

CASE REPORT

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# Anaesthetic management of Castleman's disease associated with paraneoplastic pemphigus: report of a rare case

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## Abstract

**Background:** Castleman's disease is a rare lymphoproliferative disease which can mimic other malignant tumours and can be associated with paraneoplastic pemphigus especially in the paediatric age group. This presentation can be challenging even to the seasoned anaesthesiologist when it comes to managing such a case presenting for laparotomy.

**Case presentation:** Here such a case in a 12-year-old who was initially diagnosed as retroperitoneal sarcoma complicated with paraneoplastic pemphigus and inferior venacaval thrombus is presented and its anaesthetic management and challenges are discussed. Special care needs to be taken while anaesthetizing such a patient keeping in mind the difficult airway, adrenocortical suppression, and exacerbation of skin lesions due to various anaesthetic procedures. For this case, the histopathology turned out to be Castleman's disease.

**Conclusion:** The anaesthetic challenges associated with paraneoplastic pemphigus in paediatric age group receiving perioperative thromboprophylaxis need special mention. Here we are discussing the challenges and anaesthetic management of such a case as literature on this aspect is few.

**Keywords:** Castleman's disease, Paraneoplastic pemphigus, Retroperitoneal Tumour, Anaesthetic management

## Background

Castleman's disease is a lymphoproliferative disorder with three histopathological variants and is very rare (Dinesha et al., 2014). Pemphigus induced due to neoplasm is known as paraneoplastic pemphigus (PNP) and was first described by (Anhalt et al., 1990). In the paediatric age group, there is a striking association between the two (Choh et al., 2014), but the reported cases are rare (Powell et al., 1986). It is characterised by chronic mucocutaneous erosions and blisters.

Anaesthetic concerns include primarily airway management due to facial, oropharyngeal, and laryngeal lesions leading to difficult intubation and bleeding with added risk of aspiration (Dave et al., 2007). Another concern is care of skin and mucus membranes during positioning the patient for surgery and other procedures. Since the patients are likely to be on prolonged steroid therapy, adrenocortical suppression

should be tackled in perioperative period (Lever & Schaumburg-Lever, 1977).

Here we report the anaesthetic management of a patient initially diagnosed as retroperitoneal sarcoma complicated with inferior venacava thrombus with paraneoplastic pemphigus, which after surgical resection and histopathology examination turned out to be Castleman's disease of hyaline vascular type. She was planned for exploratory laparotomy for excision of retroperitoneal tumour and removal of IVC thrombus. She had lesions over oral cavity, mucus membrane, and skin (Fig. 1) and was on long-term steroid coverage.

## Case report

A 12-year-old Indian girl presented with colicky abdominal pain associated with vomiting. On examination, a hard mass was felt in the right lumbar region and hypochondrium, immobile and tender around 10 × 10 cm. CT abdomen showed retroperitoneal mass 10.2 × 8.7 × 6.8 cm displacing the inferior venacava to the left side

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**Fig. 1** Lesions over the lips

and in close relation to the right lateral wall of inferior venacava.

Patient developed multiple raised red lesions over chest, abdomen, back, upper and lower limbs, and oral ulcers with crusts over lips over a period of 6 months after diagnosis of the tumour. Dermatology consultation was done and was diagnosed as paraneoplastic pemphigus by histopathology. Steroid therapy was initiated.

At the time of preoperative evaluation, the patient had steroid facies, with bleeding dry crusts over lips. Mouth opening was less than 1 finger and Mallampati classification grade 4, since she had painful scarred oral lesions and was unable to protrude tongue out fully. Healed hyperpigmented lesions were present over hands, chest, abdomen, and back. She weighed 57 kg with a height of 152 cm and BMI 24.7 (Figs. 2, 3 and 4).

Routine blood investigations and arterial blood gas analysis were done. Serum cortisol levels were low due to prolonged exogenous steroid therapy. Associated diseases such as rheumatoid arthritis, myasthenia gravis, collagen disease, and pernicious anaemia were ruled out through investigations. Since she had a suspicious inferior venacava (IVC) thrombus, she was on warfarin orally (3 mg/day) which was bridged with unfractionated heparin 5000 U eighth hourly intravenously (iv) during the preoperative period. Her liver function and renal function tests were within normal limits.



**Fig. 2** Healed lesions over foot

As this was a case of wide excision of the retroperitoneal tumour along with IVC thrombectomy in a paediatric patient, general anaesthesia was planned. The patient received pantoprazole 40 mg and domperidone tablet night before and morning of surgery as gastric ulcer prophylaxis. Hydrocortisone 200 mg, glycopyrrolate 0.2 mg, midazolam 1 mg, and fentanyl 100 micrograms ( $\mu\text{g}$ ) were given iv before induction of anaesthesia. Anaesthesia was induced with propofol 100 mg and succinylcholine 100 mg iv. Gentle bag and mask ventilation performed by applying minimal pressure over the face mask, keeping in mind the chances of worsening the bullous lesions if excessive pressure was applied. Intubation was done with video laryngoscope gently with cuffed endotracheal tube 7-mm internal diameter. Cuff inflated with air and cuff pressure was checked and kept at 15 cm of  $\text{H}_2\text{O}$ . Nasogastric tube was inserted and position confirmed. Both tubes were secured with single strip of elastic adhesive bandage. Eye ointment was applied and protected with wet cotton gauze. The left radial artery and two peripheral veins were cannulated with large bore cannulas and were secured with utmost care by placing minimal adhesive. Adequate pressure padding was done at all bony prominences. Right internal jugular vein cannulated under ultrasound guidance and secured with utmost care. Monitoring included standard ASA monitors along with invasive arterial blood pressure and cardiac output monitoring. Goal directed fluid therapy depending on stroke volume variation as obtained from the cardiac output monitor, with



