

Treatment of Vomiting Attacks in Patients with Williams Syndrome Using Mianserin and Telazine

The Editor,

Sir,

Williams syndrome is a genetic syndrome that comprises developmental delay and heart defects, low muscle tone, widely spaced teeth, a long philtrum and a flattened nasal bridge and premature social relationships. The ratio is 1:20 000 (1, 2). A special treatment approach is not known to change the vomiting attacks, eating disorders and premature social relationships. Here, we wish to put forward that mianserin and telazine combination had positive outcomes in a patient with Williams syndrome.

The patient is a 23-year old single woman who is flirtatious and very social. According to her mother, she focusses on unnecessary things; she shops too much and memorizes the phone numbers from the phonebook. She vomits a lot; anti-emetic drugs are insufficient. She has insomnia. She wants to be alone when she is angry. She has an unrestrained interest in men and always wants to meet with someone. There are grammar mistakes in speech but not errors of logic. She has visual impairment, 7.5 degree myopy, and is astigmatic.

Her vomiting episodes started at 15 years of age. Up to two years of age, she would only eat a cup of baby food for the entire day. In childhood, there was a problem getting her to eat but she would take fruits. She completed her primary school with some special help. Her writing skills were good. After primary school, she could not read any books. Her appearance was different from others and she did not know how to laugh and she had enuresis. Doctors said that she had some muscle issues. When she was 15 years old, her father threatened her and that caused a deep trauma. Because of that trauma, her vomiting problem started (physiologic defect with a deep psychological trauma). Her parents are not related to each other, but are from the same geographic region. Genetic studies revealed deletion in the gene for elastin and she was diagnosed as Williams syndrome.

She could not tolerate the selective serotonin reuptake inhibitor (SSRI) and tricyclic antidepressant (TCA) group of anti-depressives and atypical anti-psychotics that were given by doctors. She started to take mianserin 10 mg but her complaints continued so the dose was increased to 15 mg and 1 mg telazine was added. Her vomiting suddenly stopped and hyperphagia occured. She gained five kilograms. After these results, the mianserin dose was decreased to 5 mg per day; the telazine dose was not changed. At this dose, hyperphagia disappeared, trichiniasis and vomiting stopped and she recovered. Her psychological demanding feature and exaggerated social relationships decreased.

We therefore suggest a combination of mianserin and telazine for patients with the above-named complications in Williams syndrome.

Keywords: Mianserin, telazine, Williams syndrome

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REFERENCES

Initial Bronchoscopic Treatment of Tracheal Schwannoma: A Rarely Seen Tumour

The Editor,

Sir,

Primary tracheobronchial schwannomas are relatively rare neoplasms found in the trachea. Benign types of these tumours were rarer than 0.5% (1). They can present with chronic cough, progressive dyspnoea and obstructive symptoms but they are usually asymptomatic since the mass lesion grows significantly (1).

A 42-year old male non-smoker was admitted to our hospital with dyspnoea on exertion, having no other disease than Type 2 diabetes mellitus. Chest X-ray of the patient is given in Fig. 1. His bronchoscopic evaluation showed a mass lesion covered by mucosa on the left wall occluding 80% of the lumen (Fig. 2A). The left main bronchus was occluded at its entrance by the tracheal mass. The lesion was photocoagulated by diode laser and taken out by core out method. Coagulation with diode laser was repeated to the base to control bleeding (Fig. 2B).
Preferred diagnostic method for these tumours should be rigid bronchoscopy (3); this helps to measure the true size of the tumour, size of the tumour-free trachea that could be needed for the reconstruction after surgery, and to control bleeding that could occur after biopsy.

The definitive treatment is surgical resection with a healthy margin of tissue (4, 5). The standard management of tracheal tumours is circumferential resection with tracheal reconstruction, which often completely cures a schwannoma (4). Endoscopic laser resection was used to control the base of the tumour. The choice of treatment should be influenced by the endoscopic presentation of the tumour (ie pedunculated or sessile), the risk of tracheal resection and the presence or absence of extratracheal component (6). However, if the tumour originated with a wide base from the tracheobronchial tree or if the cartilage layer was not intact, bronchoscopic treatment will not be curative. In tumours which do not fulfil both of the criteria, surgical resection is the only curative approach. The gold standard treatment is surgical resection to protect from recurrences.

