Case report

Anesthetic Considerations of Palmoplantar Keratoderma (PPK), a Case Report

Parisa Sezari[1], Masoud Nashibi[1], Farhad Safari[1], Ahmad Naderi[2], Kamran Mottaghi[1*]

Abstract

Palmoplantar keratoderma (PPK) refers to a group of heterogeneous disorders defined by thickening of skin on the palms and soles. Due to diversity of symptoms and involved organs, the term PPK does not describe each individual patient precisely. The syndrome is quite rare and unknown to the anesthesiologists, while serious associations such as difficult airway or cardiac disease are significant challenges during perioperative management. The literature lacks enough data on anesthetic considerations in these patients, implicating case reports as the only available sources. We revised the accessible information through description of our patient.

Keywords: Palmoplantar keratoderma, Anesthetic consideration, Keratosis

1. Anesthesiology Research Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran
2. Department of Anesthesiology, Jam Hospital, Tehran, Iran

Corresponding Author:
Kamran Mottaghi, MD. Associate Professor, Department of Anesthesiology, Anesthesiology Research Center, Shahid Beheshti University of Medical Sciences; Tehran, Iran.
Email: k_mottaghi@sbmu.ac.ir
Tel/Fax: 0098-21-55419005

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Introduction

Palmoplantar keratoderma is an extremely rare disease of acquired or genetic origin. The genetic source could be hereditary (autosomal recessive or dominant) or sporadic as a mutation. Its scarcity leads to shortage of resources concerning anesthesia management. Here we report spinal anesthesia in a known case which according to our knowledge is unique in literature.

Case Report

A 38 year-old female came to our hospital with fever, chills and respiratory distress. Five days earlier, she had her pregnancy terminated due to intra uterine fetal death at her 32nd week of pregnancy. She was suffering vaginal bleeding, diarrhea, vomiting and had productive cough. She was febrile (T_{oral} of 40°C), tachychrdic (PR 105/min) and her SpO2 in room air was 90% and was candidate for emergent cervical dilation and endometrial curettage. The PalmoPlantar Keratoderma (PPK) diagnosis established when she was 2 years old and was under the treatment with daily Acitretin (25 mg). She had undergone below knee amputation of left leg secondary to untreatable infection at the age of seventeen.

On physical examination, hyperkeratosis of hands and foot, patchy circumoral defects and fizzy hair were obvious (Figures 1-4). Lab data were in normal range except for hemoglobin (7.9 g/dL) and platelet count (90000/µL). On echocardiography all the findings were normal. The Olmsted subtype of the disease consists of mutilation of digits and periorificial keratotic plaques which is similar to our case and is

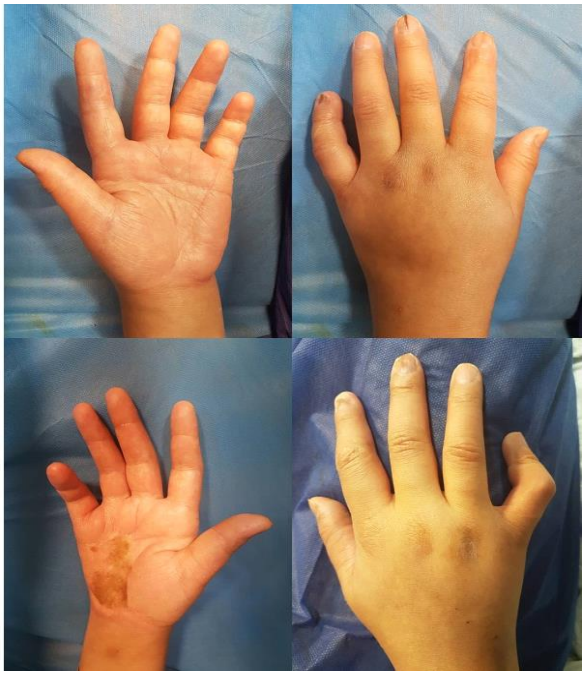

Figure 1. Hyperkeratosis of hands and nail fissuring and grooving.