**Fig. 3** Healed lesions over hand



**Fig. 4** Abdominal striae due to steroid induced obesity

balanced salt solutions and albumin was followed during the intraoperative period. Anaesthesia was maintained with 50% Air Oxygen with sevoflurane along with fentanyl and vecuronium infusions. Surgery lasted for 5 h and the procedure was wide excision of the tumour along with vascular repair of the inferior venacava. The intraoperative period was uneventful. After completion of surgery, patient was shifted to intensive care unit and electively ventilated. Steroid therapy was continued postoperatively as well as gastric ulcer prophylaxis continued with iv pantoprazole 40 mg once daily. No new lesions developed in the patient and dermatology review was obtained during postoperative period. Analgesia was provided with continuous iv fentanyl infusion (20  $\mu$ ) and paracetamol eighth hourly. She was extubated the next day, shifted out of intensive care unit on third postoperative day (POD), and discharged on seventh POD.

## Discussion

The association of PNP with Castleman's disease is around 10% (Anhalt et al., 1990). The major problems an anaesthesiologist encounters while dealing with a case of paraneoplastic pemphigus are as follows: (1) Difficult airway, (2) Adrenocortical suppression, (3) Difficulty in securing iv access, (4) Postextubation airway complications, (4) Risk of flaring up of the lesions despite adequate steroid cover.

During preoperative evaluation, the site of tumour and the centrality also should be taken into consideration as multicentric tumours can present with mediastinal widening, tracheal compression and result in central airway obstruction at induction. The association with rheumatoid arthritis, adrenal neoplasm (Shi et al., 2009), and myasthenia gravis should be ruled out in a case presenting with pemphigus (NAYSMITH & Hancock, 1976). Another serious issue that needs to be considered is the development of bronchiolitis obliterans in these patients which can cause respiratory failure (Wang et al., 2005).

Regional anaesthesia is preferred if feasible (Abouleish et al., 1997), but care should be taken to avoid needle insertion through the lesions and also introduction of infection through denuded skin (Bansal et al., 2012). Preparation for difficult airway in anticipation of the same should be made. Bag mask ventilation should be done gently as also laryngoscopy and intubation. Keeping in mind the steroid treatment, adequate dose of iv steroid should be administered before induction of anaesthesia. Particular care should be given to pressure points and adequate padding to be done. While securing vascular lines, care must be taken to avoid excessive use of adhesive bandages. Securing an epidural catheter also poses problems as the use of adhesive tapes might flare up Kobner's phenomenon (Baykal et al., 2002). Severe oropharyngeal lesions can produce airway bleeding during intubation and laryngeal edema and stridor during extubation. Due to the painful oral lesions, intake and nutrition may be poor and during preoperative evaluation, adequate nutritional screening is also warranted to rule out fluid and electrolyte imbalance and hypoalbuminemia (Lavie et al., 1984). For cases in which regional techniques are not feasible or relatively contraindicated, multimodal analgesia utilising opioid infusions via patient-controlled analgesia pumps and paracetamol iv can be safely used. Wound infiltration is not recommended for this group of patients as it can lead to sloughing (Prasad & Chen, 1989).

In this particular case, the problems described initially along with the suspicious IVC thrombus necessitating vascular repair and paediatric age group aggravated the difficulty prohibiting placement of epidural catheter and the need for sole GA and iv drugs for analgesia.

For unicentric Castleman's disease, the removal of the tumour helps resolve the associated skin and mucus membrane lesions. The patient in this report on serial reviews has shown improvement in her general condition and steroids have been stopped due to gradual resolution of the skin lesions.

## Conclusion

In conclusion, PNP in paediatric age presenting for surgery is very rare and ultimate care should be taken in dealing with the airway and providing adequate analgesia and protecting the pressure points while positioning the patient so that exacerbations of the lesions can be avoided.

## Abbreviations

iv: Intravenous; GA: General anaesthesia; IVC: Inferior venacava; POD: Postoperative day;  $\mu$ : Micrograms; PNP: Paraneoplastic pemphigus; BMI: Body mass index; CT: Computed tomography

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**Author's contributions**

RO contributed to conceptualization and design, drafting, and revision of the report. DVG contributed to design, drafting, and revision of the report. AJ contributed to drafting and revision of the report. RCK contributed to revision, supervision, and approval of the report. The other authors read and approved the final manuscript.

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**Consent for publication**

Consent has been obtained from the patient's parents for publication of personal data and pictures for academic purpose. Assent from the patient has also been obtained.

**Competing interests**

The authors declare that they have no competing interests.

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