Anaesthesia management for Winchester syndrome

Sir,

We report a case of Winchester syndrome for femur fracture fixation, which was successfully managed in our hospital. It was interesting because no prior medical literature exists on its anaesthesia management, to date.

A 15-year-old male, a known case of Winchester syndrome with distal left femur fracture was scheduled for intramedullary nailing. He had a history of seizure disorder since birth for which he was on oral medication. His older sibling also had the same genetic disorder. Apart from this, past/personal/birth/family history were all normal. On physical examination, he was a short, stubby male, afebrile, with a height of 4ft.11 inches, weight of 50 kg, a scoliotic spine, dark, leathery, dull skin, coarse facial features, heart rate of 120/min and a blood pressure of 110/60 mmHg [Figure 1]. His pulmonary, cardiac, neurological and abdominal examination were unremarkable. His airway examination revealed a Mallampati II grading, with stiffness of temporomandibular (TM) joints, inter-incisor distance

Table 1 : Lesions and concerns for the Anaesthesiologist

Lesion	Problem	Significance	Action Taken
Leathery Skin	Difficulty in venous access	Since this was a surgery with increased blood loss, proper iv access was essential	Wide bore iv access taken as urgent access intra-op would be difficult.
Scoliosis	Reduced lung function,	PFT and CXR was done to ascertain status.	Both were within normal limits.
	Cardiac involvement,	Assessed by ECHO	No cardiac abnormality
	Difficulty in giving spinal injection	Proper positioning important	Spinal given by senior anaesthesiologist
Seizures		intraoperative drug interactions	Sevoflurane avoided
Joint involvements	Difficult airway	Atlanto-axial mobility and jaw mobility abnormal	Difficult airway cart kept in standby

PFT-Pulmonary function tests, CXR-Chest radiogram, ECHO-Echocardiogram

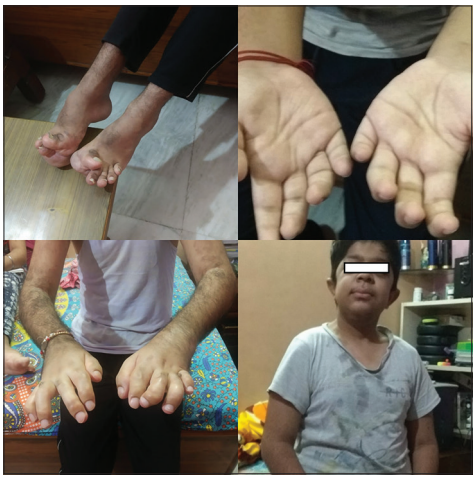

Figure 1: General appearance of the patient

4 cm and thyromental distance 6 cms. Laboratory investigations revealed a haemoglobin (Hb) level of 14.5g/dL, total leucocyte count(TLC) of 12.8 x 109/L, platelet count of 3.2 lakh/ μL ,prothrombin time of 14.7 seconds, activated partial thromboplastin time of 30 seconds and international normalised ratio of 1.3. All other blood and urine investigations were within normal limits.

In the operation suite, there was difficulty in securing venous access. External jugular vein (R) was cannulated with 18 G cannula and median antecubital vein (L) was cannulated with 18 G venflow. Patient was administered ringer lactate solution. Haemodynamic parameters were within normal limits. Patient was positioned for spinal anaesthesia. 25 G Quincke's needle was used for lumbar puncture, after giving local anaesthetic in L4-L5 space. 0.5% bupivacaine (H) 2.5 ml + 25 μg fentanyl was given. Patient was made to lie down unilaterally immediately and after the block was achieved, he was adequately positioned, making sure all joints and pressure points were adequately padded. Intraoperative course was uneventful.

Patient was given 1 mg midazolam intraoperatively for anxiolysis. Intraoperatively, he was administered 2 litres of Ringers Lactate and the procedure lasted for 2.5 hrs. Postoperatively he was shifted to the recovery bay. He was shifted to the ward only after 6 hrs and complete movement was regained after the motor block. He was administered Inj tramadol 100 mg for post-operative analgesia.

The first postoperative day was uneventful. Multimodal analgesia with Inj tramadol 100 mg SOS and Inj paracetamol 1 g thrice a day was administered. He was discharged on 4th postoperative day.

In 1969, Winchester and associates[1] described a new syndrome characterised by short stature, joint stiffening with severe contractures, peripheral corneal opacities, coarsened facial features, dissolution of the carpal and tarsal bones, and generalised osteoporosis. The presence of marked intra- and periarticular joint destruction simulated advanced rheumatoid arthritis.

The clinical and roentgenographic features of the present case are similar to those described by Winchester and associates[1] and Brown and Kuwabara[2]: onset in infancy of arthralgias and joint stiffening, dwarfism, peripheral corneal opacities, progressive symmetric flexion contractures of major and minor joints, skin lesions (more conspicuous in our patient), absence of mental retardation, generalised osteoporosis, carpal-tarsal osteolysis, and marked intra- and periarticular small joint destruction. The patient also had history of seizures, for which he was on medication.

Angiographic studies on the patient demonstrated hypervascularity of joints whose adjacent bone appeared to be undergoing resorption. The most characteristic feature was the thickened, leathery feeling imparted to the skin, although hyperpigmentation and hypertrichosis were also evident, as described in literature.[3]

But having to manage a case with no prior reports on the conduct of anaesthesia was challenging and exciting. From existing literature and our experience, areas of concern for the anaesthesiologist for this case are highlighted in Table 1.

Despite having a senior anaesthesiologist attempting to give the spinal injection, the dura was punctured only in the 4th attempt. However post-injection, the patient remained stable and had no complications intraoperatively and postoperatively and was discharged on 4th postoperative day.

In clinical practice, we do not always get a clinical case with literature published on it. So in the absence of the same, practice of 'safe anaesthesia'[4,5] and the oath 'primum non nocere' should guide management. This helped us to manage our case safely.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Submitted: 21-Jul-2020
Revised: 25-Aug-2020
Accepted: 27-Sep-2020
Published: 10-Feb-2021

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Access this article online	
Quick response code	Website: www.ijaweb.org
	DOI: 10.4103/ija.IJA_984_20

How to cite this article: Sharma A, Sharma P, Sasidharan S. Anaesthesia management for Winchester syndrome. Indian J Anaesth 2021;65:165-7.

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