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CASE REPORT

Rhabdomyomatous Mesenchymal Hamartoma Presenting as a Skin Tag in the Sternoclavicular Area

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Rhabdomyomatous mesenchymal hamartoma (RMH) is a rare congenital lesion of the dermis and subdermis. It has been described predominantly in newborns, with 30 cases reported in the English literature. Typically, it appears as a skin tag, papule, nodule or a mass involving the face or sternal notch. A 28-day-old girl presented with a 1.4 x 0.8 cm soft skin tag in the right sternoclavicular area. Physical examination revealed no congenital anomalies. A shaved biopsy showed that the core of the lesion contained striated muscle fibers mixed with hair follicles and sebaceous and eccrine glands. Thin epidermis lined the outside of the tag. We report a patient with a RMH in a site not previously reported and discuss the differential diagnosis. (Pathology Oncology Research Vol 13, No 4, 375–378)

Key words: rhabdomyomatous mesenchymal hamartoma, striated muscle hamartoma

Introduction

Rhabdomyomatous mesenchymal hamartoma (RMH) is a benign lesion of the skin, first described in 1986 as a striated muscle hamartoma.[1] To our knowledge, there are only 30 patients reported in the literature, eight with associated congenital anomalies. We report a patient with a RMH in the sternoclavicular area and review the English literature.

Case report

A 28-day-old girl was seen at Cardinal Glennon Children's Medical Center because of a congenital 1.4 x 0.8 cm soft skin tag in the right sternoclavicular area. Physical examination revealed no congenital anomalies. She was the product of a normal term pregnancy and there was no family history of similar skin lesions. Microscopically, the tag was lined by a thin epidermis (*Fig. 1*), the papillary and reticular dermis had increased hair follicles and sebaceous glands intermixed with skeletal muscle fibers (*Figs. 2 and 3*). A cluster of serous glands intimately admixed with the skeletal muscle fibers and adipose tissue was present at the base of the lesion. After 10 years of excision, the patient is well and without evidence of recurrence.

Discussion

RMH is a rare dermal lesion with 30 patients reported in the English literature (*Table 1*). This benign hamartoma was first described in 1986 as a "striated muscle hamartoma".[1] It has been reported under various names including striated muscle hamartoma, congenital midline hamartoma, and hamartoma of cutaneous adnexa and mesenchyme.[3] The term RMH given by Mills[4] has been the subject of criticism, since it emphasizes only the skeletal muscle component of the lesion, however, other mesodermal elements such as fat, erector pilli muscles and ectodermal components such as eccrine glands and elements of the pilosebaceous apparatus are clearly part of this hamartoma.

RMH is a congenital lesion in at least 70% of the 31 patients reported including our case (*Table 1*). There is no

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apparent sex predilection. Most RMH are solitary, with only three patients having multiple lesions.[6,21,29] Most lesions presented as a nodule (7/31), papule (6/31), skin tag (4/31) or mass (3/31). RMH occurs most commonly in areas where there is superficial striated muscle, as the nose (9/31) or chin (8/31), followed by the periorbital (4/31) and anterior neck areas (2/31).

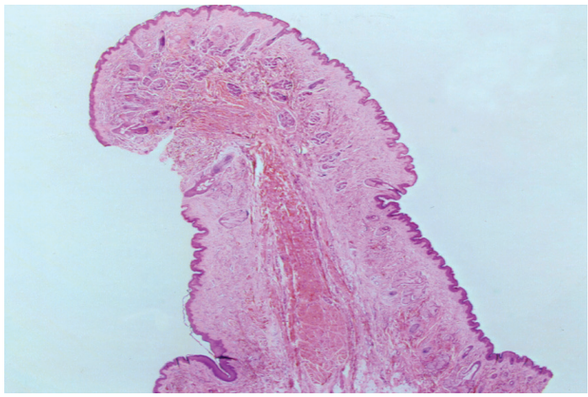

Figure 1. *Skin tag lined by thin epidermis*

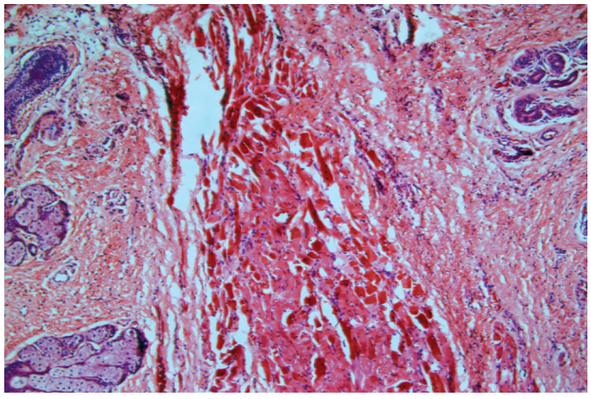

Figure 2. *Central core with skeletal muscle fibers, sebaceous glands and hair follicles are seen at the periphery*

