

Succesfull thyroid cancer surgery in Glanzmann's disease: report of a case

Glanzmann hastalığında başarılı tiroid kanser cerrahisi: bir olgu sunumu

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Summary

We present an uncommon case of thyroid cancer with Glanzmann's disease. A 54-year-old woman with symptoms like pain and mass in the lumbar region was investigated. Preoperative interventions revealed a definitive diagnosis of metastatic thyroid cancer. During the preoperative period she had an episode of recurrent epistaxis. Hematological research revealed Glanzmann's trombasthenia. Surgical excision of the thyroid was performed successfully without hemorrhagic complications.

Key words: *Glanzmann's disease, thyroidectomy, hemorrhagic complications*

Özet

Bu olgu sunumunda Glanzmann hastası olan sıra dışı bir tiroid kanser hastası sunulmaktadır. Yaşı 54 olan ve lomber bölgede ağrı ile şişlik yakınması olan bayan hasta incelemeye alındı. Ameliyat öncesi incelemelerle hastada metastatik tiroid kanserinin olduğu belirlendi. Ameliyata hazırlanan hastada, ameliyata alınmadan tekrarlayan epistaksis atakları oldu. Yapılan hematolojik incelemeler sonucunda hastaya Glanzmann trombasteni'si tanısı kondu. Tiroid bezinin cerrahi eksizyonu, herhangi bir kanama komplikasyonu olmadan, başarıyla gerçekleştirildi.

Anahtar kelimeler: *Glanzmann hastalığı, tiroidektomi, kanama komplikasyonları*

Introduction

Glanzmann's trombasthenia is a rare autosomal-recessive disorder of platelet aggregation characterized by a lifelong bleeding tendency due to abnormalities of the glycoprotein (GP) IIb-IIIa membrane complex (1). It is rare in a global context, but a relatively more common platelet function defect in communities where consanguineous marriages are more frequent (2).

In 1918 Glanzmann, who was a Swiss pediatrician, described a group of patients with normal platelet count, impaired clot retraction and prolonged bleeding time (3). Common clinical manifestations include prolonged mucocutaneous bleeding, epistaxis, menorrhage and gingival bleeding. Spontaneous bleeding is uncommon, but posttraumatic and postoperative hemorrhage may be particularly serious, causing significant morbidity (1).

Essential diagnostic features are a normal platelet count and morphology, a greatly prolonged bleeding time, absence of platelet aggregation in response to agonists like adenosine diphosphate, collagen, epinephrine, and thrombin. In response to ristocetin or bovine Von Willebrand factor, platelet aggregation occurs normally (2). The only available curative treatment for Glanzmann trombasthenia is bone marrow transplantation (2). Prophylactic and therapeutic platelet transfusion is the cornerstone of supportive treatment. Recombinant activated factor VIIa has recently been introduced as an alternative to platelet transfusion for treating bleeding episodes and to cover surgery in patients with hereditary platelet function defects (4). In this study, a clinical case of a patient with Glanzmann's trombasthenia who underwent successful thyroid surgery due to thyroid cancer is reported.

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Case Report

A 54-year-old woman was admitted to our hospital because of mass and pain in the right lumbar soft tissue of 6-months duration. The patient had no significant prior illnesses, except a history of subtotal thyroidectomy 12 years ago performed at a clinic elsewhere. She reported no bleeding episodes in her life or after her first thyroid surgery. Her family history revealed no consanguineous marriages. On admission, physical examination showed a grade II nodular goiter and right lumbar soft tissue mass, while laboratory findings revealed without particularity.

Hematological tests were as follow; hemoglobin=12gr/dL, leukocytes=4300/mm³, platelets=275.000/mm³, prothrombin time=13,9 seconds, INR=1.09 and bleeding time=6 minutes. She had an attack of spontaneous recurrent epistaxis before the operation. Her hematological markers, including thrombocyte morphology were normal. Thrombocyte aggregation test revealed agglutination rates as follow: adenosine diphosphate 26%, collagen 38%, epinefrin 16% and ristocetin 96%. These results suggested Glanzmann's thrombasthenia.

The abdominal magnetic resonance imaging confirmed the presence of the mass by revealing right iliac bone metastatic destruction. Bone scintigraphy showed iliac and sacral metastasis. She underwent a tru-cut biopsy from the mass, which concluded metastasis of a thyroid carcinoma. The initial diagnosis was a stage IV thyroid cancer and surgical excision was planned.

Before surgery, two units of thrombocyte apheresis were prepared, but only 1 unit was transfused. No support treatment with aminocaproic acid or Factor VIIa was performed. A total thyroidectomy procedure was performed without any complication. Postsurgical bleeding was not seen. Frozen section of the tissue demonstrated malignancy and gross examination showed a tumor focus 3,5 in size. Histopathologically, the tissue specimen revealed a follicular variant of papillary carcinoma. The patient had an uneventful postoperative course and the haemovac drain collected 40 ml hemorrhage during the first 24 hours. She was discharged 3 days postoperatively and was referred to the nuclear medicine department for radioactive iodine therapy.

