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Congenital Midline Nodules on the Chin and Sternum

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A 5-day old black male full-term neonate was born via vacuum-assisted delivery for non-reassuring fetal heart rate. Two asymptomatic midline lesions on the chin and sternum were appreciated on newborn screening examination. There was no history of seizures, ophthalmologic findings, or abnormalities in head circumference, height, weight or limb size. Skin biopsies were performed from both lesions for the remaining RMH congenital midline lesions of the head, neck and chest include a broad spectrum of congenital anomalies, with RMH being one of the commonest. Other congenital anomalies include dermoid cysts, thyroglossal duct cysts, bronchogenic cysts and diaphragmatic hernia. RMH is described as a broad spectrum of congenital anomalies, with RMH being one of the commonest. Other congenital anomalies include dermoid cysts, thyroglossal duct cysts, bronchogenic cysts and diaphragmatic hernia. RMH is described as a combination of skeletal muscle, adipose tissue, and adnexal elements, and is often associated with syndromes such as PHACE, LUMBAR or infantile hemangiomas.

Diagnosis

Two asymptomatic midline lesions on the chin and sternum were appreciated on newborn screening examination. A total of 63 cases of RMH have been reported in the literature to date, of which 23% (n = 11) were associated with congenital anomalies. In a report of 47 patients with RMH by Mazza et al., 70% (n = 33) were either congenital or reported within the first year of life, and of those congenital RMHs, 23% (n = 11) were associated with congenital anomalies. A wide variety of congenital anomalies have been reported with RMH and include amniotic band syndrome, cleft lip and/or palate, auricular anomalies, dermoid cyst, thyroglossal duct sinus or cyst, spinal dysraphism, infantile hemangiomas, LUMBAR or PHACE syndromes. The mainstay of treatment for RMH is surgical excision, although there have been reports of spontaneous regression. A more conservative approach of watchful waiting may be considered, as there have been reports of spontaneous regression. Reassurance should be provided as there has been no documentation of malignant transformation of RMH.

HISTORY

- A 5-day old black male full-term neonate was born via vacuum-assisted delivery for non-reassuring fetal heart rate
- Two asymptomatic midline lesions on the chin and sternum were appreciated
- No history of seizures, ophthalmologic findings, or abnormalities in head circumference, height, weight or limb size
- Newborn screening examination was unremarkable

EXAMINATION

- The submental chin had a soft, erythematous dome-shaped nodule measuring 0.8-centimeters with a circumferential ring of brown pigmentation
- The upper sternum had a light brown 2-millimeter dome-shaped nodule

DIFFERENTIAL DIAGNOSIS

- Midline congenital lesions of the head, neck and chest include a broad spectrum of congenital anomalies
- Thyroglossal cysts typically present on the midline neck and have a potential to communicate with the base of the tongue or pharynx, causing movement with swallowing
- Bronchogenic cysts present as congenital nodules or pits over the supraesternal notch
- Cartilaginous rests of the neck, also known as wattle, may present midline as a skin colored papule
- Dermoid cysts, most often located on the orbital ridge, comprise about 25% of midline neck lesions

IMAGING

- Ultrasound of the submental chin revealed a heterogeneously hypoechoic structure with a peripheral soft tissue rind

HISTOLOGY

- Skin biopsies were performed from both lesions
- There were haphazardly-arranged elongate cells in fascicles throughout the dermis
- The cells contained deeply eosinophilic cytoplasm with regular striations
- Some of the fascicles were seen inserting directly on to the epidermal rete pegs
- The proliferation was highlighted red by a Masson’s trichrome preparation and demonstrated nuclear positivity with a myogenin immunohistochemical stain
- An Alcian blue stain revealed increased dermal mucin

Figure 1. Submental chin with a dome-shaped nodule and upper sternum with a small nodule.

Figure 2. Low power. Figure 3. Medium power, and Figure 4. High power histology of haphazardly-arranged elongate cells in fascicles throughout the dermis, inserting directly into the epidermal rete pegs. Figure 5. Nuclear positivity with myogenin IHC. Figure 6. Masson’s trichrome preparation highlighting the fascicles of cells red.

REFERENCES