Bleeding Diathesis Diagnosed After Cleft Palate Repair

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Citation

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Abstract

Bleeding diathesis is a clinical definition in which patients have uncontrolled bleeding problems for different reasons. The most common etiologic factor for bleeding diathesis is Von Willebrand Disease followed by hemophilia. In the newborn period, patients with bleeding diathesis present as CNS bleeding or uncontrolled bleeding from the umbilical stump. In most cases, however, diagnosis is done later on after the development of more obvious signs of bleeding diathesis, such as easy bruising, increased menstrual or nasal bleeding and hemarthrosis. Other cases were diagnosed after an uncontrolled bleeding following a surgical intervention, such as tooth extraction, or an accident. In this case, bleeding diathesis was diagnosed following the repair of complete cleft palate. As the age of cleft palate repair decreases, it seems that more cases with bleeding diathesis and cleft lip / cleft palate will be diagnosed.

INTRODUCTION

Bleeding diathesis is the clinical final outcome of several different reasons. In the newborn period, most of the patients presented with CNS bleeding or uncontrolled bleeding from the umbilical stump. In older age groups, development of signs of bleeding diathesis, such as easy bruising, increased menstrual or nasal bleeding, hemarthrosis, uncontrolled bleeding after surgical interventions, such as tooth extraction or circumcision, or accidental injuries lead to the diagnosis the bleeding diathesis[1].

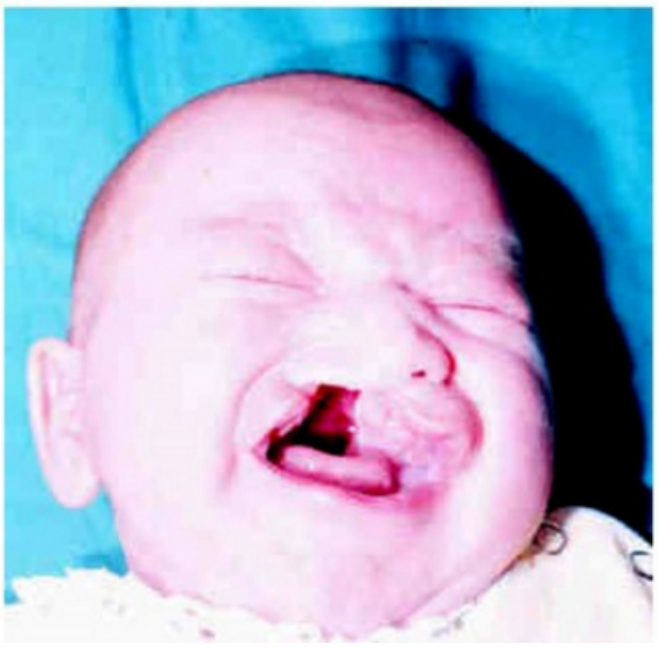
Although the incidences of cleft lip-cleft palate or isolated cleft palate differ from one nation to another (0.02-0.043% and 0.02-0.083%), figures for Turkey are 0.095% and 0.077% respectively[2, 3]. To our knowledge there is no reported bleeding diathesis case diagnosed after bleeding due to repair of cleft lip/cleft palate deformity. In our case, the clinical findings of bleeding diathesis observed also reoccurred in the circumcision of the same patient eight years later. Although none of the coagulation factor deficiencies could be demonstrated in the laboratory in both times.

CASE REPORT

A one and half year-old boy, who had been operated for repair of unilateral cleft lip at the age of three month, was readmitted for the repair of complete cleft palate (figure I).

Figure 1

Figure I: Patient with unilateral cleft lip and complete cleft palate at the age of three months



Although there was no any noticeable event in his history concerning bleeding diathesis, including the first surgical intervention for the cleft lip repair, the patient had uncontrollable bleeding in postoperative period of V-Y push back repair of cleft palate. Bleeding reoccurred several times from different locations (from rough surface of the palate, from major palatine artery and finally from mucosal surface of the uvula). Beside surgical intervention, complete blood

and fresh frozen plasma transfusions were needed for hemostasis.

The patient was evaluated by the Pediatric Hematology Department. Von Willebrand factor level, ristosetin cofactor, protrombin and partial thromboplastine time, bleeding time and thrombosit count on peripheral smear and complete blood count were found to be normal. All coagulation factor levels, including factor XIII, were within normal levels. There was no family history of bleeding diathesis. It was thought that the digestive enzymes of the saliva might interfere with the coagulation cascade in the mouth. An oral solution with -amino caproic acid was applied topically several times within the day, together with fresh frozen plasma transfusions, which helped to restore hemostasis and patient was discharged without any sequel in the 31st postoperative day.

