Case Report

Pharyngeal Angiolipoma Presenting as Neonatal Respiratory Distress

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Abstract :

Neonatal respiratory distress is an acute emergency. We report a rare case of pharyngeal angiolipoma producing glottic obstruction in a neonate, which was successfully managed avoiding tracheostomy. The baby was found to have Tetralogy of Fallot, Brachycephaly, a sacral sinus and Syringomyelia. Angiolipomata of the head and neck are extremely rare, with only 36 cases previously reported. No previous association of angiolipoma with Fallot's tetralogy, Brachycephaly or Syringomyelia were found after a search of the literature.

Keywords :

Angiolipoma, Tetralogy of Fallot, Brachycephaly, Syringomyelia

Case Report :

A female neonate presented 3 hours after birth with respiratory distress and cyanosis. Born full term as a normal delivery, the first child of her parents, she did not cry at birth, and then developed stridor, so was referred from a rural hospital. On examination a smooth, mucosa covered mass was seen in the pharynx, obstructing the glottic inlet. A 2.5mm ID endotracheal tube was negotiated beyond the mass, which was firm and did not bleed to touch, to secure the airway.

The initial plan was to perform a tracheostomy and then proceed for definitive surgery. It was decided, however, to try and avoid tracheostomy if possible. A CT scan of the neck was obtained, which revealed the mass to be pedunculated, arising from the postero-lateral wall of the pharynx (Fig 1). The presence of a sacral sinus was noted, but lumbo-sacral skiagrams excluded spina bifida. The shape of the skull did not appear normal. Surgery was performed on the third day post-delivery.

On table the endotracheal tube was carefully

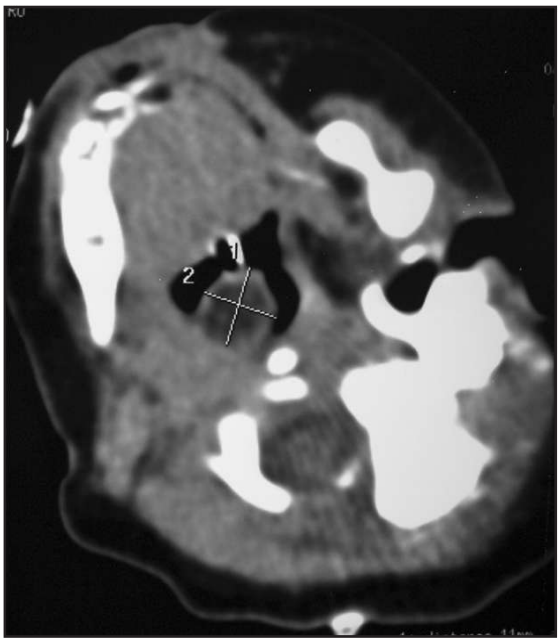

Fig 1 : Axial CT scan showing pedunculated, postero-lateral pharyngeal mass

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replaced by a 3.5mm armoured tube and a right Lateral Pharyngotomy performed. Haemostasis was maintained throughout by judicious use of bipolar diathermy. A smooth, mucosa covered, firm, pyramidal, pedunculated mass was delivered via the pharyngeal incision (Fig 2); the pedicle was transfixed with 4-0 silk, and the mass excised. The pharynx was closed with interrupted 5-0 vicryl. A braided nylon drain was introduced, and the incision closed in layers. The armoured tube was carefully replaced with a 3.5mm portex endotracheal tube, and a nasogastric tube was introduced.

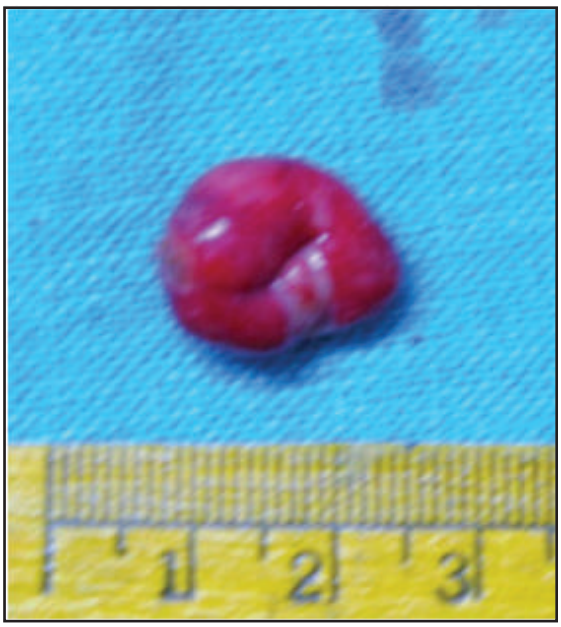

Fig 2: Excised mass

Apart from one convulsive episode the post-operative period was otherwise uneventful. She was weaned off the ventilator and extubated without problem on the third post-operative day.

The histopathology report was angiolipoma.

After extubation some respiratory distress was still apparent when the baby cried. Further investigations revealed the presence of Tetralogy of Fallot. Brachycephaly was apparent within a few months, and a lumbo-sacral MRI scan found the sacral sinus to end at the vertebral column, with a D11 – S1 syrinx. Appropriate management was organised for all of these conditions. The patient is well on five years of follow up.

Discussion :

Neonatal respiratory distress is an acute emergency. In the premature baby the commonest cause is hyaline membrane disease. In term deliveries the commonest cause of congenital stridor is laryngomalacia, followed by congenital laryngeal web, vocal cord palsy, subglottic stenosis, subglottioc haemangioma, laryngeal or laryngotracheo-oesophageal cleft, tracheomalacia and tracheal stenosis[1]. Endotracheal intubation and ventilation of premature infants was responsible for a marked increase in the number of cases of aquired subglottic stenosis, but the development of high volume, low pressure cuffs and improved ventilatory techniques has seen the incidence drop.

Angiolipomata are benign, encapsulated variants of lipomata; histologically they resemble lipomata in the presence of mature lipocytes with areas of angiomatous elements[2]. They have been described in the gastro-intestinal tract[3], the spine[4], the bones[5], as well as subcutaneously[6] and tend to be multiple[7]. To date there have been 36 documented cases of head and neck angiolipoma in the literature[8], with presentation in the supraglottic larynx[9], the parotid gland[10], the temporalis muscle[11] and the maxillary sinus[12]. Treatment is by surgical excision.

Numerous lesions are known to be associated with Tetralogy of Fallot, but angiolipoma has not been previously described as one. A Pubmed

search of the English language medical literature using the terms Angiolipoma AND Brachycephaly, Angiolipoma AND Syringomyelia yielded no results.

A rare case of neonatal pharyngeal angiolipoma is presented which was successfully managed without a tracheostomy. The lesion was associated with Tetralogy of Fallot, Brachycephaly and Syringomyelia.

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