Case Report

Challenges in Management of Infantile Giant Cervico-Oral Cystic Hygromas

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Abstract

Infantile Giant cervico-oral swellings are a threat to life. We describe here two cases of giant cervico-oral cystic hygroma who presented with life threatening airway obstruction soon after birth and were managed by multimodal therapy. Ultrasound guided aspiration; sclerotherapy and surgery were used in both. A tracheostomy and a gastrostomy were required to tide over the crisis period of acute airway obstruction and to establish nutritional support in view of poor and difficult feeding. Both infants were discharged in a stable condition on tracheostomy and gastrostomy and were regularly followed up. Their oral edema and respiration gradually settled and they could be weaned off the tracheostomy after fenestration and partial blocking over few months. Their underdeveloped swallowing reflex and esophageal motility also improved over time and their gastrostomies were removed at follow up. A multimodal therapy helped to improve outcome.

Background

Cystic hygroma, a congenital lymphatic malformation develops at sites of lymphatico-venous communications, commonly the neck, (75%), axilla (20%), retro peritoneum (2%), limbs, chest wall, groin, scrotum, mesentery, parotid (2%), cervico-mediastinal (1%) [1]. Majority of the cases may be asymptomatic and may regress over a period of time or may be managed by intra-lesional injection sclerotherapy or sometimes surgery.

Huge cystic hygromas may cause life threatening respiratory distress and feeding difficulties in infancy where the management may mandate early surgical invention, gastrostomy and tracheostomy. We present two cases of cystic neck masses causing respiratory compromise soon after birth, who received multimodal therapy.

Case 1

A healthy baby boy, delivered en route to the hospital presented with a huge left sided cervico-facial cystic swelling causing respiratory distress soon after birth. The lesion was antenatally diagnosed elsewhere as a multi-loculated cystic lesion occupying the neck, occipital and infra-temporal regions, associated with polyhydramnios. The lesion covered the whole neck and extended from lobule to lobule and lower eyelid to sternum (Figure 1). The clinical diagnosis was cystic hygroma.

The cyst was aspirated twice under ultrasound guidance and hypertonic saline was injected (50ml) at two different sites after aspiration of the straw colored cystic fluid. Mild decrease of around 2 x 2 cm was clinically observed in the swelling but it again rapidly refilled. The dyspnea worsened and the child was intubated in lateral position under ultrasound guidance as the trachea was deviated to the right and the inlet was not visible. The child was given nasogastric feeds, supplemented with parenteral nutrition. A contrast enhanced CT scan showed the extent of the cystic swelling from the skull base, occupying the left cheek, lower half of face, neck with no intra-thoracic extension.

After stabilization, the child was taken up for definitive procedure. Multiple huge macro-cysts and micro-cysts were meticulously dissected, preserving the nerves and vessels of the neck. The micro-cysts extended into the pre-auricular region, para-tracheal and para-pharyngeal region. Partial excision with marsupialization and chemical cauterization with betadine of the residual micro-cysts was done. A right jugular lymph sac was identified and excised. Direct laryngoscopy demonstrated multiple laryngeal and pharyngeal micro-cysts. The intraoral haemangiomatous components were left as such. The child was electively intubated for 7 days postoperatively and thereafter tracheostomized in view of need for prolonged ventilatory support and upper airway obstruction. Steroids and propanolol (1mg/kg each) were started for the haemangiomatous components. The lesion reduced in size significantly by the third week postoperatively but the child had difficulty in swallowing feeds. Hence a gastrostomy was performed two weeks later to take care of the child’s nutrition.

The child was discharged after a period of two months of hospital stay on tracheostomy and gastrostomy (Figure 2). At 3 months follow up, the swelling had decreased by seventy five percent with only facial swelling remaining; the child had gained weight adequately. However, the deglutination and swallowing were established later than usual due to the presence of the intra oral and laryngeal micro cysts. The baby started accepting oral feeds at last follow up.

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Case 2

A 3-month-old baby boy presented with a huge cervico-oral haemangio lymphangiomatosus swelling with intra oral sublingual extension that lifted the tongue. The soft swelling had begun as a 5x6 cm swelling over the right cheek and the submandibular region that had progressively increased in size to extend into the neck and oral cavity.

