



## Case Report

# Spinal anesthesia in a patient with Darier's disease: A case report

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## ABSTRACT

Darier's Disease (Darier-White Disease, keratosis follicularis) is a rare genodermatosis characterized by keratinisation defects that affects the skin, nails and mucosal membranes; it is autosomal and dominantly inherited. To the best of our knowledge, only one case has been reported regarding spinal anesthesia management in a patient suffering from Darier's disease—this was during Cesarean section. Here, we describe a case of spinal anesthesia with standard skin preparation for inguinal hernia surgery in a Darier's patient. Spinal anesthesia is a safe anesthesia choice when administered in lesion-free areas.

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## Introduction

Darier's disease (Darier-White Disease, keratosis follicularis) is a rare genodermatosis characterised by keratinisation defects, which affect the skin, nails and mucosal membranes; it is autosomal and dominantly inherited [1,2]. The worldwide prevalence is thought to be between 1/30,000 and 1/100,000 [1]. It is rarely symptomatic at birth, and usually presents in the first or second decade of life [3]. Darier's Disease's skin lesions and their potential for secondary infections may be problematic during spinal anesthesia. To the best of our knowledge, there is one case report in the literature describing spinal anesthesia in a patient with Darier's Disease. In that case, skin preparation before spinal anesthesia was performed using cetrimide, chlorhexidine and isopropyl alcohol [4]. Here, we report a case of spinal anesthesia with standard skin preparation in a Darier's Disease.

## Case Report

A 49-year-old-male patient (weight 65 kg, height 170 cm) with a right-sided inguinal hernia was consulted in our preoperative preparation clinic. The patient had a medical history of postoperative atelectasis after general anesthesia for tonsillectomy 12 years prior. His father and sister also had Darier's Disease. He had a history of

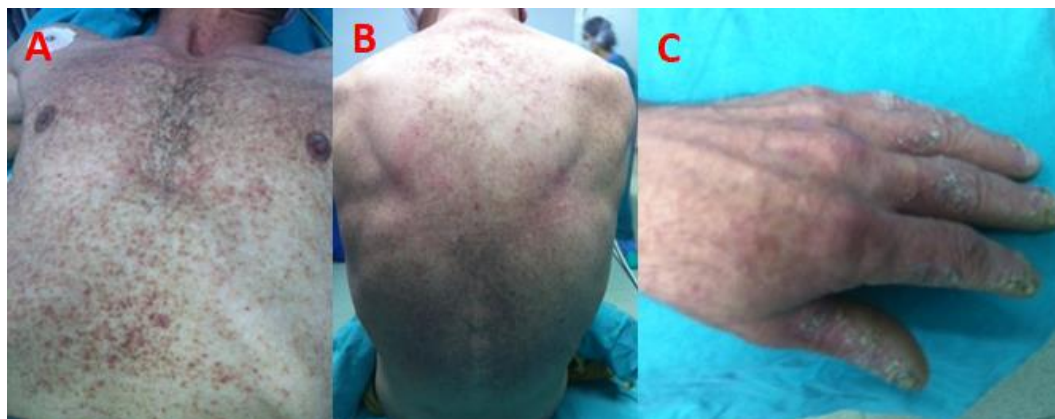
smoking 60 pack/years. On physical examination, there were brown fatty hyperkeratotic lesions over most of his body including his nail beds. There were no signs of skin infection in the spinal area. There were no lesions on his oral mucosa. His other systemic findings including laboratory results and electrocardiogram were normal. He had a mild restrictive pattern on a respiratory function test. He was classified as American Society of Anesthesiology (ASA) Physical status 2 and Mallampati 2. The patient's informed consent was taken after spinal and general anesthesia was explained. Due to his previous experience with postoperative atelectasis, it has been decided to perform the surgery under spinal anesthesia. No evidence of secondary infection was found on swabs taken from lesions on his back. The swab sample had already been taken by dermatologists during the preoperative preparation period (48 hours ago). Although there is no consensus, it can be said that it should be taken within a few days.