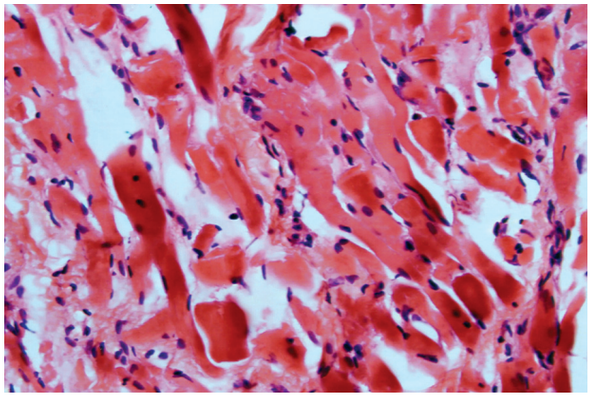

Figure 3. *Skeletal muscle fibers (40x)*

Patients with RMH occasionally have congenital anomalies like cleft lip and cleft gum as well as bilateral sclerocorneas, retinal dysplasia and amniotic band syndrome. Sanchez and Raimer[7] pointed out that some of the skin appendages associated with oculocerebrocutaneous syndrome (Delleman's syndrome) are RMH. This syndrome is characterized by orbital cysts, cerebral malformations such as hydrocephalus, central nervous system cysts, agenesis of the corpus callosum, focal cutaneous hypoplasia, and skin appendages histologically identical to RMH.

Clinically and pathologically the differential diagnosis of RMH includes accessory tragus, congenital midline cervical cleft, nasal glioma, fibrous hamartoma of infancy, rhabdomyoma, nevus lipomatosus superficialis, benign Triton tumor, acrochordon and infantile myofibromatosis.

Accessory tragus is an uncommon congenital polypoid malformation located between the pretragal (preauricular) and sternoclavicular regions, histologically composed of a thin stratum corneum and epidermis with numerous irregularly spaced hair follicles, with eccrine glands usually present. The stroma includes aggregates of mature adipose tissue, and a central plate of elastic cartilage is seen in most specimens. Small amounts of skeletal muscle may occur in some lesions.[8] Congenital midline cervical cleft is a rare anomaly of the midline anterior neck, between the mandible and manubrium. Clinically the lesion has a nipple-like configuration. Histologically, the cleft is covered by atrophic epidermis with a core of interfasciculated skeletal muscle extending into the deep dermis. Dermal adnexal structures are absent.[9] Nasal gliomas present as intranasal or extranasal masses. The characteristic histologic feature in this lesion is the presence of glial tissue.[10] Fibrous hamartoma of infancy is predominantly a mesenchymal lesion presenting as a painless subdermal nodule. Histologically, an admixture of bundles of dense fibrous connective tissue, primitive mesenchyme arranged in nests, concentric whorls or bands, and mature adipose tissue are characteristic.[11]

Adult rhabdomyoma typically occurs in the head and neck region of persons older than 40 years. Histologically, it is composed of tightly packed, large, round or polygonal cells separated by thin fibrous septa and narrow vascular channels. Fetal rhabdomyoma occurs in neonates and contains variable amounts of fibrous or myxoid tissue, with non-differentiated mesenchymal cells and striated fetal muscle.[12]

Nevus lipomatosus superficialis is recognized as papules or nodules on the hip or buttock, histologically consisting of thickened collagen bundles, increased capillaries, and mature ectopic adipocytes. Striated muscle has not been reported.[13] Benign Triton tumor (neuromuscular hamartoma) is composed of mature striated muscle fibers

intimately associated with myelinated and unmyelinated nerve fibers contained within the same perimysial-like fibrous sheaths.[14] Acrochordon or fibroepithelial polyp commonly occurs on the trunk, axial and neck skin, and may be sessile or pedunculated tumors. Histologically, it consists of a mantle of epidermis covering a protuberant fibrovascular core, and within this core occasionally nerve fibers may be observed.[15]

