Proteus syndrome: A medical rarity

Sir,

Proteus syndrome (PS), named after the Greek Sea-God Proteus, is a complex hamartoneoplastic disorder with multisystem involvement. The varied presentation and extent makes the anesthetic management challenging.

A 22-year-old female was presented to the Emergency Room with a history of road traffic accident with complaints of pain and distension of the abdomen. She had previously been diagnosed to have PS by the dermatologists at our institute with features of asymmetric overgrowth of lower limbs, lumbar scoliosis and skin lesions [Figures 1 and 2]. On admission, she was conscious and alert with a heart rate of 130/minute and blood pressure of 90/60 mmHg. Airway assessment was consistent with Mallampatti class 3. Systemic examination revealed extreme pallor and a tense, tender distended abdomen. Initial investigations revealed hemoglobin of 2.8 g/dl. Other investigations were unremarkable except the CECT abdomen which showed splenomegaly with a splenic laceration.

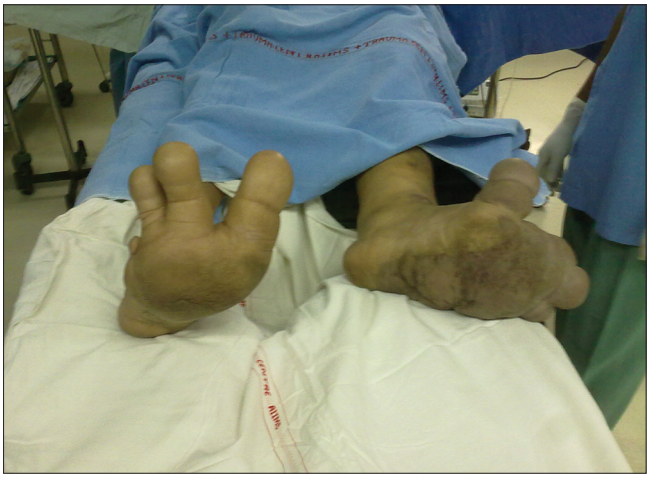

Figure 1: Asymmetric overgrowth of lower limbs

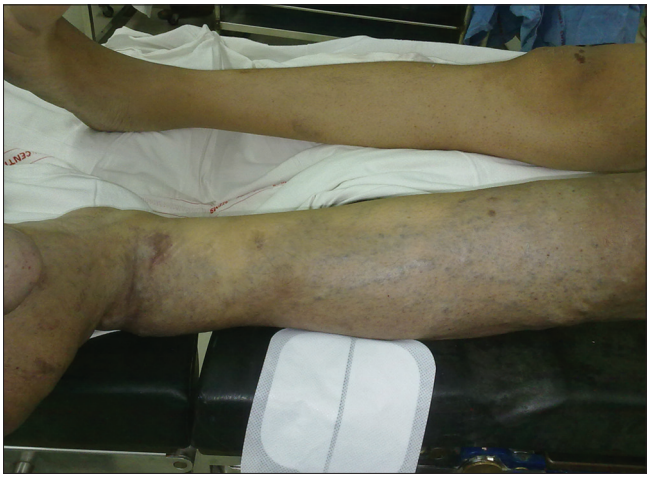

Figure 2: Multiple skin lesions and venous engorgement over the left leg

Patient was accepted for an emergency laparotomy under ASA grade 4. Laryngoscopy revealed a hanging enlarged epiglottis which necessitated the use of a Mccoy laryngoscope for intubation. Splenectomy was done and hemostasis achieved. Patient was shifted to the intensive care unit for post-operative elective ventilation and monitoring. After the repeat coagulation profile was normal, USG guided epidural catheter insertion was done. Low molecular weight heparin (LMWH) was initiated two hours later for DVT prophylaxis .The patient was extubated after 24 hours of mechanical ventilation and subsequently made a satisfactory recovery.

PS is a complex disorder which because of its rarity and protean manifestations defies easy classification or description. It has been called a hamartoneoplastic disease characterized by overgrowth of various tissues. Our patient had overgrowth of the lower limbs, varicose veins, purplish discoloration of the skin and lumbar scoliosis.

Pennant *et al.* reported a 14-year-old boy who had general anesthesia for orthopedic surgery with difficult intubation.[1] Fibreoptic-aided intubation was done under inhalational anesthesia. Pradhan *et al.* reported a 7-year-old boy who presented for finger reconstruction who had high arched palate.[2] In our patient, an elongated hanging epiglottis was revealed on laryngoscopy. Scoliosis is frequently observed in older PS patients and one-fifth of the patients have overgrowth of a vertebra.[3] Our patient was noted to have a lumbar scoliosis. Pulmonary embolism with or without deep venous thrombosis has been reported.[4] Our patient received deep vein thrombosis prophylaxis and early ambulation was ensured. The most frequently reported pulmonary anomalies in PS are cystiform anomalies emphysema, atelectasis and fibrosis. There was no evidence of any pulmonary complications in our patient.

We thought it relevant to report this case as several aspects of this condition are important for the anesthetist. A difficult airway, suboptimal positioning of the patient

because of the skeletal malformations, spine deformities, propensity for thromboembolism and respiratory disease, all add up to leave the anesthetist in a challenging situation. Since only about 200 cases of PS have been reported in literature, and even fewer requiring care from an anesthetist, it is highly probable that more is yet to be learned about this condition.

Chandni Sinha, Babita Gupta, Manpreet Kaur, Ajeet Kumar, Chandan Kumar Dey

Department of Anaesthesia & Critical Care, JPNA Trauma Centre, All India Institute of Medical Sciences, New Delhi, India

Address for correspondence:
Dr. Chandni Sinha,
211, JPN Apex Trauma Centre Hostel,
Raj Nagar, New Delhi - 110 029, India.
E-mail: chandni.doc@gmail.com

REFERENCES

1. Pennant JH, Harris MF. Anaesthesia for Proteus syndrome. Anaesthesia 1991;46:126-8.
2. Pradhan A, Sen I, Batra YK, Biswas G. Proteus syndrome: A concern for anaesthesiologist. Anesth Analg 2003;96:915-6.
3. Azauz EM, Costa T, Fitch N. Radiologic findings in the Proteus syndrome. Pediatr Radiol 1987;17:481-5.
4. Slavotinek AM, Vacha SJ, Peters KF, Biesecker LG. Sudden death caused by pulmonary thromboembolism in proteus syndrome. Clinical Genet 2000;58:386-9.

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