Post-operative airway obstruction in Noonan syndrome: An unusual presentation

INTRODUCTION

Noonan syndrome (NS) is an autosomal dominant disorder with craniofacial, cardiac, skeletal abnormalities, haemorrhagic diathesis, with reported incidence of 1 in 1000 to 1 in 2500 live births.[1] Various lymphatic abnormalities have been observed in these patients including pulmonary and intestinal lymphangiectasia and lymphoedema.[2] Craniofacial dysmorphism webbed neck and atlantoaxial instability can complicate airway management.[3] The anaesthetic considerations include difficult airway, impaired cardio-pulmonary function, mental retardation, and short stature.

We report an unusual presentation of airway obstruction due to bronchial cast on 5th post-operative day in a case of NS, who underwent laparoscopic orchidopexy.

CASE REPORT

A 15-year-old male weighing 28 kg, 112 cm tall and diagnosed case of NS was posted for laparoscopic orchidopexy for bilateral undescended testes. He had no other complaints except undescended testes. Birth history was normal, but he had delayed milestones and low intelligence. He was short statured with short neck, high arched palate, adequate mouth opening, and Mallampati airway class II. His pulse rate (PR) was 70/min, blood pressure (BP) was 90/60 mm Hg. There was a systolic murmur in the pulmonary area and two-dimensional echo showed pulmonary stenosis with pulmonary artery systolic pressure of 40 mm Hg. Chest X-ray (CXR) showed prominent broncho-vascular markings. Coagulation profile and biochemical investigations were within normal limits.

During laparoscopic orchidopexy, there was a major vascular injury, which was repaired and blood loss was adequately replaced. The rest of the intraoperative course was uneventful. Patient was extubated and shifted to Intensive Care Unit (ICU). As ICU stay was unremarkable, he was moved to wards on 2nd post-operative day.

On 5th post-operative day, the patient suddenly developed fever with breathlessness. Breath sounds were absent on left side. CXR showed homogenous opacity in left hemi thorax with collapsed lung. He was shifted back to ICU. He was conscious, oriented with a PR of 90/min, BP of 100/60 mm Hg, respiratory rate of 35/min, and oxygen saturation (SpO_2) fluctuating between 85% and 90% on room air. Patient was put on humidified oxygen 4 L/min and ipratropium and budesonide nebulisation. The cause of this sudden, event was thought to be thick mucoid secretions or aspiration of foreign body. Patient was taken up for rigid bronchoscopy.SpO_2 was 88–90% on oxygen, HR 92/min, and BP 98/62 mm Hg. Patient was pre-medicated with intravenous (IV) glycopyrrolate (4 μgm/kg), and induced with ketamine (2 mg/kg), sevoflurane and maintained on atracurium (0.5 mg/kg), and sevoflurane with the help of ventilating bronchoscope. On bronchoscopy, a tissue mass lining the left main bronchus was visualised. It was difficult to retrieve en masse even after multiple attempts with saline instillation. Intra-operative SpO_2 kept on fluctuating. Patient was then intubated with 5.5 mm cuffed oral endotracheal tube. End-tidal carbon dioxide ($EtCO_2$) was 50–55 mm Hg and SpO_2, 85–90%. Endotracheal suctioning with saline instillation brought out fibrinous material. Active physiotherapy was given. Following this, the saturation improved to 93–95% with $EtCO_2$ of 30 mm Hg. Keeping equipment ready for re-intubation, the neuromuscular block was reversed, and the patient was extubated. Immediate post-extubation, a chunk of tissue like material was coughed out by the patient [Figure 1]. It was rubbery in consistency and on spreading out, took the shape of the bronchial tree [Figure 2]. There was a dramatic improvement in saturation (SpO_2 98–99%) and air entry. Patient was put on ipratropium and budesonide nebulisation 6 hourly, which was continued for the next 48 h with IV antibiotic cover. Post-operative CXR showed significant improvement. The rest of the post-operative stay was uneventful. Patient was discharged on day 3 after bronchoscopy. On subsequent follow-up, the patient was asymptomatic. Histopathology report of the tissue mass revealed neutrophils, alveolar macrophages, fibrinous and mucinous material, diagnosed as a bronchial cast.

DISCUSSION

Lymphatic abnormality seems to be inherent in patients with NS. About 20% of NS patients may have lymphatic vessel dysplasia, hypoplasia or aplasia. This

