CONGENITAL CYST OF THE LARYNX CAUSING FATAL ASPHYXIA NEONATORUM

BY

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Congenital cysts in the region of the larynx of sufficient size to cause embarrassment of respiration at birth are rare, and the following case possesses several points of interest.

Case report

Baby S., the first child of young and perfectly healthy parents, was born after a normal pregnancy and confinement. A doctor was present at the delivery, and the child had apparently been quite all right immediately after birth. With the first respiration, however, it became cyanosed and made repeated attempts at a gasping type of respiration, finally passing into a state resembling asphyxia pallida. All possible immediate treatment was adopted; no mucus could be removed, and oxygen was obtained with little delay and administered by a nasal catheter. With continuous oxygen administration the condition of the child improved, but it remained limp, with frequent attacks of cyanosis and occasional convulsive gasps. As soon as the oxygen was diminished the child became visibly worse, and after about an hour and a half the doctor sent for my assistance, mentioning the symptoms, so that I took with me a further supply of oxygen and a small catheter for endotracheal use. I found the child in the state described above, and after ensuring that no mucus was obstructing the air passages, the possibility of a congenital abnormality was considered. It was unlikely that a tracheo-oesophageal fusion existed, as some air was evidently getting into the lungs, and the extent of the respiratory effort suggested that there was no gross failure of expansion of the lungs.

The back of the pharynx was inspected directly and digital palpation of the superior aperture of the larynx was achieved, but no abnormalities were found. We were successful in passing a nasal catheter even into the trachea, and it was concluded that the case must be one of cerebral trauma. Arrangements for a nurse to tend the child were made, and it was suggested that if no improvement took place in a few hours the child should be transferred to the hospital for better supervision. Two hours later, as it was no better, the child was sent into the North Middlesex Hospital, but became worse in transit, with increased cyanosis, and on admission it was moribund and died before I was able to see it again.
Post-mortem examination revealed the cause of the trouble: a thin-walled cyst, containing fluid and about 2.5 cm. in diameter, lay over the superior aperture of the larynx, acting as a ball-valve.

It arose from the mucous membrane of the left recessus piriformis immediately below the aryepiglottic fold. At birth it probably lay in the recess, but its attachment to the mucosa was sufficiently loose to allow of its being drawn over the superior aperture of the larynx, probably with the first breath of the infant, effectively blocking air entry. Being thin-walled and not unduly tense, it had escaped the examining finger, and allowed the entrance of a catheter beside it; even the entrance of oxygen into the pharynx and larynx would diminish the tendency for violent inspiratory effort and make the valve action less pronounced. As soon as oxygen was cut off the next deep breath sucked it in again. Death was probably due ultimately to exhaustion, as a degree of trauma had been unavoidable in the several examinations of the infant.

The accompanying photograph illustrates the condition. The specimen was removed to the museum of the Royal College of Surgeons to be fixed before the wall was sectioned. The war has intervened and interrupted the investigation of its structure, but its external appearances, with a thin wall, leave little doubt that the lining consists of a thin layer of flattened cells. It was apparent that it was confined to the mucous membrane of the outer part of the larynx and was not connected with the deep tissues, so that a branchial cleft origin is practically ruled out, and it is likely that the condition is one of simple retention cyst of a mucous gland. There is no possibility of an aerocele as a basis.

No other cysts or other abnormalities of the larynx or elsewhere were found. The lungs showed partial expansion, with a few areas of atelectasis.
Discussion

The rarity of congenital cysts of the larynx is shown by the fact that Jacobi and Roscoff (1935) in a thorough search of the literature were only able to collect eleven cases, to which they added two of their own. They mention several possible sources for such cysts: the commonest is probably closure of a mucous gland, but this is not likely to occur until after birth, though it is possible that during intra-uterine swallowing of liquor a fragment of debris may block a gland. Other possibilities are inclusions of epithelial cells, dermoids and distension of blood and lymph channels. Park and Israel (1925) suggest that birth trauma may be a factor. Tow (1937) mentions the work of Jacobi and Roscoff and also refers to cases reported by Lorie and Lux, and by Chamberlain. In the series described by Jacobi and Roscoff eleven were noted at birth, one on the second day and one in the third week. Asphyxial signs were present in all, though not always at birth, and four cases recovered either following operation or after spontaneous rupture of the cyst. In most cases the cysts were multiple, and were visible externally as bulges in the neck. The cysts varied in position, the epiglottis, right laryngo-epiglottic fold, the laryngeal wall and the deep tissues of the neck being mentioned as sites. The cause of the respiratory embarrassment was usually direct pressure on the larynx by the cystic mass, and in no case was a ball-valve effect described.

In only one case was the cyst in the site of the one described here, and in that one case the cysts were multiple. Diagnosis was made in most cases by the presence of a swelling in the neck, with in several cases direct laryngoscopic confirmation of the condition.

In the present case there was little opportunity for laryngoscopy and the child died before operative interference could be attempted. If the cyst had been palpated, it is possible that puncture might have been attempted. The cures reported have been the result of somewhat extensive operative interference in infants who survived the immediate respiratory embarrassment, or in whom the symptoms did not become severe until some months after birth; in one case a partial laryngectomy was carried out after preliminary tracheotomy in a five months old child, and in another case the cyst was perforated and did not recur.

Summary

A case is described of congenital laryngeal cyst causing asphyxia neonatorum. Diagnosis was not achieved during life.

It is shown that such cases are rare, and that, in fact, no similar valve effect of a single cyst appears to have been described.

Though rare, it is worth considering in a case of puzzling neonatal asphyxia whether a laryngeal cyst might be present, as, if diagnosed, a cure is possible.

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References