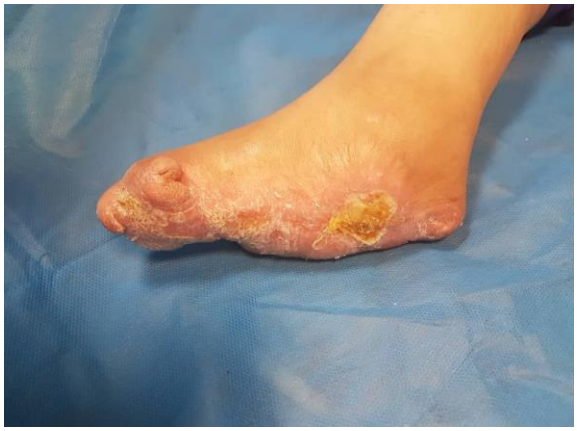

Figure 2. Toes lost and hyperkeratosis of right foot.

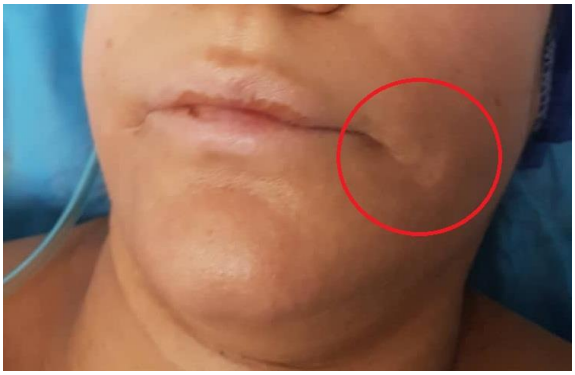

Figure 3. Patchy circumoral defects.

extremely rare (1).

Since the surgery was going to last about 5 to 10 minutes and patient was not fasting, we decided to apply spinal anesthesia. In sitting position under sterile conditions using 27G Quincke type needle in midline approach and on first attempt, 10 mg of hyperbaric 0.5% Bupivacaine was injected in subarachnoid space. Anesthesia and surgery was uneventful and after termination of procedures, she was transferred to post anesthesia care unit and then to gynecology ward.

Written informed consent was taken from the patient regarding the publication of data and pictures in scientific journals.

Discussion

Palmoplantar keratoderma is a hyperkeratosis of hands and feet. It could be accompanied by other findings which may form different syndromes and diseases such as cardiac involvement (Naxos disease) (2). The disorder may involve teeth and jaws which could lead to facial deformities and likelihood of difficult airway (3). It could be acquired or hereditary, although, the sporadic mutations might be the origin as well and inherit to offspring. Since its scarcity, the emphasis in literature is mostly on diagnosis and treatment or surgical procedure (1, 2) and anesthetic considerations are off the spotlight. The main source of evidence for this concern is derived from case reports as the trials are absent.

To our knowledge, there are only three case reports discussing anesthesia management of PPK patients. Iranpour (3) reported nail grooving and fissuring may interfere in correct pulse oximetry; moreover, they suggested teeth suturing to prevent

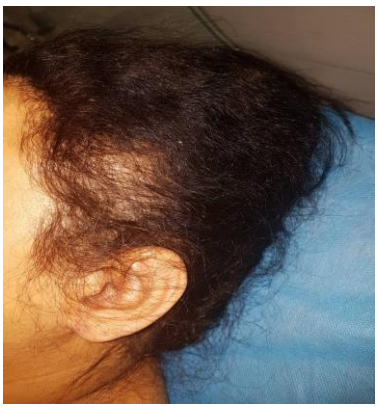

Figure 4. Fizzy and dry texture of hair.

loose teeth to fall in airway or alimentary tract. They emphasized on possible difficult airway secondary to facial deformities. Zamiri (4) used peripheral nerve blocks to provide anesthesia for biopsy and butox injection. Yildiz (5) reviewed Naxos disease, a combination of PPK and cardiac involvement. They used sedation with Ketamine for a kid during central venous catheterization.

Conclusion

Although rare, PPK could be a challenge to anesthesiologists due to potential cardiac involvement and/or difficult airway. When applicable, regional anesthesia could be an acceptable choice by avoiding airway manipulation and minimal change in hemodynamic indices.

Acknowledgment

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Conflicts of Interest

The authors declare that there are no conflicts of interest.

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