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Case report

Blue rubber bleb nevus syndrome – Anaesthetic experience of prolonged bowel surgery in a twelve year old child and review

Ankur Sharma, Ganga Prasad *, Rahul Anand

Department of Anaesthesiology, All India Institute of Medical Sciences, New Delhi, India

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Abstract Blue rubber bleb nevus syndrome (BRBNS) is a rare hereditary disease manifested by multiple vascular malformations involving skin, mucosa and musculoskeletal system. Available literature consists of mainly airway management of such patients. We describe the perioperative anaesthetic management of a child of BBRNS with gastrointestinal bleeding disorder scheduled for resection of multiple nevi and discuss the relevant anaesthetic concerns.

1. Introduction

Blue rubber bleb nevus syndrome (BRBNS) is a rare condition characterised by multiple vascular malformations in various organs of the body, most commonly the skin and gastrointestinal tract [1]. It was first reported in 1860 by Gascoyen [2]. Later on, nearly a century after the first report, it was described by Bean [3] and named as ''Blue Rubber Bleb Nevus Syndrome'' or ''Bean Syndrome'' because of colour, texture and shape of its characteristic lesions.

These patients can present with multiple surgical, medical and cosmetic complaints. Most of them will undergo surgery and anaesthesia at least once in their lifetime. We are discussing a case of BRBNS in a child who underwent prolonged bowel surgery for multiple bleeding gastrointestinal lesions and the anaesthetic concerns.

2. Case report

A 12-year-old, 25-kg girl with blue rubber bleb nevus syndrome scheduled for exploratory laparotomy and resection of multiple vascular gastrointestinal tract (GI) lesions. She had small moles all over the body since birth, which were increasing in size (nevi were present on the middle of tongue, right side of the scalp, behind left ear, upper lip, both the arms, left forearm, back, left thigh and foot- Fig. 1). She had multiple episodes of malena leading to severe anaemia for which she received multiple blood transfusions. Endoscopy revealed multiple nevi in the second part of the duodenum and computer tomography (CT scan) showed hypodense lesion in erector spinae muscles at the level of L2–3 and no other involvement. She received sclerotherapy (sodium tetra decyl sulphate) for the lesions present on face, trunk and limbs. The child had also

* Corresponding author at: Department of Anaesthesia and Intensive Care, All India Institute of Medical Sciences, Room No-5011, 5'th Floor, Main AIIMS Building, Ansari Nagar, AIIMS, New Delhi 110049, India. Tel.: +91 9968501929.
E-mail address: docmed4@gmail.com (G. Prasad).

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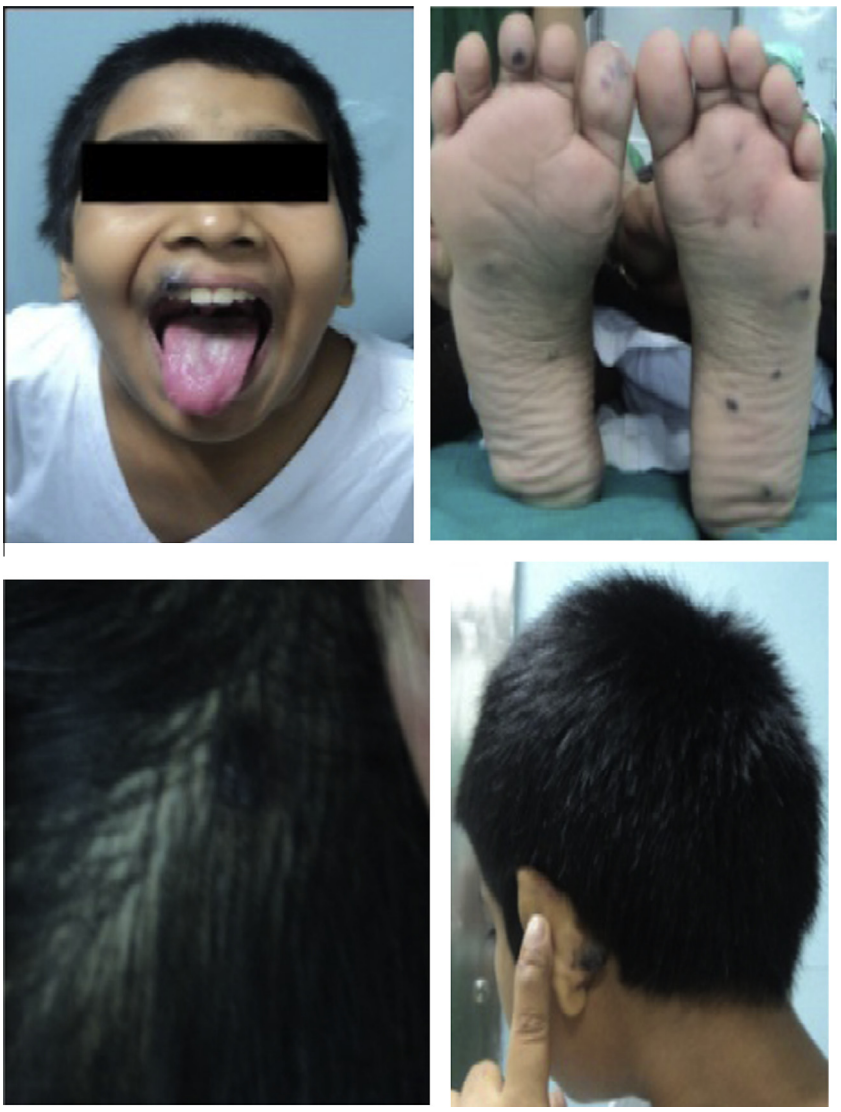

