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Case Report

Before Old Age-A Rare Case of Werner Syndrome

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Abstract

Werner syndrome or adult progeria is the most common of the premature ageing disorders. Patients usually present with all the symptoms and signs of old age very early in life, the most common being cataract, development of type II diabetes mellitus, osteoporosis, atherosclerotic changes in the blood vessels, non healing ulcers etc. Secondary complications may necessitate various surgical procedures in these patients subjecting the patient to high risks under anesthesia.

The anesthetic implications in these patients are very challenging as the airway is more often difficult owing to the morphological changes of the oral anatomy. Coexisting diseases like hypertension and ischaemic heart disease may also pose a greater threat to anaesthetize these patients.

A complete knowledge of the anesthetic challenges and preparedness for the management of complications is essential in patients with Werner syndrome coming for elective or emergency surgeries.

Keywords: Progeria; Werner syndrome; Anaesthesia; Debridement

Case Report

A 32 yr old male patient with a diagnosis of Werner syndrome is posted for debridement of non healing ulcer in the left lower limb. He was diagnosed with Werner syndrome 10 yrs back when he had developed cataract bilaterally and surgery under local anaesthesia was done for the same and vision was restored. Six years back he was diagnosed with type II diabetes mellitus and he has been on oral hypoglycemic agents since then. No history of chest pain or breathlessness. The patient has been having multiple ulcers on the lower limbs and frequently visits the dermatology clinic for regular dressings. These ulcers have been painful and so the patient was confined to the bed. Patient gives history of loss of molar teeth on either side. History obtained from the parents revealed that the patient's sister had the same disease and died of ischaemic heart disease at the age of 25 yrs.

On examination, patient appeared to be 60 yrs of age, cooperative but unable to articulate his speech correctly. The head was large, there was loss of hair extensively over the scalp with premature greying, absent outer eyebrows and pale conjunctiva. The skin appeared to be non-elastic with decreased fat in the subcutaneous tissue and thin extremities. The chest was deformed with prominent lower ribs. Chest was clear on auscultation and cardiac sounds were well heard. Multiple small ulcers were seen on the upper and lower extremities. A large ulcer measuring 7x8 cms was seen in the left ankle which needed debridement under anaesthesia.

Airway examination revealed mouth opening of 1.5 cms with Mallampati grade IV. Neck extension was slightly restricted. Thyromental distance was normal. Mild scoliosis was present on examination of the spine. Investigations showed hemoglobin -9.6 gms%, platelet-3.6 lakhs/dl, INR-1.1, RBS-217gms/dl, FBS-126gms/dl, HbA1C-7.0, renal function tests, serum electrolytes were within normal limits. Left lower limb arterial doppler study revealed generalized atherosclerotic changes of lower limbs with biphasic spectral waveform. Electrocardiography and echocardiography were normal.

The risk associated with this syndrome and anaesthesia was explained to the relatives, but the need for surgery was also emphasized and written informed high risk consent was obtained. The case was accepted under American Society of Anesthesiologists-physical status III classification. Patient was advised to be fasting for 6 hrs.

In the operating room, all the emergency drugs and the difficult intubation cart including fibreoptic bronchoscope were kept as stand by on the day of surgery. Monitors such as electrocardiography, non invasive blood pressure and saturation probe were connected and baseline parameters were recorded. Intravenous infusion of Ringer lactate solution was started through the patent intravenous cannula. In the left lateral position, under aseptic precautions, after subcutaneous infiltration with local anesthetic Xylocaine 1%, 2 ml of 0.5% heavy Bupivacaine was injected into the subarachnoid space following free flow of cerebrospinal fluid. Analgesia was adequate with level of analgesic block being T10 dermatome. Surgery was done in the supine position. Hemodynamic were stable throughout the surgery which lasted for 20 mins. Patient was shifted to the post anaesthesia care unit for monitoring and observation for 6 hrs. He was discharged from the hospital on third postoperative day.

Discussion

Otto Werner originally described Werner syndrome in 1904 on the basis of scleroderma like tight skin and bilateral cataract. It is an autosomal recessive disorder affecting 1 in 4-8 million population and there are about 1000 cases reported with maximum incidence in Japan and Sardinia. It is also called Progeria of adulthood where the manifestations begin after the first decade of life when the affected individual fails to undergo the usual growth spurt and there appears

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the evidence of premature ageing with pathological manifestations. The average life span is around 30-35 yrs. In children it is called Hutchinson Gilford Syndrome when the average life span is only upto 13.6 yrs with manifestations as early as 3 yrs [1]. Progeria in the Greek language means "before old age" [2].

The complication of ageing in a normal population is accelerated in Werner syndrome to as early as 20-30 yrs of age. They include the development of type II diabetes mellitus, early cataracts, extensive atherosclerosis of the arteries which results in hypertension, stroke, ischaemic heart disease and peripheral vascular disease [3]. In females, premature menopause is an additional sign. The facial sign is typically called the "bird like" face [4].

Major concern for the anesthesiologist to anaesthetize these patients are difficult airway attributed to multiple craniofacial abnormalities like micrognathia, mandibular and maxillary hypoplasia, large head, short stiff neck and poor dentition [5]. Securing an intravenous canula is also quite challenging owing to the non elastic nature of the skin. Intraoperatively, the risk of accelerated hypertension, myocardial ischaemia, and stroke is higher [6].

When propofol is used for induction of anaesthesia in patients with Werner syndrome, the incidence of "Propofol infusion syndrome" resulting in lipemia has been reported [7].

Conclusion

Authors conclude that all these anesthetic implications and the increased frequency of surgical complications in patients with Werner syndrome poses a greater challenge to the practicing anesthesiologist.

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