Massive infiltrating cystic hygroma of the neck in infancy

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Barrand, K. G., and Freeman, N. V. (1973). *Archives of Disease in Childhood, 48, 523.* Massively infiltrating cystic hygroma of the neck in infancy. Nine cases of massive infiltrating cystic hygroma of the neck are reviewed; it was in this group that major difficulties in treatment arose. Unexpectedly high morbidity and mortality were found after surgical removal. Conservative management of these tumours is therefore suggested.

**History**

Redenbacher, in 1828, first described cystic hygroma which he termed ‘ranula congenita’ and Wernher (1843) first used the term cystic hygroma. In 1872 Koester suggested that cystic hygromas were derived from lymphatic tissue. At the turn of the century, two parallel theories regarding the development of the lymphatic system arose.

Sabin (1909), following the work of Langer (1868) and Ranvier (1897a, b), developed the centrifugal theory of lymphatic development. She considered that the lymph sacs developed from veins, and that subsequent sprouting of these primary sacs developed into the lymphatic system. Sala (1899) showed that lymph vessels in the chick embryo are formed independently of the venous system, and Huntington and McClure (1907), working with cats, while agreeing that the jugular lymph sacs arose from veins, showed peripheral lymphatic vessels along side the veins before the formation of the lymph sacs. Huntington and McClure (1908) considered the lymphatic system as beginning in mesenchymal slits in the 9 to 12 mm embryo. These coalesced to form lymphatic cavities, which secondarily opened into the venous system. This view was supported by Kampmeier (1931) working with human embryos.

Two main theories regarding the aetiology of cystic hygroma thus similarly arose.

Goetsch (1938) considered that the hygroma began centrally and grew peripherally. He observed fibrillary sprouts arising from the lining wall of the cystic spaces which he thought led to enlargement of the tumour and penetration of the surrounding tissues. He supported the idea of Dowd (1913) that the tumour originated in lymphatic rests—remnants from the primitive jugular lymph sacs.

Godart (1966) supported the views of Huntington and McClure and described cystic hygroma as being due to a failure of the primary lymph spaces to join the central system. The tumour then grew because atresia of the main collecting vessels prevented its emptying. In *cavernous lymphangioma* the sequestration is more peripheral and in *lymphangioma simplex* it is the result of still more localized sequestration.

In 1960 Willis compared cystic hygromas with haemangiomas and believed that fluid accumulation, the formation of collateral channels, supraventricular on thrombosis and organization, and accounted for their growth.

**Definition**

Landing and Farber (1956) have classified lymphangiomas into 3 groups. (1) **Lymphangioma simplex** composed of capillary-sized thin-walled lymphatic channels. (2) **Cavernous lymphangioma** composed of dilated lymphatic channels. (3) **Cystic lymphangioma** (hygroma) composed of cysts from a few mm to several cm in diameter.

Bill and Sumner (1965) believe that there is no dividing line between the types. Where tissues are dense, e.g. tongue, there is more likely to be a lymphangioma. Where tissues are lax, e.g. neck, there is more likely to be a hygroma. In Bill and Sumner's cases the different varieties of lymphangioma frequently coexisted.
Case selection

Most authors employ the term 'cystic hygroma' to describe any tumour containing large or small cystic spaces lined by endothelium.

In selecting the group of 9 cases seen and treated at this hospital between 1958 and 1972, we have limited our discussion to the massive infiltrating hygromas of the neck presenting in infancy. Whether or not infiltration was present was judged at operation, and not by the size or situation of the tumour. During the same time, 763 cases of lymphangioma and haemangioma were diagnosed and treated at this hospital, thus giving some idea as to the relative frequency of the massive hygromas.

Signs and symptoms

In our cases, a cervical swelling was noticed at birth or soon after, except in Case 3 whose swelling was first noticed at the age of 11 months. Respiratory distress was noted in 5 cases (Table).

Case material

Case 1. A right cervical swelling was noticed at birth. This increased in size and caused difficulty in feeding. 30 days after birth a cyst was aspirated after which there was a rapid increase in size, and at 40 days operation was performed. A large multicystic tumour was discovered ramifying behind the trachea and around both carotid arteries. The right jugular vein was sacrificed. After operation a palsy of the cervical branch of the right facial nerve was noted and laryngoscopy revealed a right recurrent laryngeal nerve palsy. Histology of the excised mass showed a multiloculated lymphangioma with evidence of inflammation. The patient developed recurrent respiratory infection and was unable to feed orally. A Dionosil swallow was suggestive of pharyngeal incoordination.