Figure 1 Bluish rubbery nevi present on middle of tongue, right side of upper lip, both soles, scalp and behind the left ear.

been diagnosed with neurocysticercosis (frontal area) one year back following an episode of seizure and was currently on tablet Carbamazepine 200 mg bd. She was anaemic and her haemoglobin was 8.5 gm/dl, all other routine haematological investigations and Chest X-ray were within normal limits. Airway was assessed with indirect laryngoscopy and did not reveal any lesions in the uvula, posteriorpharyngeal wall and epiglottis.

Her anaesthetic management includes: intravenous premedication with midazolam 1 mg was given five minutes before induction and the baseline ECG, pulse oximeter and non-invasive blood pressure (NIBP) were recorded. Child was preoxygenated with 100% oxygen and anaesthesia was induced with intravenous fentanyl 50 μg and propofol 60 mg, after confirming adequacy of mask ventilation, atracurium 12.5 mg was given for muscle relaxation. After gentle laryngoscopy, trachea was intubated with a 6.0-mm internal diameter cuffed orotracheal tube, and anaesthesia was maintained with 40% oxygen in air and 0.8–1.2% end-tidal isoflurane. A radial arterial line was placed for invasive blood pressure monitoring. Patient was ventilated to keep end tidal CO_2 ($EtCO_2$) in the range of 35–45 mm Hg.

Surgery lasted 9 h, and a total 67 lesions were excised. Blood loss was 250 ml and 150 ml of PRBC was given intraoperatively. Overall intraoperative course was uneventful. In the end neuromuscular block was reversed with injection neostigmine 1.25 mg and glycopyrrolate 0.25 mg and trachea was extubated when the child was conscious. For postoperative analgesia, fentanyl infusion was started 10–20 μg/h with titration to keep visual analogue scale (VAS) less than 30 mm on a 0–100 mm scale. Postoperative recovery was uneventful and patient was discharged after four days.

3. Discussion

Blue rubber bleb nevus syndrome (BRBNS) is a rare condition characterised by multiple malformations of the venous system. These malformations are most commonly present in the skin

and gastrointestinal tract, however any organ may be involved (Table 1). Most cases of BRBNS are diagnosed at birth or during first few years of life but few cases remain undiagnosed until adulthood. The clinical manifestations depend upon the site of the lesions. These lesions where ever present carry significant potential for bleeding.

4. Anaesthetic concerns

Haemangiomas of nose, lips, tongue, uvula, pharynx, and trachea [4] have been noticed. Nasal and lip lesion may make mask holding an issue as any pressure may lead to skin breech and a bleeding thereafter. Parotid gland lesions and lip lesions may be painful and associated with decrease mouth opening and render the airway difficult. Cervical vertebra lesions can reduce neck movement and paravertebral lesions may lead shift of trachea away from midline. All these associated findings may lead to difficult bag and mask ventilation as well as difficult intubation [4]. MRI and indirect laryngoscopy (IDL) should be done in advance to map any upper airway haemangiomas. Fibreoptic bronchoscopy (FOB) guided intubation or gentle laryngoscopy is desirable to avoid any bleeding from these lesions. In our patient we did a careful airway examination and after induction of anaesthesia, gentle laryngoscopy was done uneventfully. Gonzalez-Pizarro et al. [4] suggested the use of fibre optics to avoid any airway bleeding in these patients.

Meticulous positioning and padding of pressure points are required, especially during gastrointestinal excision of lesions because of the long duration of procedures. Vertebral lesions may cause vertebral collapse or spinal cord compression [5] and preoperative MRI is essential. Epidural venous plexus haemangioma may cause epidural haematoma during needle placement [6]. The chances of bleeding in a patient with chronic anaemia, thrombocytopenia and altered coagulation [7,8] make neuraxial anaesthesia risky. Because of these reasons we decided to avoid epidural placement in this patient. Our patient was having lesions in both the arms; therefore, we inserted an arterial cannula to avoid repeated friction and compression applied by NIBP and possible bleeding from the denuded lesions.

