Clinician's Corner

Whistling an unfamiliar tune

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CASE PRESENTATION

A female term infant, born uneventfully after a normal pregnancy, presented with fever at 24 h of life. Initial workup revealed a leukocytosis with elevated C-reactive protein. Empirical antimicrobial therapy was immediately initiated. On the third day, it presented with recurrence of a fever (39.2°C) associated with stridor and a few oxygen desaturation events. On examination, the infant exhibited no significant features that may have explained respiratory distress (e.g., hemangiomas, neck mass, signs of choanal atresia). The baby was transferred to the neonatal intensive care unit, where a full septic workup was undertaken, and antimicrobial therapy broadened. Progressive upper airway obstruction leading to hypercapnia required non-invasive ventilation with conventional nasal cannulation (0.5 L/min). Echocardiography was normal and showed no vascular rings. Fibre-optic laryngoscopy showed neither laryngomalacia nor vocal fold paralysis, but demonstrated an important left pharyngeal mass. Neck ultrasound followed by magnetic resonance imaging (MRI) promptly led to the diagnosis.

Figure 1. Female term infant presenting with fever and stridor. Neck magnetic resonance imaging demonstrates a mass further established as an infected fourth branchial pouch anomaly.

DISCUSSION

Both neck ultrasound and MRI (Figure 1) helped to delineate a $2.2 \times 2.4 \times 4.2$ cm³ mass with an opening facing the apex of the left pyriform sinus and displacing the trachea. Based on the location of that mass, the presence of an orifice, and its septic presentation, a diagnosis of an infected fourth branchial pouch anomaly was established.

The branchial arches develop between the fourth and seventh weeks of gestation and ultimately form the facial, cervical and pharyngeal structures. The fetal branchial apparatus is composed of six paired pharyngeal arches. Anomalies related to the branchial apparatus result from an incomplete obliteration of the apparatus during embryogenesis and usually present themselves as sinuses, fistulae or cysts in childhood or adulthood. While the epidemiology of branchial anomalies has not been fully ascertained, they probably represent approximately 20% of paediatric congenital head and neck masses (1). Of these, second branchial arch anomalies account for >90% of cases, followed by first branchial anomalies (8–10%), third branchial anomalies (2–8%) and finally, fourth arch anomalies, whose reported prevalence is very low (1–4%). The latter anomalies typically occur on the left side (>95%), are usually found later during childhood (mean age of presentation: 9 years), and are most commonly present along with acute suppurative thyroiditis or neck abscesses (2). Rare neonatal cases have been reported, with most of them presenting themselves with respiratory distress.

Barium swallow studies have been described as among the most used diagnostic tools for the investigation of branchial arch anomalies. While highly specific for the latter conditions, their sensitivity ranges only from 50% to 80% (3) as, during infectious processes, inflammation may lead to sinus obstruction, which prevents passage of the contrast material. Ultrasound imaging is a good tool to explore soft neck tissue and to identify a collection but usually requires further imaging studies. According to the current consensus, computed tomography (CT) is the proper choice for investigating the fourth branchial anomaly, and for detecting air density in the sinus tract as well as thyroid involvement. CT can also help to differentiate branchial arch anomalies from other cystic lesions of the neck. In the present case, MRI was preferred since it is readily available and avoids exposing a very young patient to radiation.

Surgery is the treatment of choice for branchial anomalies, for which numerous options can be considered. Incision and drainage are frequently performed, but they are associated with a high recurrence rate. Endoscopic obliteration of the orifice in the pyriform fossa allows a lower rate of recurrence. In the present case, an initial treatment was performed by incision with

imaging-guided drainage. One week later, endoscopic cauterization of the left pyriform fossa orifice was carried out. The post-operative course was uneventful, stridor quickly resolved and normal nocturnal oximetry was confirmed before discharge. At 10 months, the patient was totally asymptomatic, growing and developing well.

Manifestations of upper airway obstruction usually result from a variety of much more frequent conditions ranging from benign to life-threatening. Laryngomalacia is the most common cause of stridor in neonates and can be diagnosed with fibre-optic ('flexible') laryngoscopy. Laryngomalacia-associated stridor is not always present at birth but generally develops within the first week of life and disappears completely within the first 24 months of life. Such stridor varies with activity and is more noticeable upon agitation or crying, during upper respiratory infection episodes, and in the supine position. The differential diagnosis of infected fourth branchial pouch anomaly in neonates includes nasopharyngeal obstructive disorder, vocal fold paralysis, laryngeal cyst or web and subglottic lesion (including hemangiomas, tracheal stenosis or vascular rings). Other causes such as upper airway infections (laryngitis, epiglottitis, tracheitis), neurologic anomalies, trauma (secondary to inhalation injury or caustic ingestion), electrolyte anomalies (hypocalcemia), angioedema resulting from allergic reactions, while tumors are usually found in older subjects.

CLINICAL PEARLS

- Neonatal stridor is a manifestation of airway obstruction and should always be promptly evaluated by clinicians.
- Branchial anomalies, albeit rare conditions, should be considered in the differential diagnosis of neonatal upper airway obstruction.
- Surgery following proper antibiotherapy stays the treatment of choice for infected branchial arch anomalies.

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