation procedure was carried out in a usual manner. Teeth arrangement is done and anterior and posterior trial were done with patient consent. The disadvantage with overlay prostheses is excess exposure of the front teeth which has been explained to the patient. Then upper and lower overdenture insertion performed and a minor occlusal error has been corrected. Chairside soft relining performed in both the dentures (Fig. 9) and post-operative instruction regarding insertion and hygiene maintenance given to the patient (Fig. 10). The patient was asked to come after 24 hours for the first follow up visit and the second follow up was done after one-week post- insertion. It has been instructed to the patient to come for regular follow up every three months.



Fig. 5. Cementation of upper metal copings



Fig. 6. Cementation of lower metal copings with dolder bar



Fig. 7. Peripheral Tracing

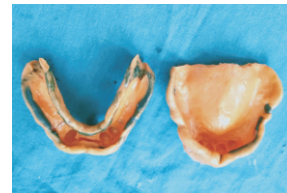


Fig. 8. Final Impressions



Fig. 9. Chair side soft-relining of both dentures



Fig. 10. Final insertion

Discussion:

Managing patient suffering from BSS is a very challenging task which also depends upon the severity of the disease and its invasiveness. In most instances, patients need to be transfused. Factor VIIa can also be used as an alternative medication system, but it must be supplemented with the

transfusion of platelets. One should keep in mind that the use of Factor VIIa can lead to thromboembolic events.¹⁰ If feasible, human leukocyte antigen matching should occur when platelet transfusion is scheduled. In some patients, the administration of desmopressin has been shown to shorten the bleeding time. BSS can be a candidate for potential gene replacement treatment with megakaryocyte progenitors that are virally transduced.¹¹ Spontaneous and recurrent gingival bleeding is caused by hyperemic gingiva. Periodontitis is a leading cause of tooth morbidity necessitating extractions. So in our case, intense care has been taken during extractions, endodontic therapy, and tooth preparation. Tranexamic acid for antifibrinolytic therapy avoids post-operative bleeding through inhibits of the plasminogen activation to plasmin and promotes the stability of the clots. In patients with inherent bleeding disorders, tranexamic acid is recommended for seven days after dental extractions.¹¹ Nerve block anesthesia is contraindicated in patients with BSS unless better alternatives are found and prophylaxis occurs as an anesthesia solution deposited in an extremely vascular region that carries a danger of formation of a hematoma.¹² The patients are preferred for endodontic therapy over - extraction whenever feasible. Endodontic therapy generally does not present a significant danger of bleeding and is regularly practicable.¹³ Taking into consideration the fragility of the mucosa and gingiva the finish line of the preparation is kept at equi - gingival level. To avoid any laceration a hemostatic gingival retraction cord is used before making an impression. Excess amount of pressure is avoided during peripheral tracing and final impression procedure. Removable prosthetic dentures can be fabricated without any complications. To enhance the retention of the prostheses we used a Dolder bar joint attachment with a header and runner bar. The use of dolder bar offers periodontally involved teeth an improved crown/root ratio and splinting of the teeth. Because the bar is near to the alveolar bone, chewing forces have a much lower impact on the teeth.¹⁴ The bar joints offer slight vertical and rotational movement of the denture as well as stress breaker action.¹⁴ Many studies have supported the use of bar supported overdenture, as practitioners are looking for simplified treatments that can provide cost – effective alternatives to more complex prosthodontics procedures.^{15–18}

Due to the presence of boney irregularities in the maxillary and mandibular edentulous ridge temporary soft liner is recommended to avoid any trauma to the underlying mucosa in a syndromic patient. Denture soft lining materials provide a spongy, cushioned interface between the hard base of a denture and the oral mucosa. They are used to resurface the tissue side of the removable dental prostheses with the new base material, thus producing an accurate adaptation to the denture foundation area.¹⁹ Parker reduced the effect of traumatic impact over the edentulous ridge by sandwiching a resilient liner layer within a denture base. This soft liner acts as a “shock absorber” and stress distributor.²⁰ Soft liners have also been shown to promote the growth of candida Albicans.²¹ Denture cleanliness is stressed to prevent fungal growth and subsequent staining and malodor.

Summary :

This clinical report explains the prosthodontic management of a patient suffering from Bernard – Soulier syndrome. Taking the support of the left natural teeth in these patients, a removable over denture with soft-liner is always a better treatment alternative for the patient. The use of dolder bar will aid in retention of the lower denture.

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