An early excision of the 15 x 10x 10cm multi-septated lesion was done. The lesion consisted of micro and macro cysts of varying sizes filled with haemorrhagic, serous and amber-coloured fluid. Though most of the lesion could be excised, the haemangiomatous component was left as such (Figure 3). Post operatively, the baby did not tolerate a trial of extubation and required a tracheostomy. Steroids and propanolol (1mg/kg) were started for the haemangiomatous components. Nasogastric feeding was started. Even after a week, oral feedings could not be resumed due to poor swallowing. A gastrostomy was done and the baby was discharged after 45 days of hospital stay.

Two weeks after discharge, the tracheostomy came out accidently. Multiple residual cystic spaces refilled. Sclerotherapy with sodium tetradecylsulphate was done twice at intervals of 3 weeks. The baby became better at 4.5 months follow up and had no respiratory distress. He could be weaned off the tracheostomy at 5 months follow up. However, the baby continued to take gastrostomy feeds till 6 months follow up. The gastrostomy was gradually clamped for increasing durations and finally taken out at 7 months follow up. The baby had gained weight adequately at 8 months follow up.

Discussion

Cervical cystic hygromas/lymphangiomas occur due to incomplete connection established between the normally placed lymphatic sacs. An early antenatal diagnosis of cystic lesion in the neck may suggest Turners syndrome with chromosomal abnormalities in 39% cases [2]. An institutional delivery is recommended...
for prenatally diagnosed cases. If the cystic hygroma is large, a cesarean section may be performed and may require an ex utero-intrapartum treatment (EXIT) procedure if the airway is potentially compromised [3]. After birth, infants with persistent giant cervical cystic hygroma must be monitored for such life-threatening airway obstruction that may require intubation, needle aspiration or early excision. Case 1 had a difficult intubation.

Cystic hygromas has a varied treatment ranging from observation and spontaneous regression in a few asymptomatic cases to surgical excision. Surgical excision is strongly recommended in cases causing significant cosmetic deformity, obstructive symptoms, bleeding or recurrent infections [4]. The other treatment modalities include aspiration, radiation and injection sclerotherapy using bleomycin, sodium tetradecylsulfate, hypertonic saline and OK 432 [5]. In the cases described, acute airway obstruction causing respiratory distress would have led to death if not managed by a combined approach of aspiration, sclerotherapy and surgical excision in time. As residual lesions are common, for achieving complete resolution of the lesions, a combination of tailored multiple surgeries and sclerotherapy have been adopted [6]. Acute airway obstruction is more common in cases with laryngeal involvement [7]. Large lesions are notorious to reoccur despite treatment but recurrences are less after excision [8].

The surgical excisions of such huge cystic hygromas are associated with high complications like recurrent laryngeal nerve injury accounting to fatality [9]. In a series of 8 operated cases, the lesion could not be completely removed in all; important structures were damaged in 5 and 4 cases died including 3 with recurrent laryngeal nerve injury [9]. 7 of the 8 cases had respiratory distress post operatively necessitating tracheostomy in 3 cases.

Also both the babies reported here had feeding problems due to poor swallowing reflex. The presence of polyhydramnios in the first baby during the antenatal period suggests this. This issue has been under reported in literature and needs to be addressed by a temporary gastrostomy to allow adequate weight gain.

Learning Points/Take Home Messages

- An Institutional delivery is recommended for antenatal large cervical cystic hygromas
- A difficult airway may be encountered secondary to mass effect by the swelling and associated pharyngeal, laryngeal and tracheal micro-cysts
- Cyst aspiration may be helpful temporarily
- Timely temporary tracheostomy and gastrostomy is helpful to tide-over the period of acute edema and swelling in early postoperative period
- Steroids and propanolol may prove beneficial for haemangioma-tous lesions
- Multimodal therapy improves outcome for these challenging cases

References


Figure 3: (a) Operative photograph of Case 2 showing lymphohaemangious lesion (b) post operative photograph after excision of lesion.