Case report

The management of thoracic inlet syndrome associated with Hurler's syndrome: a novel surgical technique

Rauf M. Ahsan a, Sarah A. Early a, Anne O'Meara b, Lars Nølke a, *

a Department of Cardiothoracic Surgery, Our Lady's Children's Hospital, Crumlin, Dublin 12, Ireland
b Department of Paediatric Oncology, Our Lady's Children's Hospital, Crumlin, Dublin 12, Ireland

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Abstract

A 21-year-old male developed significant swelling of his tongue after a respiratory arrest. The patient had a history of Hurler's syndrome. Magnetic resonance imaging (MRI) angiogram delineated that the swelling was due to compression of his internal jugular veins at the level of the first rib, resulting in thoracic inlet obstruction. The standard surgical treatment of thoracic inlet obstruction was not suitable in this patient's case due to his short thick neck and his characteristic Hurler's syndrome body habitus. Therefore, a novel surgical strategy was used to decompress his head and neck vessels. The manubrium was widened using an iliac crest bone graft, stabilised using internal fixation plates and reconstructed with a pectoral muscle flap.

1. Case summary

A 21-year-old male with a known diagnosis of Hurler's syndrome was admitted for the management of a lower respiratory tract infection. He was treated with intravenous (IV) antibiotics; however, on the fourth day he decompensated acutely and had a cardio-respiratory arrest. He was intubated, ventilated and transferred to the intensive care unit. While fully ventilated, over the course of the following 3 days, his tongue and head became progressively swollen, complicating the ventilator management and causing considerable difficulty in maintaining oral care. To prevent tongue ulceration, his mouth was kept in the open position using specialised dental splints (mouth prop and silicon putty splint). A tracheostomy was performed due to the degree of tongue and oro-pharyngeal swelling to maintain the airway and assist ventilation.

Over the following 5 weeks, there was progressive enlargement of the tongue (Fig. 1A). A computed tomography (CT) thorax was performed; however; it was inconclusive in determining the cause of the patient's tongue swelling. A magnetic resonance imaging (MRI) angiogram was performed, which confirmed the diagnosis of thoracic inlet obstruction with bilateral internal jugular vein obstruction (Fig. 1B). After a multidisciplinary team meeting involving oral surgeons, anaesthetists, otolaryngologists and orthopaedic, vascular and cardiothoracic surgeons, a decision was made to attempt surgical decompression. The standard operation for thoracic inlet obstruction is resection of the first rib using either a transaxillary or video-assisted thoracic surgery (VATS) approach. Neither of these techniques was deemed appropriate in this patient's case and it was decided to decompress the patient's thoracic inlet by widening the manubrium and sternum anteriorly. This procedure was chosen over the standard approach for several important reasons. An anterior decompression requires minimal dissection, is reasonably straightforward and can be performed safely in a patient with massive head and neck swelling. It uses techniques that are performed everyday in our department (partial sternotomy) and allows excellent visualisation and access to the manubrium. Most importantly, this technique does not require any single lung ventilation, which is required in both a transaxillary and a VATS resection of the first rib. It was for these reasons that this novel approach was used over a transaxillary or VATS first rib excision.

2. Surgical technique

A low incision was made over the manubrostellar junction, away from the tracheostomy site. The manubrium and sternum were divided vertically to the level of the
Fig. 1. (A) This picture demonstrates the typical habitus of a patient with Hurler’s syndrome. The patient’s short neck can be easily appreciated. The tongue is massively congested with a dental split in place to protect the tongue from the teeth (inset close up of gross tongue swelling). (B) MRI angiogram depicting the extent of internal jugular vein compression at the level of the first rib (arrow). There is no obvious obstruction of the arterial tree.

Fig. 2. (A) Illustration of sternal reconstruction. The manubrium was incised vertically to between the 2nd and 3rd ribs. An iliac crest bone graft was used to stent open the lower portion of the incised manubrium (shaded area). Two compression plates were used to secure the bone graft in position. The superior plate was fixed using four screws (black circles) one screw was placed on either side of the manubrium. The lower plate was fixed using two screws, placed on either side of the manubrium. The manubrium was reconnected to the sternum using a steal wire on either side. (B) Post operative CXR demonstrating the internal fixation device.

second intercostal space and transversely into the same space. The manubrium was stabilised 3 cm apart with a cortical bone graft harvested from the right iliac crest and held in place with two dynamic compression plates (DCP) and 3.5 mm cortical screws and sternal wires. Closure was completed with pectoral muscle flaps that were approximated over the plates and the sternum (Fig. 2A and B).

2.1. Postoperative recovery

Over the next 3–5 days, the tongue reduced in size and the patient was partially able to return his tongue into his mouth. He was also noted to have a bilateral hypoglossal nerve praxia which recovered over the following 5 weeks. The tracheostomy was removed 6 weeks postoperatively. At the time of discharge 43 days following surgical decompression, he had regained full movement of his tongue and was completely able to maintain his tongue within the oral cavity.

3. Discussion

Mucopolysaccharidoses (MPSs) are a group of inherited lysosomal storage diseases resulting from deficiency in the lysosomal enzymes responsible for the degradation of glycosaminoglycans (GAGs), also known as mucopolysaccharides. Type I mucopolysaccharidosis or Hurler’s syndrome is the most common disorder, with a reported incidence of 1.19 per 100,000 population in Europe [1,2]. Interestingly, the prevalence in the Republic of Ireland is considerably higher at 1 per 26,206 births [3]; this has been attributed to the high prevalence of Hurler’s syndrome found in the Irish traveller community.

Clinical features associated with this condition include coarsening of the facial features and upper airway obstruction. Storage of GAGs within the oropharynx with associated enlargement of the tonsils and adenoids can contribute to upper airway complications along with narrowed trachea, thickened vocal cords, redundant tissue in the upper airway and an enlarged tongue [4,5]. Vertebral anomalies, compounded by substrate deposition around the cervical cord, have been reported to cause spinal cord compression [6].

Survival into the third decade is uncommon in Hurler’s syndrome, with less than 100 survivors worldwide in their third decade [7]. The thorax can be very deformed in these patients due to bony abnormalities including kypho-scoliosis, short clavicle, wide, thick and canoe-paddle-shaped ribs and deformed scalpula [6]. There are no reports in the literature detailing the appropriate management of thoracic inlet syndrome in patients with Hurler’s syndrome. Typically, internal jugular vein compression is not a feature of thoracic outlet syndrome that usually affects the brachial plexus and subclavian vessels. The usual causes of internal jugular vein thrombosis and possibly subsequent superior vena cava syndrome include complications of surgical procedures, presence of an indwelling venous catheter, tumour invasion, hypercoagulability, caudal extension of sigmoid sinus thrombosis, compression from adjacent tumour or nodes, reaction to an adjacent infectious process and direct venous injection [8,9].

The classical surgical approach of rib resection in treating thoracic inlet syndrome was deemed inappropriate in this situation. Consideration was given to dental clearance and hemi-glossectomy; however, this would only have dealt with the consequence rather than the cause of the thoracic inlet syndrome. To deal with this unique condition, a novel technique as described above was devised to enlarge the thoracic inlet with minimal dissection of the neck, involving input from a multidisciplinary team, with considerable experience dealing with patients with Hurler’s syndrome. The result was an immediate reduction in tongue swelling and marked improvement in the clinical status of the patient.

To the best of our knowledge, internal jugular venous compression at the thoracic inlet due to Hurler’s syndrome has not been previously reported. The procedure herein described proved successful resulting in reduction of macroglossia and upper airway congestion enabling ultimate discharge home and resumption of premorbid activity.

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