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Case Report
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Anaesthetic Management of a Known Case of Werner Syndrome by Peripheral Nerve Block in the Orthopaedic Surgery of Forearm

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Abstract

Werner syndrome (WS) is a rare hereditary disease, characterised by the clinical signs and symptoms of premature ageing. Patients with WS usually have difficult airway due to anatomic malformation of the oral cavity. General anaesthesia with endotracheal intubation poses a high risk for these patients. On the other hand, the risk associated with the peripheral nerve block is minimal. Here we report the successful management of a known case of WS by using a peripheral nerve block (axillary brachial plexus block) without any significant complications. The patient was a 39-year-old man, a known case of WS, admitted to the hospital with chief complaint of non-healing ulcers on his wrist and elbow due to the compression effect of the abnormal ulna bone on the overlying soft tissue. To the best of our knowledge, this is the first case report of using peripheral nerve block in the anaesthesia of a patient with WS.

Keywords: Anaesthesia, nerve block, Werner syndrome

Introduction

Werner syndrome (WS) is a rare hereditary disease, characterised by the clinical signs and symptoms of the premature ageing. The patients develop diabetes mellitus type II, atherosclerotic changes of blood vessels, ischaemic heart disease and hypertension early in life. Non-healing ulcers and early cataract changes of the lens other co-existing disorders of this syndrome.

Patients with WS usually have a difficult airway due to anatomic malformation of the oral cavity. General anaesthesia with endotracheal intubation poses a high risk for these patients due to the above manifestations. On the other hand, the risk associated with the peripheral nerve block is minimal.

We report the successful management of a known case of WS by using peripheral nerve block without any significant complications. To the best of our knowledge, this is the first case report of using a peripheral nerve block in the anaesthesia of patients with WS.

Case Presentation

The patient was a 39–year-old man, a known case of WS, admitted to the hospital with a chief complaint of non-healing ulcers on his wrist and elbow due to the compression effect of the abnormal ulna bone on the overlying soft tissue (Figures 1, 2).

The orthopaedic team had planned for osteotomy of distal part of the ulna bone. An anaesthesiologist was consulted the day before the surgery to find the best plan for anaesthesia, considering the patient's comorbidities and problems.

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On physical examination, the patient was cachectic with senile appearance, short stature, poor dental condition, grey hair, alopecia, scleroderma-like skin changes and skin ulcers. The thyro-mental distance was 3 cm, and the mouth opening was limited (Figure 1).

The patient had a history of myocardial infarction 5 years before (taking Acetyl-Salicylic-Acid (ASA) and captopril), and the cardiologic evaluation showed moderate aortic stenosis, moderate mitral regurgitation, an ejection fraction of approximately 20%–25% with global hypokinesia and changes of old inferior myocardial infarction on the electrocardiogram, and he was determined to be at a high risk for general anaesthesia.

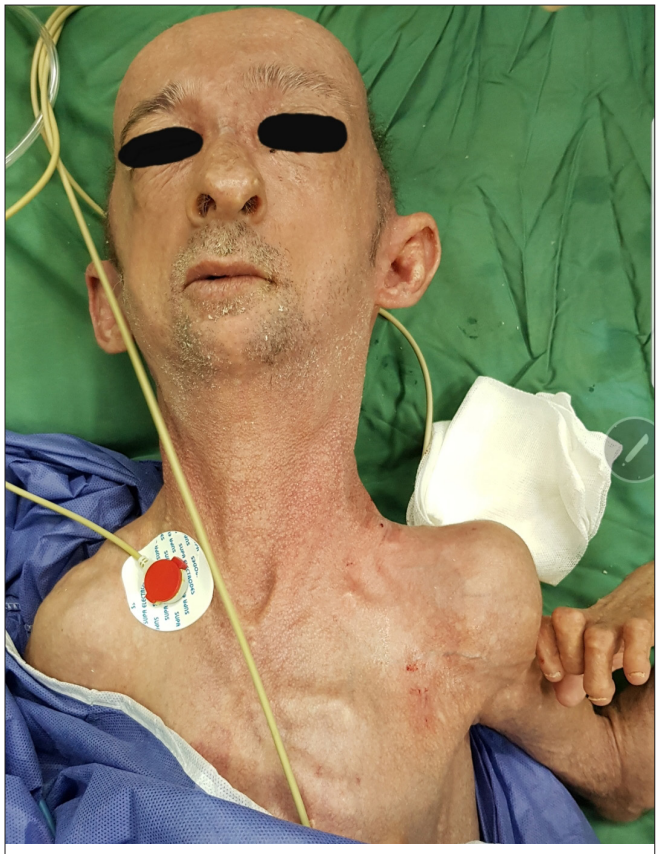

Figure 1. The physical appearance of the patient with Werner syndrome

Main Points:

- Airway management is highly difficult in Werner syndrome during surgery.
- Regional anaesthesia can successfully reduce complications compared to general anesthesia.
- Peripheral nerve block helped to prevent intra-operative hypoxic periods in a patient with Werner syndrome underwent osteotomy for distal part of the ulna bone.
- Regional anesthesia contributed to prevent myocardial ischemia and stroke in the case of Werner syndrome.
- Regional anesthesia caused more hemodynamic stability than general anesthesia in the case of Werner syndrome.

He also had seizure disorder treated by Depakin. The internal medicine specialist had predicted a high-risk surgery with general anaesthesia due to uncontrolled diabetes mellitus. Also, the patient had respiratory distress due to moderate obstructive airway disease. A pulmonologist was consulted, and he ordered the use of bronchodilators before anaesthesia. The pulmonologist had also recommended avoiding general anaesthesia if possible in this patient.

Considering all the risks mentioned above and the difficulties we encountered during intubation resulting from patient's facial and upper airway anatomy, we found the peripheral nerve block as the best choice for our patient.

