

Spinal anesthesia for a rare case of Proteus syndrome

Nadir görülen Proteus sendromlu olguda spinal anestezi

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Abstract

Our purpose is to present an uneventful spinal anesthesia experience in a rare case with Proteus syndrome (PS) suffering from urolithiasis. A 17-year-old male patient with previously diagnosed PS admitted to the urology polyclinic for urolithiasis. Physical examination showed disproportionate growth of the lower extremities, amputated toes and superficial and large veins in the legs. Spinal anesthesia was successfully administered at sitting position through the intrathecal route via the L₃₋₄ space in the midline. Since it also includes difficult airways, spinal canal deformities, thromboembolisms, and certain pathologies such as cystic abnormalities in the lung, emphysema, atelectasis and fibrosis, this syndrome should be considered further with respect to anesthesia. The most significant cause of mortality in PS is pulmonary thromboembolism induced by deep vein thrombosis caused by growing extremities. Spinal anesthesia has provided a more reliable method of anesthesia due to the lower risk of development of thromboembolism in this case.

Keywords: Proteus syndrome; spinal anesthesia; urolithiasis

Özet

Amacımız ürolitiazisi olan ve nadir görülen Proteus sendromlu (PS) bir olguda sorunsuz olan spinal anestezi deneyimimizi sunmaktır. On yedi yaşında önceden tanı almış PS olan erkek hasta ürolitiazis nedeniyle üroloji polikliniğine başvurdu. Fizik muayenesinde alt ekstremitelerde orantısız büyüme, ampute ayak parmakları ve bacak venlerinin yüzeysel ve büyük olduğu görüldü. Hastaya başarılı bir şekilde oturur pozisyonda, orta hat L₃₋₄ aralığından spinal anestezi uygulandı. Zor hava yolu, spinal kanal deformiteleri, tromboembolizm, ve akciğerde kistik anomaliler, amfizem, ateletazi ve fibrozis gibi patolojilerin eşlik etmesi bu sendromun anestezi açısından daha çok dikkate alınmasını gerektirmektedir. PS'da en önemli mortalite nedeni büyüyen ekstremitelerden kaynaklanan derin ven trombozuna bağlı gelişen pulmoner tromboembolilerdir. Spinal anestezinin tromboemboli gelişiminde daha az risk taşıması bu olguda bize daha güvenilir bir anestezi yönetimi sağlamıştır.

Anahtar kelimeler: Proteus sendromu; spinal anestezi; ürolitiazis

Introduction

Proteus syndrome (PS) is a sporadic, multi-systemic disease characterized by the overgrowth of various parts of the body in an asymmetrical and disproportionate manner accompanied by the skeletal system disorders (1). The incidence of the disease is less than 1/1 000 000 live births. Although its exact cause is unknown, this disease is sporadic and genetically involves all blastoderms, several findings on PS can be defined. Here, we report on a patient with PS for whom we administered spinal anesthesia due to urolithiasis.

Case Report

A 17-year-old male patient with previously diagnosed PS admitted to the urology polyclinic for pain in the right side and burning during urination. An abdominal ultrasound revealed multiple calculi,

the largest of which were 11 mm and 15 mm in length in the right ureter and urinary bladder, respectively. Urethrocystoscopy was planned for the patient along with the preoperative evaluation and laboratory results. Based on his medical history, the patient has had increasing swelling in his body, particularly in his feet, from an early age; he previously underwent an operation for this reason, and has no family history of any similar disease.

Physical examination showed disproportionate growth of the lower extremities, amputated toes and superficial and large veins in the legs (Figures 1 and 2). His height was 162 cm and weight was 58 kg. Preoperative evaluation of laboratory tests and X-ray radiography of the lumbar and thoracic vertebral columns were within normal limits. Spinal anesthesia was planned for the patient. The patient was premedicated with diazepam (0.15 mg/kg) one hour before the procedure. Vascular access was established and was given isotonic saline (3-5 ml/kg) before the commencement of spinal anesthesia and monitorization of the patient (electrocardiography, pulse oximetry, non-invasive blood pressure)

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followed by skin disinfection for spinal anesthesia. Then 12.5 mg levobupivacaine was administered to intrathecal space with using 26 G atraumatic needle (B. Braun, Melsungen, Germany) at the first attempt. The duration of surgery was 90 minutes. The patient was transferred into the recovery room after the operation without any complications and then to the ward. Paracetamol 20 mg/kg was added as postoperative analgesia after 8 hours from the operation. Spinal anesthesia complications as hypotension, shivering, post-dural puncture headache and urinary retention were not seen during the intraoperative and postoperative period.



Figure 1. Huge lower extremities in Proteus syndrome. Full written permission for using the photos of the case has been obtained from the patient

Discussion

Proteus syndrome is complex hamartoneoplastic pathology with multi-system involvement (2). It was initially defined by Cohen and Hayden in 1979 (3). Although many patients exhibit clinical symptoms and deformities at birth, the disease is diagnosed at a later stage in most of cases. In our case, the patient was diagnosed at the age of 2 years by pediatricians at a university hospital, and excess lipomatous tissues were removed from the dorsolateral area (Figure 3); moreover, his toes were amputated due to overgrowth under general anesthesia at 9 years old. Skeletal system abnormalities serve as a significant characteristic of this syndrome, in which

disproportionate growth of extremities is the most frequent manifestation. Other than the enlargement of the extremities, vertebral dysplasia, kyphosis, scoliosis, and hip dislocation may be involved, among the other skeletal system disorders.

Scoliosis occurs in older cases with PS with an approximate proportion of 20% (4). In our case thoracic and lumbar X-ray radiographies were within normal limits and there were no pathologies like scoliosis, vertebral dysplasia and other skeletal system disorders.



Figure 2. Superficial and large veins in the legs. Full written permission for using the photos of the case has been obtained from the patient.



Figure 3. The patient's position while sitting. Full written permission for using the photos of the case has been obtained from the patient.

There are eight cases referred to anesthesia for several surgical procedures reported in the literature. Therefore, it is important for anesthetists to consider specific situations. Because it is

accompanied by difficult airways, skeletal system disorders, spinal canal deformities, thromboembolisms, and certain pathologies, including cystic abnormalities in the lung, emphysema, atelectasis and fibrosis, this syndrome should be further considered (5,6). Publications have particularly emphasized that difficult airways and difficult intubation may be encountered in such cases (2,7). A large epiglottis, maxillomandibular dysmorphism, and lymphangiomatosis masses may increase the airway complications. In our case Mallampati test score was class one showing that there was no risk of difficult airway. No pathologic lesion was found in his Chest X- ray. We preferred spinal anesthesia in this case because there was no serious deformity of the upper extremity and also surgical area was suitable for the spinal anesthesia.

The most significant cause of mortality in PS is pulmonary thromboembolism induced by deep vein thrombosis caused by growing extremities (5). The likelihood of developing deep vein thrombosis is less for spinal anesthesia in comparison to general anesthesia (8). In our case low molecular weight heparin was initiated after surgery for deep vein thrombosis prophylaxis. Further, spinal anesthesia has provided a more reliable and suitable choice of anesthesia due to the lower risk of development of thromboembolism in this case.

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