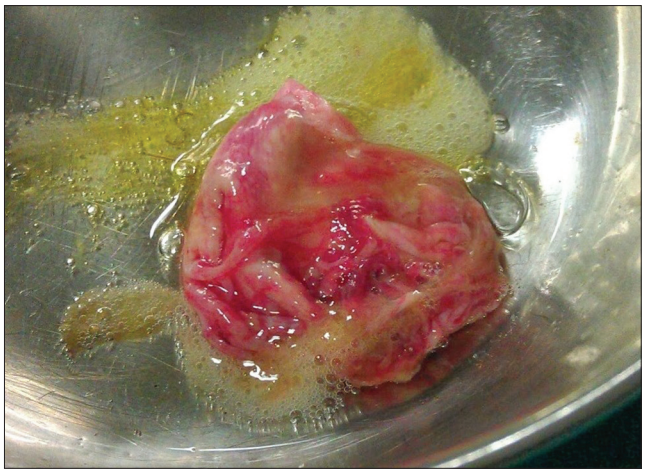

Figure 1: Coughed out tissue

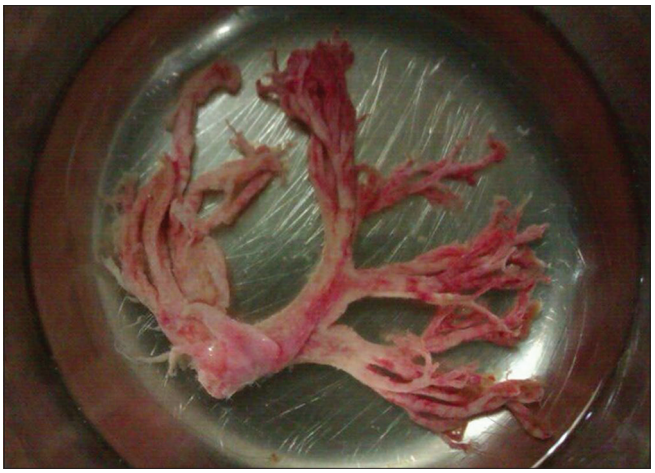

Figure 2: Spread out bronchial cast

may cause generalised or peripheral lymphoedema, pulmonary, and intestinal lymphangiectasia.[2] Borgan *et al.* reported bronchial cast formation in NS following surgery for Tetralogy of Fallot.[4] In the present case, no thoracic intervention was done and the patient had no previous history of cough or respiratory difficulty. In retrospect, the reason for the cast formation could be attributed to elevated pulmonary arterial pressure. A literature search revealed that bronchial casts are formed in a condition called plastic bronchitis (PB). PB is a rare condition described more commonly in children than adults. It has been reported following surgeries for congenital heart disease, especially Fontan's procedure, asthma, cystic fibrosis, aspergillosis, and smoke inhalation.[4] Due to acute and vague presentation, these patients may have a mortality rate as high as 29%.[4,5] The cause is usually asphyxiation due to airway obstruction. The composition of these casts is consistent with lymphatic fluid. Probably high pulmonary or intrathoracic lymphatic pressure and/or presence of undetected lymphobronchial fistulae may contribute to the formation of these casts.[6] The most common clinical presentations include respiratory distress and coughing up of material like 'chicken meat' or 'noodles'.[7] Recurrence of the cast with repeated bronchoscopies has been reported.[8]

Seear *et al*. classified these casts into type I (cellular) and type II (acellular).[9] The casts are cohesive in nature and take the shape of bronchi in which they are formed. They are adherent but friable. They may be spontaneously expectorated or may require urgent bronchoscopy. Flexible bronchoscopy may help in diagnosis but rigid bronchoscopy is required for removal of the cast with the use of forceps during bronchoscopy. Irrigation with normal saline causes these casts to swell and thus easier to grasp. We irrigated with saline but still could not retrieve it and ultimately patient coughed it out after extubation.

In addition to bronchoscopy, adjuvant medical treatment with mucolytics, macrolide antibiotics, and tissue plasminogen activator have been used for cast disruption.[4] Chest physiotherapy with steroid and saline nebulisation is frequently employed as an adjunct for cast mobilisation.

CONCLUSION

Awareness and ability to diagnose PB leading to airway obstruction in patients with NS are crucial to reduce morbidity and mortality with prompt and effective treatment.

ACKNOWLEDGEMENTS

Authors acknowledge support from Dr. Shilpa V. Deshmuhk and Dr. Debasis Debata, Resident Anaesthesiologists for support in patient management.

Sheetal R Jagtap, Hemalata R Iyer, Rochana G Bakhshi, Hemant N Lahoti

Department of Anaesthesiology and Paediatric Surgery, Pad. Dr. D. Y. Patil Medical College, Hospital and Research Centre, Mumbai, Maharashtra, India

Address for correspondence:
Dr. Sheetal R Jagtap,
Department of Anaesthesiology and Paediatric Surgery,
504, Sayia CHS, Plot E/01, Sector 12, Kharghar,
Mumbai - 410 210, Maharashtra, India.
E-mail: drsheetaljagtap@gmail.com

REFERENCES

1. van der Burgt I. Noonan syndrome. Orphanet J Rare Dis 2007;2:4.
2. Hernandez RJ, Stern AM, Rosenthal A. Pulmonary lymphangiectasis in Noonan syndrome. AJR Am J Roentgenol 1980;134:75-80.
3. Samra T, Banerjee N. Anaesthesia for emergency ventriculo-peritoneal shunt in an adolescent with Noonan's syndrome. Indian J Anaesth 2014;58:452-5.
4. Brogan TV, Finn LS, Pyskaty DJ Jr, Redding GJ, Ricker D, Inglis A, *et al.* Plastic bronchitis in children: A case series and review of the medical literature. Pediatr Pulmonol 2002;34:482-7.
5. Do TB, Chu JM, Berdjis F, Anas NG. Fontan patient with plastic bronchitis treated successfully using aerosolized tissue plasminogen activator: A case report and review of the literature. Pediatr Cardiol 2009;30:352-5.
6. Shah SS, Drinkwater DC, Christian KG. Plastic bronchitis: Is thoracic duct ligation a real surgical option? Ann Thorac Surg 2006;81:2281-3.
7. Eberlein MH, Drummond MB, Haponik EF. Plastic bronchitis: A management challenge. Am J Med Sci 2008;335:163-9.
8. Pawar SS, Chun RH, Rao AR, Kerschner JE. Management of plastic bronchitis in a child with mild intermittent asthma. Ann Otol Rhinol Laryngol 2011;120:697-9.
9. Seear M, Hui H, Magee F, Bohn D, Cutz E. Bronchial casts in children: A proposed classification based on nine cases and a review of the literature. Am J Respir Crit Care Med 1997;155:364-70.

Access this article online	
Quick response code	Website: www.ijaweb.org
	DOI: 10.4103/0019-5049.160961