Conclusions

RMH is a rare benign hamartomatous lesion, with a peculiar histology, treated by excision alone. No recurrences have been reported. Although the clinical significance of RMH is only of cosmetic importance, several patients have had associated congenital anomalies. The histologic similarity between RMH and the cutaneous

Table 1. **Reported cases of rhabdomyomatous mesenchymal hamartoma**

Case no. (reference)	*Sex*	*Age*	*Clinical presentation*	*Congenital anomalies*
1[21]	M	NB	Periorbital, nasal septum, skin tags	Delleman´s syndrome
2[1]	M	NB	Sternal notch, midline, pedunculated papule	None reported
3[1]	M	NB	Upper lip, hornlike protuberance	Cleft lip, cleft palate, amniotic bands, syndactyly
4[4]	M	NB	Chin, midline pedunculated nodule, 0.6 cm	None reported
5[6]	M	NB	Multiple periorbital and periauricular polyps and nodules	Bilateral leukocoria from sclerocornea, low-set ears, preauricular sinuses
6[22]	NI	NI	Nostril	Lipoma of corpus callosum
7[5]	M	4m	Chin pedunculated midline nodule, 0.7 cm	None reported
8[19]	M	4y	Anterior midline neck, pedunculated nodule, present since birth	None reported
9[18]	F	4y	Chin, papule, 0.3 cm, present since birth	None reported
10[24]	M	4y	Chin, nodule, 1.2 cm, present since birth	None reported
11[24]	M	4y	Chin, midline nodule, 2.0 cm, present since birth	None reported
12[7]	F	3½m	Anterior neck, skin tag, 0.5 cm, present since birth	Thyroglossal duct sinus
13[7]	F	12y	Nasal alae papule, 0.4 cm, present since infancy	None reported
14[7]	M	54y	Nose papule, 0.6 cm, present 10 year	None reported
15[7]	M	48y	Nasal alae papule, uncertain duration	None reported
16[7]	M	NB	Chin skin tag, 0.6 cm	None reported
17[3]	M	71y	Lateral forehead subcutaneous nodule, 1.4 cm, present for unknown time	None reported
18[3]	M	4m	Medial eyebrow fingerlike projection, 1.3 cm	Amniotic bands, craniofacial clefts, microphthalmia, bilateral cleft lip and palate
19[17]	F	11m	Nostril sessile mass, 0.5x0.5 cm, present at birth	None reported
20[17]	F	15y	Nostril sessile mass, 0.7x0.7 cm, present at birth	None reported
21[20]	M	9y	Nostril papillomatous pedunculated lesion, 0.5 cm	None reported
22[16]	F	7m	Perianal hemangioma appeared 15 d after birth, hemangioma regressed leaving polypoid lesion	None reported
3[23]	F	14m	Sternal notch, flesh-colored papule, presents ince birth	None reported
24[2]	M	6m	Lower eyelid, smooth flesh-colored cystic-like mass, 0.8x0.7 cm, congenital	Upper lid coloboma, corneal leukoma, limbal dermoid
25[25]	F	NB	Intranasal mass with papillated surface, 0.4 cm	None reported
26[26]	F	40y	Flesh-colored plaque-like lesion above the chin	None reported
27[27]	F	18m	Subcutaneous tumor of the forehead, present since birth	Nasofrontal meningocele and dermoid cyst
28[28]	M	6m	Chin, papillomatous lesion	None reported
29[29]	F	15d	Multiple polypoid masses of tongue	None reported
30[29]	M	1m	Single polypoid mass of the left palatinic tonsil	None reported
31 (current case)	F	28d	Sternoclavicular area, skin tag, 1.4x0.8 cm, present since birth	None reported

NI: no information, NB: neonate, d: days, m: months, y: years

appendages found in Delleman syndrome is intriguing. However, at the moment it cannot be reasonably ascertain if RMH is a feature of the syndrome. Therefore, the complete and careful evaluation of a patient with RMH is of paramount importance, specifically looking for congenital anomalies associated with the above mentioned syndrome.

The striking histologic similarities between RMH and the accessory tragus and congenital midline cervical cleft are interesting. Possible hypotheses to explain the etiology of these peculiar lesions include a genetic predisposition or aberrations in the embryonic migrations of mesodermally derived tissues.

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