Postoperative Incisional Bleeding Due To Amyloidosis: A Case Report

M. Sinan BEKSAÇ, Tülay ÖZLÜ, Doruk KATLAN
Ankara-Turkey

Here, we represent a case of a 36 years old woman who had massive bleeding from the site of cesarean incision on the same day with the surgery. She was known to have Familial Mediterranean Fever (FMF) for 22 years and a renal biopsy performed 10 years ago confirmed a diagnosis of renal amyloidosis.

(Gynecol Obstet Reprod Med 2006; 12:000-000)

Key Words: Amyloidosis, Pregnancy, Cesarean section, İncisional hemorhorgia

Case Report

This was a 36 years old women who admitted to our outpatient clinics at 9th weeks of gestation of her second pregnancy for routine antenatal follow up. She had one previous abortion 6 years ago. She was diagnosed to have FMF 22 years ago and was using colchicine since then. A renal biopsy performed 10 years ago confirmed a diagnosis of renal amyloidosis. On admission, her biochemistry showed a creatinine value of 1.6 mg/dl and a blood urea nitrogen value of 25 mg/dl. A 24 hr urine sample showed 4300 milligrams/day of proteinuria. She was evaluated together with the nephrology unit. She had hypertension and therapy with alpha-methyl dopa was initiated. During her follow up, she tended to be hypertensive in spite of increased dosages of alpha methyl dopa. After the 22nd gestational week she was hospitalized to our high risk pregnancy service and nifedipine was added her antihypertensive therapy. She continued colchicine therapy throughout her pregnancy. She had protein S and protein C deficiency and used enoxaparin sodium 2000 units/day after the 20th weeks of gestation. At 30 weeks of gestation she had severe hypertension, nausea, vomiting. She also had effective contractions but did not have any cervical dilatation on pelvic examination. She was thought to have superimposed preeclampsia and MgSO4 therapy was initiated for prophylaxis of eclampsia and for tocolysis. On follow up the fetal heart rate pattern became non reassuring and she was taken to emergency cesarean delivery. 1780 grams of male infant with an apgar score of 4-7-8 was delivered. At the 6th to 8th postoperative hours, leakage of some blood beneath the cover of her incision was noticed. Her vital signs showed mild tachycardia. On follow-up, her tachycardia worsened and her hemoglobin level decreased from 8.8 to 4.9. Since she had hyperkalemia and disturbed renal function tests blood replacement was done (4 units) in accompany with hemodialysis through a femoral catheter. She was also given 12 units of fresh frozen plasma. But her bleeding did not stop and her tachycardia did not resolve. As a result she was taken to a revision surgery. Revision showed that the origin of bleeding was the subcutaneous tissues. Local hemostasis was maintained after the amyloid deposits at the incision site was debrided and the incision was reclosed after inserting a haemovac drainage system. Retansion sutures (Ethicon Prolene/0, USA) were also used to support the incision. After the surgery, slight bleeding continued a few days more and than stopped. The drain was removed after 10 days. Her activated partial tromboplastin time and prothrombin time were normal and she did not have bleeding from any other site in her body. Also the factors V, VII and X were measured to be at normal levels.

Hacettepe University medical Faculty Department of Obstetrics and Gynecology, , Ankara, Turkey

Address of Correspondence *Tülay Özlü*
Öncebeci Mh. Cevher Sk. 5/5
06420-Cebeci-Ankara-Turkey

Submitted for Publication: 28.02.2006
Accepted for Publication: 10.03.2006

Discussion

Amyloidosis is the intercellular deposition of a proteinous substance in certain clinical situations. It refers to extracellular deposition of insoluble protein fibrils. These fibrils are associated with other proteins and glycosoaminoglycans. The amyloid fibrils have a typical electron microscopic appearance consisting of polypeptides arranged in a twisted ß-pleated sheet. The polypeptide component varies with different sub-types of amyloidosis. The protein precursor accumulating in FMF related amyloidosis is serum amyloid A (SAA).

Amyloidosis associated with FMF is the only form of systemic amyloidosis known to be inherited as a recessive trait.[1] Like other forms of AA amyloidosis nephropathy is the most important clinical feature and a significant cause of death. Colchicine is the drug of choice in the prevention of amyloidosis in FMF patients.

Systemic amyloidosis may often be complicated with hemorrhagic tendency. This may be caused by factor deficiencies, hyperfibrinolysis and vasculopathy. A role of a platelet aggregation defect in hemorrhagic diathesis complicating systemic amyloidosis was also mentioned. It was concluded that desmopressin may therefore benefit patients with this disease in case of bleeding and before surgical interventions.[2]

Acquired coagulopathies, especially factor X deficiency, are common in patients with primary amyloid light-chain

amyloidosis. Factor X deficiency is the most common coagulopathy associated with life-threatening hemorrhagic complications when surgery is indicated. Recombinant human factor VIIa and Bebulin may allow for successful perioperative management of bleeding disorders in patients with primary amyloidosis.[3]

Another commonly observed clinical problem related with amyloidosis is intracranial hemorrhage. Cerebral amyloid angiopathy is one of the most common chronic vascular diseases that lead to intracranial hemorrhage (the other one is chronic hypertension).[4]

Examination of blood clot evacuation specimens in cases of intracranial hemorrhage can lead to a discovery of the etiology of the hemorrhage in a subset of cases, particularly if neural tissue is part of the specimen. The routine use of Congo red stain in all adult cases in which brain tissue is present proved to be of diagnostic utility in screening for amyloidosis.[5]

Perivascular deposition of amyloid may cause vascular fragility.[6] In our case, the patient had bleeding from the incision site from the subcutaneous vessels. Probably due to the vascular fragility caused by amyloidosis these vessels at the incision site started to bleed again in spite of a good surgical hemostasis. Local hemostasis was maintained, the amyloid deposit at the incision site was debrided and the incision was reclosed after inserting a haemovac drainage system. After the surgery, slight bleeding continued a few days more and than stopped. Factor V, VII and X levels were checked and were found to be normal in our case.

Just like this case, we have to be alert for the hemorrhagic complications that can be expected to occur in all types of amyloidosis patients that will undergo surgery. Maybe it will be a good approach to do a debridement at the incision site and to insert a subcutaneous drainage system before closing the incision to prevent formation of incisional hematomas.

References

1. Pras M, Bronshipigel N, Zemer D, Gafin J. Variable incidence of amyloidosis in familial mediterranean fever among different ethnic groups. The John Hopkins Medical Journal 1982; 150:22-6.

2. Demiroglu H, Barista I, Gursoy M, Oymak O, Dundar S. The effect of desmopressin on platelet aggregation defect in systemic amyloidosis: a preliminary report. Eur J Haematol 1996; 56:283-6.

3. Kazuaki Takabe, M.D, Ph.D, Peter R. Holman, M.D, Kenneth D. Herbst, M.D, Catherine A. Glass, R.N, Michael Bouvet, M.D. Successful Perioperative Management of Factor X Deficiency Associated With Primary Amyloidosis. J Gastrointest Surg 2004;8:358-62

4. Badjatia N, Rosand J. Intracerebral hemorrhage. Neurologist 2005; 11:311-24.

5. Neil A. Abrahams and Richard A. Prayson . The role of histopathologic examination of intracranial blood clots removed for hemorrhage of unknown etiology: A clinical pathologic analysis of 31 cases. Ann Diagn Pathol 2000; 4:361-6.

6. Hoshino Y, Hatake K, Muroi K, et al. Bleeding tendency caused by the deposit of amyloid substance in the perivascular region. Intern Med. 1993; 32:879-81.