The patient's electrocardiogram, blood pressure and O<sub>2</sub> saturation were monitored. An 18-g cannula was used for IV access on the left dorsum of the hand and an isotonic saline solution was administered at 10 ml/kg. Hemodynamic parameters were normal and the SpO<sub>2</sub> was 96-98% throughout the procedure. The patient was placed in the sitting position. There were papular lesions on the vertebral and paravertebral region, but none at the L3-L4 level. The skin was prepared with chlorhexi-

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dine and isopropyl alcohol. A 25-g spinal needle was used, and 12 mg of bupivacaine was administered to the

subarachnoid space. After spinal anesthesia, the patient's head and shoulders were kept at a 30-degree angle.



**Fig. 1.** Presentation of Darier's disease in our patient: (a) Lesions of thoracoabdominal region, anterior aspect; (b) Lesions of thoracodorsal region, posterior aspect; (c) Patients left hand and lesions.

We used a pin prick to measure sensory and motor blockage. Sensory block was achieved at 10th minute to dermatome T10. The patient's surgery lasted 60 minutes. His hemodynamics was normal, and SpO<sub>2</sub> was over 96% throughout the procedure. There were no postoperative complications due to spinal anesthesia. The patient was externalised at postoperative 30th hour and at postoperative 5th day, there was no evidence of complications including skin infection. There were no neurologic or neuraxial anesthesia-related complaints.

#### 4. Discussion

Darier's disease is a rare dermatological disease caused by the mutation of ATP2A2 gene located in area 12q23-q24 on the first chromosome [1]. It is commonly seen in the first or second decade of life. Our patient's first symptoms appeared at 17 years of age. Darier's disease is characterised by converging papular lesions in the seborrheic areas of the face, torso and back with nail bed involvement seen in all patients [4]. Our patient demonstrated widespread lesions. (Fig. 1) Secondary bacterial, viral and fungal infections can be seen on these lesions, leading to malodorous lesions of the scalp and genital area [1]. Our patient had a history of malodorous lesions during the summer months but none were present at this time. Patients with Darier's disease may also show psycho-neurological pathologies [5]. This was not seen in our patient.

Darier's disease is known to flare up with heat, perspiration, humidity, sunlight, corticosteroid use and mechanical trauma [5]. We believe it is therefore important for patients to protect themselves from sunlight and to use oral or IV corticosteroids preoperatively. During the procedure, anesthesiologists and surgeons should avoid the lesions.

Sharma et al. [4] reported the use of spinal anesthesia for Cesarean section in a 26-year-old woman with Darier's disease who presented with cephalopelvic discordance. The authors reported that cetrимide, chlorhexidine and isopropyl alcohol were safe to use for skin

preparation before spinal anesthesia in areas with no lesions or secondary infection.

We selected chlorhexidine and isopropyl alcohol to prepare the site of spinal anesthesia. There were no lesions or secondary infections in this area. There were no postoperative complications related to spinal anesthesia. Similar to Sharma et al. [4], we believe that spinal anesthesia in areas without a lesion is safe.

Sharma et al. [4] used cetrимide, chlorhexidine and isopropyl alcohol possibly with the aim of minimizing infectious complications caused by neuraxial blockade. We did not find any literature describing the use of cetrимide for preoperative skin preparation in spinal anesthesia. Many studies have demonstrated the benefits of chlorhexidine and isopropyl alcohol for skin preparation before neuraxial blockade [6,7]. In light of these studies, we used chlorhexidine and isopropyl alcohol for skin antisepsis. We do not believe that there is a need for cetrимide.

In conclusion, Darier's patients can safely undergo spinal anesthesia with standard skin preparation techniques when administered through lesion-free areas with no evidence of secondary infection.

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#### Conflict of Interest

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