Recurrent blood loss from the gastrointestinal tract leads to anaemia, and if the blood loss is acute and large in amount can lead to cardiovascular decompensation [9]. These haematological abnormalities require meticulous preoperative evaluation in the form of peripheral blood smear and coagulation profile, and may be associated with increase blood loss intraoperatively; therefore blood and blood products should be arranged for perioperative period. Use of NSAIDs should be avoided because of possibility of platelet defects.

Haemangiomas of brain can affect the blood supply to the brain may cause nervous system dysfunction, which may be associated with significant morbidity and mortality. Focal neurological deficits, strokes, epilepsy and dementia have been reported with BRBNS. A smooth induction without any intubation response (reflex hypertension and tachycardia) is advocated and desirable to avoid any sudden cerebral haemorrhage. Drug interaction of antiepileptic drugs with anaesthetic drugs should be kept in mind.

We preferred general anaesthesia as the first option. Neuraxial blockade could not be given as there was a hypodense lesion (probably a vascular nevus) in the erector spinae muscle at the level of L2–3. Apart from routine monitoring (ECG, NIBP, pulse oximetry), invasive BP monitoring was also done.

In our case, the various concerns were paediatric age group, prolonged duration of surgery, meticulous airway manipulation, hypothermia, IV fluid and blood loss management. Postoperative analgesia was achieved with fentanyl infusion 10–20 μg/h. If neuraxial blockade is not contraindicated, patient-controlled epidural analgesia (PCEA) or epidural infusion of morphine or fentanyl may be given for analgesia.

The treatment of the GI lesions and bleeding is usually supportive and symptomatic in the form of blood transfusions and

Table 1 The various organ systems involvement and their anaesthetic concerns in a patient with blue rubber bleb nevus syndrome.

Organ involvement	Characteristic findings	Anaesthetic concerns	Management
Airway [4]	Haemangiomas of nose, lips, tongue, uvula, pharynx, trachea	Leads to difficult in bag and mask ventilation, possible airway bleeding during intubation, painful lesions leading to decrease mouth opening, tracheal shifting away from midline	Detailed airway examination, indirect laryngoscopy, fibre-optic bronchoscopy guided ETT intubation
Orthopaedics and spine [5]	Bony deformity, pathological fractures, articulation problem, painful joints, paravertebral lesions, vertebral collapse, epidural haemangiomas	Difficulty in positioning, increased propensity to fracture, restricted flexion-extension, epidural haematoma and difficult central neuraxial blockade because of vertebral collapse	Careful positioning, MRI evaluation of spine, avoid multiple attempts for neuraxial blocks
CNS [6]	Haemangiomas, dementia, strokes, seizures	Focal neurological deficits, intraoperative strokes and seizures	Smooth induction, avoid intubation response, sudden increase in blood pressures
Haematological [7] and CVS [8]	Chronic anaemia, Chronic DIC (Consumptive coagulopathy)	May present with CHF Intraoperative excessive bleeding	Evaluation, iron folic acid supplementation, coagulation profile prior to surgery, arrange adequate amount of blood and blood products
Gastrointestinal (GI) [9]	GI haemangiomas, chronic spontaneous bleeding to fatal acute bleeding, acute abdomen	Anaemia and emergency surgery	Prolong duration of surgery

iron supplements. The role of surgery is dependent on the presentation, location and extent of the GI haemangiomas. Number of lesions cannot be predicted preoperatively, usually numbers of lesions are huge and extensive surgery is required. Long midline incision is given usually and duration of surgery is quite prolonged. Hypothermia is a risk because of prolong duration of surgery and bowel exploration. We used hot line fluid warming system (HL-90, Level 1, Smiths Medical, USA) and warming blankets to prevent hypothermia. Blood loss in GI haemangioma excision is not much unless until there is some baseline bleeding abnormality but blood products should be arranged.

In conclusion BRBNS is a rare disease entity and may require routine as well emergent surgical procedures. Careful evaluation and inspection are the key for fruitful anaesthetic management. Haemogram, coagulation profile, liver function tests, renal function tests, chest X ray, MRI brain, head and neck and spine should be done. Detailed airway and spine examination should be done along with inspection of joint lesions which may lead to difficulty during positioning. Other organ involvement should be sought and managed accordingly. Blood should be cross-matched and arranged. Airway management should be done with extreme care and use of FOB may be considered, with difficult airway cart in vicinity. Smooth induction is very important along with prevention of hypothermia intraoperatively. Postoperative pain can be managed with IV or epidural opioids.

Conflict of Interest

None.

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