The plan of surgery and anaesthesia was explained to the relatives, and written informed high-risk consent was obtained.

In the operating room, all preparations including emergency drugs and difficult airway instruments, such as those for fiber-optic laryngoscopy, were kept as stand by.

We positioned the patient supine and abducted the arm to 90 degrees. After the skin and transducer preparation, we placed a linear 38 mm, high-frequency 10–12 MHz transducer in the transverse plane along the axillary crease. After the selection of an appropriate depth of field (within 1–2 cm) and focus range (within 1 cm) and gain, we identified the axillary artery by using a colour Doppler. Afterwards, we visualised the median, ulnar, radial and musculocutaneous nerves in the transverse short axis view around the axillary artery.

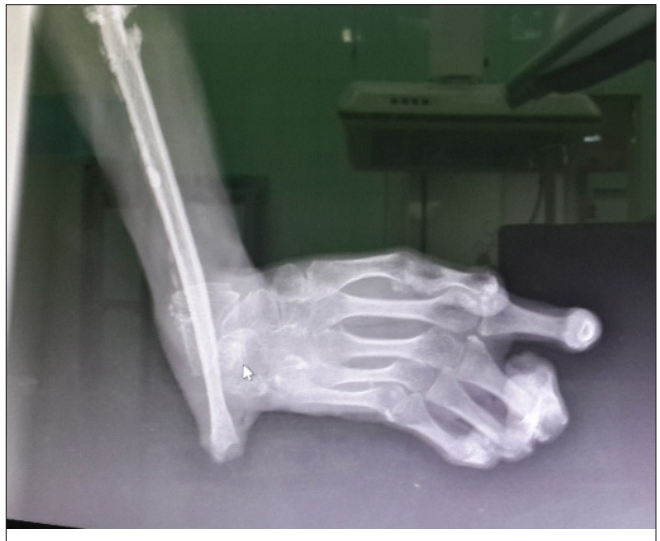

Figure 2. The left forearm X-Ray of the patient who was the candidate for osteotomy of the distal part of the ulna under peripheral nerve block

Then, we inserted a 5 cm 23 G needle parallel to the long axis of the transducer, in line with the ultrasound beam. We injected 4 mL of bupivacaine 0.5% at each nerve location and ensured that the local anaesthetic spread around each individual nerve.

After 20 minutes, the surgery was initiated. The ulnar osteotomy was successfully performed by an orthopaedic surgeon. The heart rate (HR), blood pressure (BP), blood oxygen saturation (SpO_2) and electrocardiogram monitoring were performed throughout the surgery. For control of anxiety and sedation during operation, a low dose of midazolam 0.05 mg kg^{-1} intravenous bolus was administered as needed. There were no significant changes in the HR and BP values intraoperatively. The SpO_2 was maintained between 92% and 94%. After 90 minutes, the patient was transferred to the post-anaesthetic care unit (PACU). He was awake and stable hemodynamically in the PACU and had no significant problems or adverse effects. Twenty-four hours after admission to hospital, the patient was discharged successfully.

Discussion

Werner syndrome or adult progeria is a rare autosomal recessive premature ageing disorder beginning after puberty. It has an overall incidence of 1:1,000,000–1: 10,000,000. WS is related to mutations of the WRN gene, which participates in DNA repair and replication, telomere maintenance and apoptosis, thus causing a multi-organ involvement. Clinical findings include senile appearance, voice abnormalities, a short stature, hypogonadism, alopecia, grey hair, bird-like face, scleroderma-like skin changes, skin ulcers, cataracts, osteoporosis, type II diabetes mellitus, ischaemic heart disease and difficult airway. This syndrome becomes apparent in adolescence but is usually undiagnosed until the third or fourth decade of life. The patients usually die at the age of 40–50 years due to malignancy or atherosclerotic complications (1, 2).

The major concerns for the anaesthesiologist to anesthetise patients with WS are difficult airway, securing an intravenous cannula, acceleration of hypertension, myocardial ischaemia and stroke. Difficult airways are attributed to the craniofacial anomalies such as micrognathia, a large head, short stiff neck, poor dentition and mandibular and maxillary hypoplasia.

The complications associated with difficult airway included brain and cardiac injury due to hypoxia, aspiration of gastric content, dental, pharyngeal and laryngeal injury, laryngospasm, bronchospasm and death (3). To prevent these adverse effect, it was necessary to use alternative anaesthesia techniques, such as regional anaesthesia. As in our case, there were no such complications with peripheral nerve block.

In patients with previous myocardial infarction (similar to our case), general anaesthesia with laryngoscopy and tracheal intubation leads to dangerous complications such as hypertension, hypotension, hypoxia, arrhythmia, cerebrovascular accident, new ischaemic electrocardiogram changes, tachycardia, bradycardia and death (4). We had none of these complications during and after the axillary brachial plexus block. It was due to avoidance of laryngoscopy stress.

Our patient had a history of asthma and bronchodilator application. Choosing general anaesthesia with endotracheal intubation in this patient might lead to severe bronchospasm, hypoxia and extubation failure due to respiratory failure after bronchospasm, need for the mechanical ventilation (MV) support in the intensive care unit, complications associated with MV and death (5). Using the peripheral nerve block in our patient prevented the occurrence of such hazardous adverse effects because laryngoscopy and tracheal intubation that could exacerbated bronchoconstriction was avoided.

Conclusion

Our experience reveals that using a peripheral nerve block, such as axillary brachial plexus block, under ultrasound guidance prevents many dangerous complications of general anaesthesia, such as cardiac arrest and stroke in patients with WS.

Informed Consent: Written informed consent was obtained from the patient's parents who participated in this case.

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Conflict of Interest: The authors have no conflicts of interest to declare.

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