She developed cyanotic attacks and had several attacks of pneumonia in the first year of life. Her vocal cord paralysis persisted. She was only rarely out of hospital and on 6 February 1961 she died suddenly. At necropsy aspirated food was found blocking the left main bronchus.

Case 2. Forceps delivery, weight 3·1 kg. A large cystic hygroma was noticed on the right side of his neck 10 days after birth. This was observed and it gradually increased in size. There was no respiratory distress and the child fed normally. At the age of 14 months the swelling increased further in size, x-ray of the chest showed extension of the swelling into the right upper lung field and the trachea was displaced forwards and to the left. Hb was 7·9 g/100 ml and this was corrected by blood transfusion. It was decided not to operate in the hope that spontaneous regression would occur, but at age 2 further increase in size precipitated surgical excision. Via a transverse cervical incision, a large multilocular cystic hygroma was excised with extreme difficulty. The lesion was closely related to the right internal jugular vein and extended to the floor of the mouth and behind the trachea. The intrathoracic extension could not be removed. A few hours after operation the child developed marked stridor with rib recession. On laryngoscopy both cords moved normally but there was subglottic oedema. This resolved after a course of hydrocortisone. The child was discharged 2 weeks after operation.

Histology showed a large mass of intercommunicating cystic spaces, and it was reported as a benign lymphangioma with inflammatory changes. He was reviewed regularly until the age of 3 and up to this age no recurrence was noted.

Case 3. Normal term delivery, weight 2·7 kg. The pregnancy was complicated by rubella infection at 2 months' gestation. A right cervical swelling was noticed at the age of 11 months, and he was first seen when he was 14 months old. The swelling was extensive, lying deep to the sternomastoid muscle. The hygroma increased in size and at the age of 16 months operation was carried out via a right crease cervical incision. A large cyst and multiple smaller cysts were removed. The hygroma extended around the right carotid sheath, and was associated with numerous lymph nodes. Excision was incomplete. Histology confirmed that it was a benign lymphangioma with multiple cysts. At 17 months of age there was a sudden reappearance of the swelling which increased to its former size, and therefore a second operation was performed. This involved a

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difficult dissection around the right carotid sheath. 12 hours after this second operation the child developed severe stridor with rib recession. Laryngoscopy showed marked laryngeal oedema and a cystic swelling above the larynx. A tracheostomy was performed. 2 weeks later a repeat laryngoscopy showed that the anatomy of the larynx had returned to normal. The tracheostomy tube was removed and the child made a good recovery. No further recurrence of the hygroma was noted up to the age of 10 years. He developed a cough with a whoop which persisted; no obvious cause was found and it was considered that the whoop might be due to his old tracheostomy.

Case 4. Admitted at 12 days, weight 3·0 kg. He was noted to have swellings in both submandibular regions of the neck extending into the tongue and the floor of the mouth. He had respiratory stridor and rib recession. The following day there was bleeding from the floor of the mouth. The cervical and tongue swellings increased in size rapidly and he had an apnoeic attack. 2 days after admission tracheostomy was performed. After this the cervical swellings continued to increase in size, and the child required feeding via a nasogastric tube. He developed a respiratory infection. A course of radiotherapy was given but this caused the tumour to increase further in size. At 30 days of age, the main cystic mass in the neck was removed via a transverse cervical incision. The histology showed benign lymphangioma with some haemangiomatosus formation, with haemorrhage into the interstitial tissues. After operation the remaining cysts increased in size and were aspirated. The tongue continued to increase in size. At age 6 weeks a horizontal wedge excision of the tongue was performed and after this he fed orally. At 10 weeks laryngoscopy showed anterior-posterior compression of the trachea. When 3 months old a further cervical recurrence was excised at operation, and after this his tracheostomy was removed. The child was discharged home at the age of 4 months with a small residual lymphangioma in the left orbit.

