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Case Report

Spinal Anesthetic Management for Proteus Syndrome: A Case Report

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Abstract: Proteus syndrome is a rare, complex, and progressive disorder characterized by tissue overgrowth, deformities, and vascular anomalies. We present a case of a 56-year-old patient with Proteus syndrome who underwent successful incision and drainage an ulcerative lesion on a Cerebriform connective tissue nevus on the left plantar surface under spinal anesthesia. The patient had a history of mild lumbar scoliosis and a previous hospitalization for pulmonary embolism. Due to the patient's difficult airway and increased risk of thromboembolism, spinal anesthesia was chosen as the anesthetic technique. The procedure was carried out without complications, and the patient had a satisfactory postoperative recovery.

Keywords: Proteus syndrome, spinal anesthesia, scoliosis, pulmonary embolism, difficult airway.

INTRODUCTION

Proteus syndrome is a rare and highly variable disorder caused by sporadic somatic mutations in the AKT1 gene [1]. It leads to progressive and asymmetric tissue overgrowth, affecting multiple systems and organs. The hallmark of Proteus syndrome is the presence of various hamartomatous and vascular malformations, such as Cerebriform connective tissue nevi. These nevi, also known as cerebriform plantar hyperplasia, are often found on the soles of the feet and can cause significant functional impairment and discomfort [2]. Here, we describe the spinal anesthetic management of a patient with Proteus syndrome undergoing surgical incision and drainage an ulcerative lesion on a Cerebriform connective tissue nevus on the left plantar surface.

CASE PRESENTATION

A 56-year-old male with previously confirmed Proteus syndrome admitted to the Plastic surgery for ulcerative lesion on a Cerebriform connective tissue nevus on his left plantar surface (Fig 1.). The patient had been hospitalized two years ago for a pulmonary embolism. Physical examination showed airway Mallampati class IV, disproportionate growth of the lower extremities, and large veins in the legs. Preoperative evaluation of laboratory tests were within normal limits, and X-ray radiography of the lumbar vertebral columns showed mild scoliosis. Considering the difficulties associated with securing the airway and the thromboembolic history, the decision was made to proceed with spinal anesthesia for operation. The patient was placed in a lateral decubitus position for the spinal anesthesia procedure. After sterile preparation and draping, a spinal needle was inserted into the subarachnoid space, and 10mg of hyperbaric bupivacaine (0.5%) was injected. Adequate sensory block was achieved up to T10, providing sufficient anesthesia for the planned operation. The duration of surgery was 60 minutes. The patient was transferred into the recovery room after the operation without any complications and then to the ward. Postoperatively, the patient was given DVT prophylaxis with subcutaneous low–molecular weight heparin that was continued until he became ambulatory.

Figure 1: Ulcerative lesion of a cerebriform connective tissue nevus on the left sole of a patient with Proteus syndrome

DISCUSSION

The anesthesia management of patients with Proteus syndrome requires careful consideration of the associated challenges and comorbidities. In our case, the patient had Mallampati class IV, which could potentially limit the view of the airway and make intubation challenging. Overgrowth of soft tissues of the airway and asymmetric hyperplasia of tonsils and adenoids occur commonly in Proteus syndrome, causing airway obstruction and difficult intubation [3]. Moreover, the history of pulmonary embolism raised concerns about the use of general anesthesia and the potential risks of thromboembolism during the procedure. The most significant cause of mortality in PS is pulmonary thromboembolism induced by deep vein thrombosis caused by growing extremities [4]. The likelihood of developing deep vein thrombosis is less for spinal anesthesia in comparison to general anesthesia [5]. These factors prompted us to opt for spinal anesthesia, which offered several advantages in this scenario. Spinal anesthesia allowed for a reliable and rapid onset of anesthesia without compromising the patient's airway. The sensory block achieved was adequate for the surgical procedure, and the patient remained conscious and responsive throughout the surgery.

Avoiding the need for general anesthesia also reduced the risk of aspiration, which is particularly critical in patients with Proteus syndrome, who may have difficult airway. Spine deformities including asymmetric vertebral bodies, kyphoscoliosis, and scoliosis are frequently seen in Proteus syndrome; these add to difficulties in giving central neuraxial blocks [6, 7]. However, in our case, the scoliosis was not severe, therefore we performed spinal anesthesia without difficulty because we had sufficient experiences in providing spinal anesthesia to scoliosis patients. For orthopedic and reconstructive surgery in patients with Proteus syndrome, use of regional anesthesia with central or peripheral neuraxial block as opposed to general anesthesia has the advantage of avoiding possible problems of difficult airway and respiratory complications. Besides providing excellent analgesia and facilitating patients' early movement, regional anesthesia can help reduce the incidence of DVT. Despite the successful outcome in this case, it is crucial to acknowledge that spinal anesthesia may not be suitable for all patients with Proteus syndrome. Individual patient characteristics, the nature and duration of the surgical procedure, and the surgeon's preference must all be taken into account when selecting the appropriate anesthetic technique.

CONCLUSION

In this case report, we described the successful incision and drainage of a Cerebriform connective tissue nevus on the left plantar surface in a patient with Proteus syndrome under spinal anesthesia. The decision to use spinal anesthesia was justified by the patient's difficult airway and history of thromboembolism, and the procedure was completed without complications. Spinal anesthesia proved to be a safe and effective alternative to general anesthesia in this challenging patient population

Conflict of interest

No conflict of interest was declared by the authors.

Financial disclosure

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Consent for publication

Written informed consent was obtained from the patient for publication of this case report and the accompanying image.

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