References

1. Van Buuren HR, Wielenga JJ. Successful surgery using recombinant factor VIIa for recurrent, idiopathic nonulcer duodenal bleeding in a patient with Glanzmann's thrombasthenia; *Dig Dis Sci.* 2002;47:2134-6
2. Nair S Ghosh K, Kulkarni B, Shetty S, Mohanty D. Glanzmann's thrombasthenia: updated. *Platelets.* 2002;13:387-93
3. Glanzmann E. Hereditaire hamorrhagische thrombasthenic .Ein Beitrag zur pathologie der Blutplattchen. *Jahrbuch Kinderheilkde.* 1918;88:1-42

Discussion

Glanzmann's thrombasthenia is a functional disorder of platelets (2) and as far as we know has not been the subject in the field of thyroid surgery. Patients with this hematologic disorder may present with symptoms like mucosal bleedings, epistaxis, petechias, purpuras, menorrhages and gastrointestinal bleeding (5). In this case, only the presence of epistaxis and anemia before surgery was noted.

Laboratory tests are required for the diagnosis, where platelet aggregation is of utmost importance; a platelet aggregation test normal to ristocetin is essential. With this disorder, bleeding can be an extremely serious problem and also difficult to solve. Because of this possible clinical complication, platelet transfusions, administration of antifibrinolytic agents and local hemostatic substances are recommended during surgical procedures (6,7). Sometimes conventional treatment can not stop the bleeding (8). Activated coagulation Factor VII which is produced by recombinant DNA technology using a hamster cell kidney line, is structurally very similar to human plasma-derived activated Factor VII. Thus, administration of rF VIIa seems to result in local thrombin formation at the site of bleeding and to enhancement of the natural physiological coagulation cascade leading to hemostasis. rF VIIa has been used mainly for bleeding in patients with hemophilia and inhibitors of factor VIII or IX (7,9). Number of reports indicates that it may also be useful as prophylaxis during surgery and against bleeding in Glanzmann's thrombasthenia (10), Bernard-Soulier syndrome (4) and various acquired thrombocytopathies (11).

According to two studies, blood products may induce self-immunization or form isoantibodies against GP IIb-IIIa, promoting bleeding in patients with Glanzmann's disease (6, 12). Our case with Glanzmann's thrombasthenia demanded only one unit platelet-concentrate transfusion preoperatively and thyroid surgery resulted in no bleeding and no signs of immune reactions. In case of major surgery, platelet transfusions can be used if necessary. Whether a rF VIIa administration is required should be evaluated in future studies. Loyalty to basic surgical principles with minimal preoperative cautions provides a safe surgery in the thyroid patient with Glanzmann's thrombasthenia.

4. Monroe DM, Hoffmann M, Allen GA, Roberts HR. The factor VII-platelet interplay: effectiveness of recombinant factor VIIa in the treatment of bleeding in severe thrombocytopathia. *Semin Thromb Hemost.* 2000;26:373-7
5. French DL, Seligsohn U. Platelet glycoprotein IIb/IIIa receptors and Glanzmann's thrombasthenia. *Am Heart Assoc* 2000;20:607-10
6. Kirita T, Okamoto M, Horuici K, Sugimura M. Management of a deep space infection of the neck in patient with Glanzmann's thrombasthenia. *J Oral Maxillofac Surg* 2997;55:882-5
7. Rodriguez-Merchan EC, de la Corte H. Orthopaedic surgery in haemophiliac patients with inhibitors: a review of the literature. In: Rodriguez-Merchan EC, Goddard NJ, Lee CA, eds. *Musculoskeletal Aspects of Haemophilia.* Oxford: Blackwell, 2000: 136-42.
8. Chuansumrit A, Sangkapreecha C, Hathirat P. Successful epistaxis control in a patient with Glanzmann's thrombasthenia by increased bolus injection dose of recombinant factor VIIa. *Thromb Haemost.* 1999;82:1778
9. Lusher JM. Recombinant Factor VIIa (NovoSeven) in the treatment of internal bleeding in patients with Factor VIII and IX inhibitors. *Haemostasis.* 1996;26 (suppl 1):124-30
10. Tengborn L, Petruson B. A patient with Glanzmann thrombasthenia and epistaxis successfully treated with recombinant factor VIIa. *Thromb Haemost.* 1996; 75:981-82
11. Meijer K, Sieders E. Effective treatment of severe bleeding due to acquired thrombocytopathia by single dose administration of activated recombinant factor VII. *Thromb Haemost.* 1998;80:204-5
12. Rakocz M, Lavie G, Martinowitz U. Glanzmann's thrombasthenia: the use of autologous fibrin glue in tooth extractions. *J Dent Child* 1995;62:129-31