Thereafter he made satisfactory progress apart from occasional blood staining of saliva and recurrent monilial infection of the mouth and tongue. When aged 2, 20% saline was injected into the tongue and 6 months later a wedge excision of lymphangioma of the lower lip was performed. Prognathism had developed, but he could close his mouth satisfactorily. The family then moved and were lost to further follow-up.

Case 5. Normal term delivery, weight 4·2 kg. His mother had noticed rattling breathing from birth, and at the age of 2 weeks his respiration became gasping and bubbly. He was diagnosed as having laryngitis and was treated at home, but was admitted to hospital at age 3 weeks in gross respiratory distress requiring intubation. A fluctuant swelling 3 cm x 3 cm which, when trans-illuminated, presented behind the right sternomastoid muscle. The cervical swelling increased rapidly in size. At the age of 1 month a large multilocular cystic mass was excised from the right cervical region via a crease incision. The cystic mass was attached to the carotid sheath and extended above the mandible and down into the thorax. Total excision was not possible, and 3 days after this operation a tense right cervical swelling reappeared. The child developed bronchopneumonia with frequent cyanotic attacks and was reintubated. At age 7 weeks laryngoscopy showed a right abductor palsy and therefore a tracheostomy was performed. After this the child began to thrive and gain weight. At 10 weeks a left lower lobe pneumonia developed which responded to treatment. At 3 months the tracheostomy tube was removed, but 2 weeks later the child collapsed, became cyanosed, and was reintubated. 48 hours later the endotracheal tube was removed, but he continued to have apnoeic and spluttering attacks and died aged 3½ months. Necropsy revealed residual multicystic benign lymphangioma around the right carotid sheath passing between tracheal and pharyngeal walls and extending up to the nasopharynx. Bronchopneumonia was the immediate cause of death.

Case 6. Admitted aged 4 days, weight 3·5 kg. He had been difficult to resuscitate at birth and had to be tube fed. There was a large left cervical swelling which rapidly increased in size, occupying the whole left side of his neck. The swelling appeared multiloculated, and involved the floor of the mouth. The child was nursed prone on a Burston frame (Burston, 1969). At the age of 1 week he was given a 2-week course of radiotherapy, but the swelling did not prove to be radio sensitive; it increased in size and became indurated. When aged 24 days, the respiratory obstruction became worse with a markedly raised Pco2 and operation was performed. On laryngoscopy, the aryepiglottic folds were noticed to be oedematous and nodular with tumour, leaving a slit-like opening to the trachea. Via a collar incision, the cystic mass was dissected off the strap muscles, the sternomastoid muscles, both carotid sheaths, and both parotid glands. There was retrosternal prolongation and extension into the tongue; neither extensions could be removed. Tracheostomy and gastrostomy were carried out. During dissection the right internal jugular vein and the left facial nerve were injured. After operation the child developed a persistent respiratory infection. Because of haemorrhage into the remaining cysts, the swelling reappeared in the neck. Blood was aspirated from several cysts. The tracheostomy tube had to be changed frequently due to obstruction with debris. When aged 43 days he was found dead in his incubator. Necropsy showed haemorrhagic areas around the oesophagus and trachea. Clinically and at operation this was thought to be a haemangiolympangioma. Histology showed a benign, partly calcified angioma.

Case 7. Admitted when aged 2 days, weight 4·5 kg. He had a large fluctuant, brilliantly transilluminable swelling of both sides of the neck extending from mandible to the sternum and involving the floor of the mouth (Fig. 1a).

He had early signs of respiratory distress which were
Case 7. Weight 4.5 kg. Admitted a few hours after
birth with a very large multicystic swelling of the left side
of her neck. The trachea was deviated to the right and
the head was held at right angles to the body (Fig. 2).
There was no respiratory distress, but x-ray of the neck
showed the trachea to be pushed markedly forward (Fig. 3).
When aged 3 days, the main mass of tumour was
excised via a left crease incision. The larynx, though
deviated, was normal on intubation. The tumour
ramified among vital structures at the base of the skull,
around the trachea, and down to the cervical spine. It
was composed of multiple small cysts and could not be
completely excised. Fluid taken from one of these
showed a protein content of 750 mg/100 ml. Histology
confirmed a benign cystic lymphangioma. After
operation plasma-like fluid drained from the wound and
the wound became infected. The residual cysts
increased in size and on aspiration were found to contain
blood. She developed cyanotic attacks during feeding
and was therefore tube fed. After operation, left facial
tissue and left recurrent laryngeal nerve palsies were
noted. A Dionosil swallow showed spillage into the
trachea and very little propulsion down the oesophagus.
She developed respiratory distress and when aged 25
days tracheostomy and gastrostomy were performed.
Due to dehiscence, the abdominal wound required

Fig. 1.—Case 7. (a) At age 2 days; (b) at age 3 months.
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FIG. 2.—Case 8 at age 24 hours.