Keywords: Diode laser, interventional, rigid bronchoscopy

Histopathological examination of the specimen revealed a tumoral infiltration composed of loose fascicles of spindle shaped cells with hypo-hyperchromatic nuclei consistent with mesenchymal neoplasm (Fig. 3A). Immunohistochemical staining was done to differentiate benign mesenchymal tumours. Pathology results identified fusiform cell mesenchymal tumour with diffuse and widespread S-100 and focal EMA positivity (Fig. 3B). The tumour was immunohistochemically and histopathologically compatible with benign schwannoma. The patient was sent to surgery to provide curative treatment. The patient was followed-up for a year and no recurrence was observed.

Schwannoma is a benign, slowly growing neoplasm of Schwann cells that may arise in any nerve. The endobronchial origin is extremely rare. Tracheal primary neurogenic tumours usually have a long natural history, causing symptoms only after they have reached a significant size (2).
Airway Obstruction and Inability to Ventilate Due to Swollen Uvula following Adenotonsillectomy in a Three-year Old Child

Dear Editor,

Sir,

Adenotonsillectomy is one of the commonly done day care surgical procedures in paediatric patients. Airway obstruction following adenotonsillectomy requiring intubation in paediatric patients is one of the potential complications, and has been reported only in two cases in the literature (1, 2). In neither of the two cases was ventilation reported to be difficult. The inability to ventilate can be catastrophic. We present the first reported case of airway obstruction and inability to ventilate due to swollen uvula following adenotonsillectomy.

A three-year old girl, weighing 15 kg, with recurrent attacks of tonsillitis was scheduled for elective adenotonsillectomy. She also had features of obstructive sleep apnoea (OSA). Apart from being allergic to penicillin, the preoperative assessment was unremarkable. The child was induced with propofol (30 mg), fentanyl (30 µg) and cisatracurium (2 mg). Following uneventful mask ventilation, orotracheal intubation was achieved on first attempt with 5 mm uncuffed tracheal tube. The patient was positioned with neck extension and mouth opened with Davis Boyle mouth gag. Adenoids were removed using a curette. Both tonsils were dissected and removed by applying a tonsillectomy snare to the lower poles. Haemostasis was achieved with packing and diathermy of bleeding vessels. In this child, the dissection of tonsils was technically difficult, resulting in longer operative time. Hence, laryngoscopy was done prior to extubation and a moderate uvular swelling was noted (Figure). Dexamethasone 4 mg and ranitidine 15 mg were administered intravenously. Neuromuscular blockade was reversed and extubation was performed when the child was awake in the post-tonsillectomy position.

Following extubation, the child was initially breathing normally but progressively developed obstructed breathing which was thought to be due to the observed uvular swelling. Over the next three minutes, SpO₂ started to fall below 90% and she became bradycardic. Attempts to perform mask ventilation with oral airway were unsuccessful. She was thus administered glycopyrrolate IV, followed by suxamethonium (0.5 mg/kg). Mask ventilation was still not possible, suggesting supraglottic swelling only. Urgent laryngoscopy was done and the trachea was intubated with a 5.0 mm uncuffed tube. Following intubation, oxygen saturation and heart rate returned to normal. Nasal endoscopy confirmed worsening swelling of the uvula with no further abnormal findings. The patient’s sedation, dexamethasone and ventilation were con-tinued for 24 hours. The patient’s intensive care unit course remained uneventful and after 24 hours, direct laryngoscopy showed that uvular swelling had subsided significantly. The child was extubated uneventfully and was discharged home on the third postoperative day.

Compared with adults, children have a two-fold higher incidence of fatal respiratory events in the postoperative period following adenotonsillectomy (3). One of the rare causes of airway obstruction following adenotonsillectomy is uvular swelling (4). In this patient, the technical difficulty in performing adenotonsillectomy would have resulted in uvular swelling leading to airway obstruction. The patient had OSA which can cause vascular engorgement of the uvula and compound the problem (5). It is possible that the negative pressure generated in the airway at the onset of spontaneous breathing gradually drew the swollen uvula toward the glottis, resulting in worsening uvular swelling and leading to complete airway obstruction. Hence, bag mask ventilation, even with oral airway, was unsuccessful. Failure to ventilate even after suxamethonium administration rules out laryngospasm. No other causes of airway obstruction were apparent on direct laryngoscopy. In retrospect, having noticed moderate uvular swelling before extubation, elective ventilation until the swelling subsided could have been a better option.

In conclusion, uvular oedema should be recognized as one of the potential causes of airway obstruction following adenotonsillectomy. The obstruction could lead to poten-