5-2020

Congenital Midline Nodules on the Chin and Sternum

Allison Zarbo
Henry Ford Health System, azarbo2@hfhs.org

Kevin M. Luk
Henry Ford Health System, kluk1@hfhs.org

Tor Shwayder
Henry Ford Health System, tshwayd1@hfhs.org

Ben J. Friedman
Henry Ford Health System, bfriedm1@hfhs.org

Congenital Midline Nodules on the Chin and Sternum

Allison Zarbo, MD,[1] Kevin M. Luk, MD, MPH, [1] Tor A. Shwayder, MD,[1,2] Ben J. Friedman, MD[1,3]

[1]Department of Dermatology, [2]Division of Pediatric Dermatology, [3]Department of Pathology and Laboratory Medicine Henry Ford Health System, Detroit, Michigan

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 differential diagnosis
- 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 suprasternal notch
- Cartilaginous rests of the neck, also known as wattles, 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

CLINICAL IMAGE

Figure 1

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

PATHOLOGY

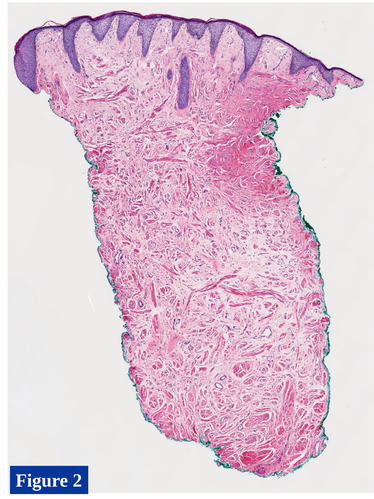

Figure 2

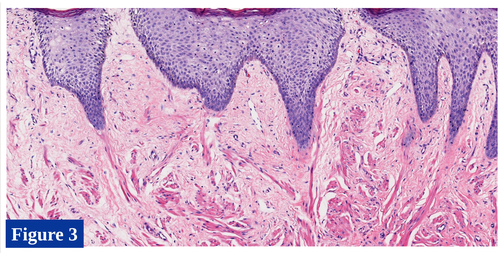

Figure 3

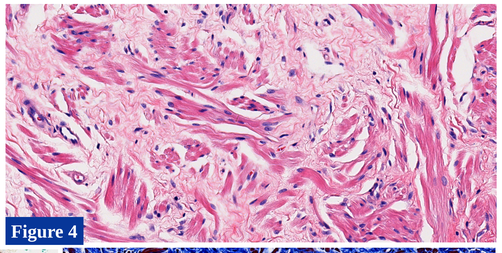

Figure 4

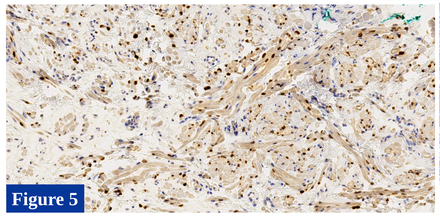

Figure 5

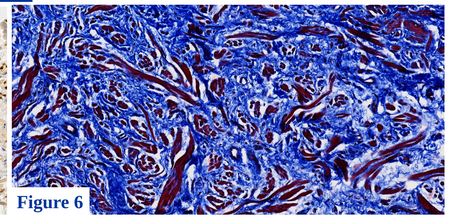

Figure 6

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.

DIAGNOSIS

- Rhabdomyomatous mesenchymal hamartoma

TREATMENT

- Punch biopsy of the rhabdomyomatous mesenchymal hamartoma (RMH) on the upper sternum was both diagnostic and excisional
- Conservative management with clinical monitoring for the remaining RMH on the submental chin was preferred by the guardians

DISCUSSION

- Rhabdomyomatous mesenchymal hamartoma (RMH), originally termed striated muscle hamartoma in 1986, is a rare and benign tumor composed of skeletal muscle, adipose tissue, and adnexal elements
- Congenital RMHs are most often midline and located on the head and neck
- Clinical presentation varies and includes plaques, papules with or without pedunculation, or subcutaneous nodules
- A total of 63 cases of RMH have been reported in the literature to date
- 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
- Of these 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, ocular 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 are reports of recurrence
- 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

REFERENCES

1. Mazza JM, Linnell E, Votava HJ, Wisoff JH, Silverberg NB. Biopsy-proven spontaneous regression of a rhabdomyomatous mesenchymal hamartoma. *Pediatr Dermatol.* 2015;32(2):256-262.
2. Stefanko NS, Davies OMT, Beato MJ, et al. Hamartomas and midline anomalies in association with infantile hemangiomas, PHACE, and LUMBAR syndromes. *Pediatr Dermatol.* 2020;37(1):78-85.
3. Orozco-Covarrubias L, Carrasco-Daza D, Diaz-Noriega A, Lara-Mendoza L, Ruiz-Maldonado R. Rhabdomyomatous Mesenchymal Hamartoma: A Deep Subcutaneous Lesion in the Sternoclavicular Area. *Pediatr Dermatol.* 2016;33(1):e36-37