Abstract:
A 5 days old male child presented with antenatally detected huge Cystic Hygroma, with a characteristic heart shaped double headed appearance. Child presented with respiratory distress and was managed with early surgical excision successfully. Early surgery was performed because the Cystic Hygroma was compressing the structures in the face and neck, besides causing severe disfigurement. The paper is being presented for the interesting appearance of the child at birth and the successful surgical outcome. Residual disabilities were corrected with Bleomycin. These combination modalities resulted in an almost normal looking child.

Keyword: Cystic Hygroma, antenatal diagnosis, surgical treatment (fig:1)

Clinical History:
Five days old male child admitted with antenatally detected cystic lesion of face involving the entire right half of the face. At birth child had a heart shaped double headed appearance (fig:1), with the lesion stretching the eye and pushing the eye ball outside. The child had respiratory distress, tachypnoea with stridor and difficulty in sucking and taking feeds.

On general examination, the child was active with a good cry and hydration. No history of seizure disorder. Local examination revealed a soft cystic swelling over the face involving forehead, cheeks, maxillary and mandibular region, with extension into the neck. There was a bulge over the inner buccal mucosa and lateral oral cavity with thinning of the bones and stretching of the eye lids. The ear lobe on the whole was pushed back. It was a fluctuant and brilliantly trans-illuminant swelling (fig: 2)
without any pulsation.

The child was put on antibiotics, and supplemented with nasal oxygen and IV supplements. Ophthalmology and ENT opinions were obtained. The antenatal USG was suggestive of a giant cystic hygroma (fig: 3 & 4). The MRI revealed a huge cystic swelling involving right parietal, maxillary and mandibular region with slight extension into the neck. There was compression over the maxilla and the mandible with thinning of the bones and medial displacement with thinning of orbital wall and displacement of eyeball (fig: 5).

(fig:2)

There was no evidence of any intracranial extension or loculations or septations. It was associated with well defined wall (fig: 6) with mild displacement of the vessels (fig: 7). The other investigations were within normal limits and the child was taken up for surgery and a near total excision of the cyst was done through a curvilinear incision, leaving a part of the cyst wall adherent to the vital structures (fig: 9 & 10). The child was progressively improving in the post operative period except for minimal collections which were drained and supplemented with 2 courses of Bleomycin intralesional injection. The child improved significantly well (fig: 12) and is on regular follow up. The biopsy report was consistent with lymphangioma.

Discussion:
Lymphangiomas are localized, congenital malformations of the lymphatic system that involve the skin and subcutaneous tissues. In 1843, Wernher gave the first case report of a cystic Hygroma. This term comes from the Greek word “hygroth” meaning fluid and “oma” meaning tumor. Lymphangiomas are sequestration of lymphatics that fail to communicate with the vascular tree. Lymphangiomas are more common the left as the Thoracic duct joins the Brachio Cephalic Vein here. Spillage or sequestrations at these junctures leads to Cystic Hygroma. They are multi lobular and multi locular cystic lesions. The Cyst walls consist of single layer of flattened endothelium. 50% present at birth and up to 90% are evident by the end of second year of life. They are found most commonly in the head and neck, with the posterior cervical region the commonest site and also in the axilla and abdomen. Cervical facial Lymphangiomas are relatively uncommon, representing fewer than 6% of benign tumors of childhood. Incidence has been reported to be 1.2 to 2.8 per thousand populations. Most studies report an equal incidence among males and females.
Lymphangiomas are associated with Turner’s, Klinefelter’s and Noonan’s syndromes and exposure to alcohol during pregnancy. It is necessary to differentiate these lesions from other cervical congenital masses or malignancy. The cystic nature and the bluish hue frequently resemble characteristics of a Hemangioma. Lesions in the floor of the mouth, particularly when they bulge in the sub mental region, may simulate a plunging Ranula. Other differential considerations in the head and neck include a Branchial cleft cyst, a Thyro glossal duct cyst, and goiter. Investigations of choice are a Prenatal USG and MRI to rule out airway compression or obstruction. If present EXIT Procedures are considered. If diagnosed in 1st trimester with a Karyo typing abnormality there is a 59% chance for poor outcome. If diagnosed in 1st trimester without a Karyo typing abnormality there is a 89% chance for viability and 20% would go for a dysmorphic sequelae. If diagnosed in 2nd and 3rd trimester (i.e. <30 weeks) prognosis is guarded. If diagnosed in 3rd trimester (i.e. >30 weeks) —

The treatment modalities are surgical excision or partial excision and medical management — intra lesional bleomycin. Excision is the only effective available treatment because of the risks of spontaneous infection, progressive growth, airway compression and obstruction or dysphagia is relieved by surgical excision. Post operative complications include infection, Horner’s syndrome, recurrent laryngeal nerve palsy, damage to spinal accessory and marginal mandibular nerves. Histologically, these tumors are composed of dilated lymphatic channels. The large empty lymphatic channels that are lined by a flattened layer of endothelial cells, they contain proteinaceous fluid. The intervening stroma consists of fat, fibrous tissue and skeletal muscle. The dilated spaces can vary from microscopic to large and cystic. They vary in size depending on the location and surrounding tissues. Landing and Farber were the first to divide these lesions into two major groups based on the size of these abnormal lymphatic vessels. When a lymphangioma is confined to rather dense tissue like the tongue it presents as a micro cystic lesion, but generally when it develops in the relatively loose fascia of the neck a macro
OTHER MODES OF TREATMENT: CO₂ laser treatment for superficial lesions, radiation, steroids, sclerotherapy and intralesional injection of OK – 432, bleomycin A₅ and vincristine. Lymphangioma is not a fatal disease. There is a 3% mortality rate. These deaths are usually due to bronchospasm, atelectasis, or airway compromise from edema. There is no risk of malignant transformation. The growth rate is variable but most lesions tend to progress slowly. Recurrence rates following complete excision range from 0 to 27% in various studies, and after partial excision the recurrence rate is reported to be between 50 to 100%. Most recurrences occur within the first year of resection, but delayed recurrences as long as 10 years after the initial operation have been reported.

**Conclusion:**
Cystic Hygroma varies from a simple cyst to life threatening conditions like CHAOS (Congenital High Airway Obstruction). Treatment also follows suit from Observation, Injections to extirpation of the tumour. Here we have successfully, managed a grossly disfiguring tumour which was life threatening and also causing gross distortion of the anatomy employing all the modalities. Hence we advocate intervention when Compressive features, and complications like infection and bleeding supervene. The benign nature of the disease must be kept in mind and no unduly aggressive surgery entailing permanent loss must not be performed at the other extreme.

**References:**

(fig:9) (fig:10)