The same patient was readmitted to our clinic for circumcision eight years later (figure II). His family informed us that he had a home accident and have a big cut in the scalp two years ago, which healed without any bleeding problem. All his hematological tests were controlled again and none of them revealed any finding of bleeding diathesis. Circumcision was done without any problem and the patient was discharged one day after the operation. However, the patient was hospitalized again due to the bleeding from phrenilum on 8th postoperative day. The bleeding was started when they removed the crust from the phrenilum. There was 2 sq. mm. rough surface on the phrenulum, probably the area under the crust that the patient removed, was the source of the bleeding. Following surgical homeostasis, Fibrin glue and compressive dressing was applied to the surface of the phrenulum[4]. He was followed up with fresh frozen plasma transfusions and opioid analgesics for five days and discharged without any sequel.

Figure 2

Figure II: Same patient with repaired cleft lip and palate at the age of eight year

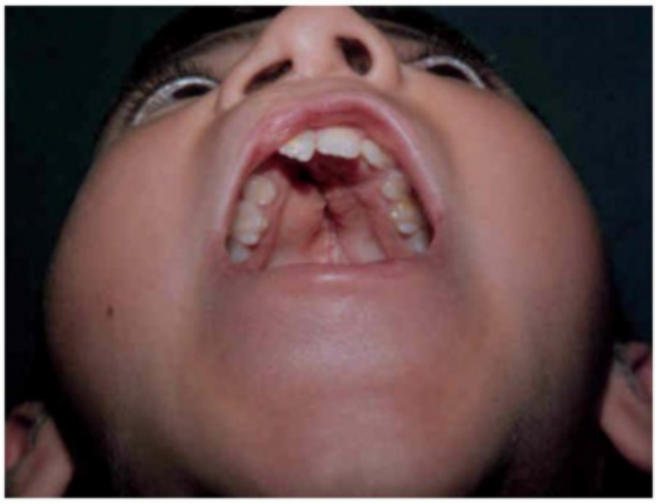


Figure 3

Figure III: Fibrin plug at the phrenulum, indicated with arrow

DISCUSSION

Hemostasis is a series of events, initiated by injury to vascular wall, leading to the deposition of platelets adhering to components of the subendothelium. Degranulation of preformed granular contents of these platelets reinforces the recruitment and adhesion of more platelets to the site of injury and therefore enlargement of the platelet plug. Vasoconstriction of injured vessel walls and beginning of the coagulation cascade that in turn ends up with conversion of fibrinogen to fibrin, which increases the platelet plug mass, strengthen and stabilize the plug. Therefore, coagulation is the end result of platelet aggregation, vasoconstriction and fibrin formation cascades. Although these events seem to be

separate sets of coagulation phenomenon, in fact there are several overlapping and control steps in these cascades. Therefore, a disorder of one of these cascades does not only upsets its own cascade but also disturbs or slow down the other cascades of the coagulation and clinical outcome is the prolonged and intractable bleeding.

There is no reported bleeding diathesis case diagnosed after bleeding due to an operation for repair of cleft lip/cleft palate deformity. As the age of cleft palate repair decreases, it seems that more cases with bleeding diathesis and cleft lip / cleft palate will be diagnosed. This case was presented with prolonged intractable bleeding problem, occurring not always but from time to time in different occasions (after cleft palate repair and circumcision). Our Hematology Department evaluated him for the differential diagnosis and treatment of bleeding diathesis. The entire laboratory tests, described in the literature for differential diagnosis on bleeding diathesis were performed[5], all the tests revealed all hematological parameters were within normal limits.

Although the etiology cannot be clarified exactly, history of the patient (development of bleeding diathesis after elective surgical interventions but no bleeding diathesis after an acute traumatic cut) was implying the presence of VWD. In mild form of VWD, adrenalin discharge in an acute trauma may lead to discharge of von Willebrand factor from vascular endothelium, and therefore prevent the bleeding but same patient may present the findings of bleeding diathesis later in a more elective surgery.

Only supportive treatment with fresh frozen plasma transfusions was applied. The fibrin formation seem to be intact after circumcision but the formed fibrin plug (figure III) could not manage to stop the ongoing bleeding. It is interesting that the bleeding problems occurred after cleft palate repair and circumcision. The common denominator is that both, uvula and penis are mobile and under tension in soft palate movements and during erection respectively. Therefore, opioid analgesics were preferred after circumcision to prevent the penile movement due to the erection.

Although all the coagulation factors levels were measured and found to be normal, there are reports in the literature that vascular disorders may lead to bleeding diathesis[6].

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