

A case of Hemophilia A presenting with paraparesis following lumbar puncture

Lomber ponksiyon sonrası paraparezi ile belirti veren Hemofili A olgusu

Cahide Yılmaz¹, Fatmagül Başarslan¹, Ahmet Sami Güven², Hüseyin Çaksen², Ahmet Faik Öner²,
Nebi Yılmaz³

¹Department of Pediatrics, Mustafa Kemal University Faculty of Medicine, Hatay, Turkey

²Department of Pediatrics, Yüzüncü Yıl University Faculty of Medicine, Van, Turkey

³Department of Neurosurgery, Mustafa Kemal University Faculty of Medicine, Hatay, Turkey

Geliş Tarihi / Received: 05.08.2011 Kabul Tarihi / Accepted: 29.09.2011

ABSTRACT

Besides large intra-joint bleedings that are frequently observed in Hemophilia A, bleeding may also rarely occur in spinal joints. Additional to traumatic or spontaneous hematomas, cord suppression may be occurred due to various reasons such as epidural tumor and infections, myelosclerosis and bone tissue suppression that occurs to the enlargement in the bone marrow because of hemolytic anemia and thalassemia. In the present study a 6 months old male who developed paraparesis as a result of spinal compression of a hematoma that occurred after lumbar puncture and then diagnosed with Hemophilia A presented on account of the present case, our aim is to emphasize that perispinal hematoma may lead to paraparesis and that paraparesis may develop due to neurological retention that is rarely seen in children with Hemophilia A.

Key words: Hemophilia, Paraparesis, Children, Spinal Puncture

INTRODUCTION

Hemophilia is a hereditary hemorrhagic disease caused by deficiency of clotting factors.¹ The condition possesses a recessive inheritance due to the X chromosome which develops due to the Hemophilia A Factor 8. Central nervous system bleeding in Hemophilia A is quite frequent whereas intraspinal bleeding is very unusual.^{1,2} Intraspinal hematoma is a rare complication of lumbar puncture; however it occurs more frequently in patients with a bleeding diathesis and may display miscellaneous clinical symptoms that varies from back pain to paraplegia.²

ÖZET

Hemofili A da büyük eklem içi kanamalar sık görülmesinin yanısıra, nadiren spinal eklemlerde de oluşabilir. Travmatik ya da spontan hematomların yanısıra kordun baskılanması, epidural tümör ve infeksiyonlar, myeloskleroz, hemolitik anemi ve talasemi gibi kemik iliğinde büyüme yaparak kemik dokuyu bastıran pek çok nedenlerle oluşabilir. Bu çalışmada lomber ponksiyon sonrası gelişen hematomun yaptığı spinal kompresyon sonucu paraparezi gelişen 6 aylık erkek olgunun yapılan incelemesi ile ilk Hemofili A tanısı konulması üzerine sunuldu. Amacımız bu vaka ile perispinal hematomun parapareziye yol açabileceğini ve hemofili A olgularında görülen nörolojik gerilemenin buna bağlı olabileceğini vurgulamaktır.

Anahtar kelimeler: Hemofili, paraparezi, çocuk, lomber ponksiyon

In this study a Hemophilia A case who developed paraplegia paraspinal hematoma following lumbar puncture (LP) is presented.

CASE

A six months old male was referred to our clinic because of the complaints including cough, fever, dispne, edema on the back and thigh and unable to stand. The patient was examined by a physician 5 days before due to the symptoms of cough episode that continued for a period of 2 weeks and fever that continued for a period of 1 week. LP was applied to

Yazışma Adresi /Correspondence: Dr. Fatmagül Başarslan

Mustafa Kemal University Faculty of Medicine, Department of Pediatrics, Hatay, Turkey Email: fatmagulbasarslan@hotmail.com
Copyright © Dicle Tıp Dergisi 2011, Her hakkı saklıdır / All rights reserved

the patient, nevertheless echymosis and edema was observed at the LP site nearly 1 hour after LP, and then the patient failed to step on his feet. According to the information obtained from the personal history the patient successfully began to hold his head upright at the 2nd month and was able to sit and stand up without support; but unfortunately his male sibling died because of uninterrupted bleeding after a circumcision procedure which was performed when he was 1.5 years old.

The patient's physical examination revealed the following: Body weight and height were measured respectively, 10 kg (97th percentile) and 70 cm (90th percentile) respectively. General condition was moderate, body temperature VS: 37°C, Pulse: 150/minute, respiratory rate: 48/minute, blood pressure: 120/Pulses. Thorax examination displayed sub costal regressions and bilateral rale were present during auscultation. The liver was palpable 2-3cm. A mass (hematoma) was present within the muscle 2x3cm on femoral. A hematoma was found at dimensions of 5x4cm on the lumbar region. Deep tendon reflexes on the lower extremities were also reduced. The muscle strength on the right lower extremity was 4/5 and 3/5 at the left lower extremity. Deep tendon reflexes and muscle strength on the upper extremities were normal. Plantar response was bilaterally unconcerned.

Laboratory studies revealed the following results: Hemoglobin: 6.2 g/dL, hematocrit: 18% (32%), white blood count: 15.300/mm³, Platelet count: 265.000/mm³, prothrombin time (PT): 14.7 sec, active partial thromboplastin time (aPTT): 67.6 sec, bleeding time: >20 min. Factor 8 level was 1%. Other factor levels and routine biochemical studies were normal. Staphylococcus epidermis cultured from in the blood culture. Para cardiac pneumonic infiltration was present on the X- ray graphy. Abdominal USG was normal.