Resuturing. At age 7 weeks the child began oral feeding and the gastrostomy was allowed to close; however, her respiratory problems persisted and weight gain was slow. The left facial nerve palsy began to improve. When she was 2 months old she collapsed and died. Necropsy showed a widely infiltrating cystic mass extending from the clavicle to the floor of the mouth. Death was thought to be due to a plug of mucus in the tracheostomy.

Case 9. She was noticed to have a haemangioma of the lower lip and cystic swellings on both sides of the neck (Fig. 4).

When 1 week old she was admitted with bubbly respiration and apnoeic attacks. The bilateral cervical swellings were soft and extended to the parotid regions. X-ray of the neck showed forward deviation of the trachea. Laryngoscopy showed cysts at the base of the tongue. The swellings in the neck increased in size and the child developed respiratory difficulty, especially after oral feeding. A Dionosil swallow showed normal passage of the contrast medium into the stomach. Respiratory difficulty continued, and when aged 22 days, a tracheostomy and gastrostomy were performed. The cystic mass overlying the trachea was excised during tracheostomy and showed the histological features of benign lymphangioma. The protein content of the fluid aspirated from the cysts varied between 3·9 and 5·4 g/100 ml with a similar electrolyte pattern to normal plasma. After operation the gastrostomy leaked and had to be abandoned, the child being fed by nasogastric tube. When 32 days old, several cysts around the glottis were diathermied. The child was slow to thrive and developed respiratory infection. The cervical mass increased in size with hard nodules within the hygroma, which on aspiration proved to be haemorrhage into the cysts. A course of sodium tetra-decyl injections was given into the cysts. These temporarily caused an increase in the size of each cyst injected; there was no dramatic sclerosant effect and there was no long-term decrease in size. At age 5 months the child was gaining weight and feeding orally, but still required the tracheostomy. The cystic mass seemed to be growing in proportion to the rest of the body. When 9 months old the tracheostomy was removed, the child breathed and fed normally, and she was allowed home.

Treatment

In our series of 9 cases the indications for treatment were respiratory distress 5, dysphagia 2, and
sudden increase in size of the tumour. In most of the series reported the main indication was the presence of a mass. Thus, it is difficult to compare this group with the results from other centres. Previous authors have not distinguished the massive infiltrating lesions from the smaller superficial lymphangiomas, which present no surgical difficulties in removal and are not associated with any morbidity or mortality.

In Goetsch's (1938) cases the youngest was 6 months old; the baby had a massive cystic hygroma and was the only patient that died.

The types of treatment which have been employed are as follows. Aspiration—when used alone this was ineffective on two occasions in Case 1 and on one occasion in Case 4. Aspiration + injection of sclerosant, 20% saline produced an unpleasant cellulitis when injected into the tongue in Case 4. There was no effect in Case 9. Radiotherapy was used in Cases 4 and 6 causing increase in size in both sufficient to precipitate urgent surgery. Surgical excision was carried out in 8 cases. In none could the tumour be completely removed, and in 5 important structures were damaged. 4 cases out of the 8 died. All 3 cases in which a recurrent laryngeal nerve was divided died. 7 of the 8 cases had respiratory distress after removal of the main cervical mass, and in 3 cases tracheostomy was later performed.

There is no agreement on the age at which operation should be undertaken. Ravitch (1962) stated that operation should be avoided in premature babies, Broomhead (1964) that 18 to 24 months is the most suitable age, Gross (1953) and Ward, Harris, and Downey (1970) that early removal is advisable to prevent further increase in size.