The patient was hospitalized with the diagnosis is of bleeding diathesis etiology, bronchopneumonia and paraparesis. Antibioterapy was initiated, a 10 cc/kg erythrocyte suspension was administered. Fresh frozen plasma was also given administered to the patient every other day till the factor level increased whereas the aPTT level was high. The patient received 0.6 mg/kg/day of 4 dose dexamethasone for a period of 1 week due to the hematoma located at the spinal region. The patient a surgical

intervention and he was discharged on day 16 with recovery after the paraparesis had improved. Now the patient is regularly followed with Factor 8 infusions and he is symptom-free.

DISCUSSION

Hemophilia A is the most frequently seen coagulopathy¹. Hemophilia occurs due to deficiency of factor 8, and is commonly presented with hemarthrosis form³. Hemophilic patients might be in need of several elective or emergency procedures during their entire life. Surgical procedures must never be applied to these patients without factor replacement⁴. Intraspinial bleeding is a rare condition in hemophilic patients. In a study among 1410 hemophilia patients carried out by De Tezanos Pinto et al, intraspinal bleeding was reported only in two cases⁵. In another study, 6 out of 2500 hemophilic patients displayed intraspinal bleeding.³ In a study performed by Eftekhari et al, a 9 years old patient with no any background of trauma was diagnosed with Hemophilia A due to quadriplegia, confusion and meningismus.¹ In a study performed by Faillace et al, a 3 months old infant was diagnosed with Hemophilia A due to paraparesis that developed after lumbar puncture because of sepsis.⁶ In our patient paraparesis developed after lumbar puncture and he was diagnosed with hemophilia A based on the laboratory findings.

In a study performed by Domenicucci et al, there were many patients among non-traumatic acute spinal subdural hematoma cases with a bleeding diathesis or patients without a lumbar puncture⁷. Clinical symptoms of intraspinal hematoma may vary from waist pain to paraplegia. It may be helpful to determine the existence of computed tomography and magnetic resonance imaging.⁶ In a study performed by Harve et al, tetraparaparesis developed in a hemophilia A patient due to spontaneous intraspinal bleeding, and myelography displayed a lesion extending to the extradural area while this patient responded dramatically well to a high dose factor 8 infusion.⁸

In the study of Freger et al, it was demonstrated that only 4 out of 11 hemophilic patients developed intraspinal epidural hematoma due to mechanic trauma whereas surgical intervention was applied to these patients after factor replacement.⁹ Surgical intervention may be necessary during therapy,

however some cases may recover by conservative treatment.⁵ In our case we assumed that paraparesis developed due to hematoma as a result of lumbar puncture causing suppression to the paraspinal structure and as a result of edema that developed at the medulla spinalis. The paraparesis in our patient recovered by conservative therapy and surgical intervention did not require.

Consequently, on account of this case, we would like to assume that children with Hemophilia A may refer to clinics due to rare neurological complications such as paraparesis, and therefore we must emphasize that it is very important to investigate the patient with a paraspinal hematoma for bleeding diathesis which may be the cause of paraparesis.

REFERENCES

1. Eftekhari B, Ghodsi M, Ketabchi E, Bakhtiari A, Mostajabi P. Spinal subdural hematoma revealing hemophilia A in a child: A case report. *BMC Blood Disord* 2003; 3(1): 2.
2. Pai SB, Krishna KN, Chandrashekar S. Post lumbar puncture spinal subarachnoid hematoma causing paraplegia: a short report. *Neurol India* 2002;50(3): 367-9.
3. Eyster ME, Gill FM, Blatt PM, Hilgartner MW, Mallard JO, Kinney TR. Central nervous system bleeding in hemophiliacs. *Blood* 1978;51(6): 1179-88.
4. Akçay A, Ünüvar Ö. Preparation of patients with hemophilia for minor and major surgery. *Türkiye Klinikleri J Int Med Sci* 2006; 2(2):15-9.
5. de Tezanos Pinto M, Fernandez J, Perez Bianco PR. Update of 156 episodes of central nervous system bleeding in hemophiliacs. *Haemostasis* 1992; 22(5): 259-67.
6. Faillace WJ, Warrier I, Canady AI. Paraplegia after lumbar puncture. In an infant with previously undiagnosed hemophilia A. Treatment and peri-operative considerations. *Clin Pediatr (Phila)* 1989; 28(3): 136-8.
7. Domenicucci M, Ramieri A, Ciappeta P, Delfino R. Nontraumatic acute spinal subdural hematoma: report of five cases and review of the literature. *J Neurosurg* 1999; 91(1): 65-73.
8. Harvie A, Lowe GD, Forbes CD, Prentice CR, Turner J. Intraspinal bleeding in haemophilia: successful treatment with factor VIII concentrate. *J Neurol Neurosurg Psychiatry* 1977; 40(12): 1220-3.
9. Freger P, Meneses M, Creissard P, Vanier JP, Tadie M, Godlewski J. Intraspinal epidural hematoma in the hemophiliac. *Neurochirurgie* 1986; 32(6): 486-9.