**Discussion**

In planning treatment, several points of interest arise. Spontaneous disappearance has been reported in 7 out of 44 cases by Broomhead (1964), but other authors such as Bill and Sumner (1965), Galofre *et al.* (1962), and Gross (1953) failed to find this. A single aspiration and injection of a hygroma have both been reported as leading to complete cure (Broomhead, 1964).

It is commonly agreed that about half of all of cystic hygromas are present at birth and that the remainder first appear in the early years of life. In Galofre *et al.*'s (1962) series of 141 cases, 32% presented at birth.
We believe that in massive infiltrating cystic hygroma of the neck it should be accepted that the lesion cannot be completely excised. There is a high risk of operating in the first few months of life and surgery should be confined to urgent measures such as tracheostomy and gastrostomy. When the child is thriving, provided there is no evidence of regression and preferably after the age of 1 year, excision of the tumour should be performed.

In Case 9 we were interested to know if the cysts intercommunicated, as has been suggested (Lynn, 1963). If intercommunication does exist, then sclerotherapy could be expected to be successful for the whole tumour. We thought in terms of draining a group of cysts via a cystopleural shunt using a Spitz-Holter valve, but this would only be effective if there were free intercommunications. We therefore injected 'ultra-fluid Lipiodol' into a single cyst after aspiration and followed progress with serial x-rays. After 3 weeks the Lipiodol was still well localized in the cyst, indicating minimal intercommunication (Fig. 5).

At the age of 4 months, 10 μCi radio-iodinated serum albumin (RISA) was injected into the lymphangioma and serial estimations of activity were performed over the next 22 days by Dr. R. Spencer, radiotherapist. There was a slow but definite diminution of activity compared with the expected reduction due to decay. This loss of RISA was presumably into the blood capillary system (Fig. 6).

Shortly afterwards, lymphangiography was performed via the foot and via the left side of the neck. The lymphangiogram showed normal cervical lymphatics and a normal though displaced chain of left cervical glands. The thoracic duct was seen during injection on one film and appeared normal. This supports the idea that the lymph-

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**Fig. 5.—Case 9. X-ray of neck showing contrast medium (a) immediately after injection into a cyst; (b) after 3 weeks.
angioma formation was peripheral rather than being derived from the jugular lymph sac, which is known to form the proximal thoracic duct (Huntington and McClure, 1908).

We were interested in the accumulation and drainage of fluid in the hygromas. According to Godart (1966), the volume of tumour usually increases after inflammation because the fluid balance between arteries and veins is disturbed and the filtration and lymph formation are consequently increased. The accumulated fluid separates bundles of muscle fibres, vessels, and nerves which are soon surrounded by fluid and which then degenerate under fluid pressure. Vaughn (1934) injected iodized poppy seed oil into a massive cystic hygroma after aspiration of its contents. X-rays taken 30 minutes later revealed complete disappearance. This was not our experience with 'ultra-fluid Lipiodol' in Case 9, in which intercommunication and drainage were virtually nil. 2 ml ultra-fluid Lipiodol injected into a single cyst was still present 3 weeks later (Fig. 6).

The microvascular circulation of lymphangiomas was studied in 5 patients aged 2 to 5 months by Touloukian et al. (1971) using $^{135}$Ze in saline. 100 $\mu$Ci were injected in 0.1 ml saline into the centre of the lesion and counts were carried out for 90 minutes. This study in smaller and peripheral lymphangiomas showed the existence of a lymphatico-venous circulation which is comparable to normal values for blood circulation within the skin. An inverse correlation of rate of disappearance to the gross size of the lymphangioma was shown, suggesting that a functional lymphatico-venous block contributes to size of lymphangiomas.

Using RISA in the massive multicystic lesion of Case 9, very slow clearance was obtained over 22 days. This indicates that in the massive lesion the lymphatico-venous block is almost complete.

Conclusion

The mortality of massive infiltrating cystic hygroma of the neck presenting in infancy is high (4 out of 9 cases). Surgical excision is never complete and the morbidity is high. Unless respiratory distress or dysphagia necessitates urgent operation, the treatment should be expectant. Tracheostomy without excision of the hygroma is preferable initially.

Cases 1 to 5 were under the care of Miss I. Forshall, Mr. P. P. Rickham, and Mr. J. H. Johnston.

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