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Giant Connective Tissue Nevus of the Inguinal Region in a 14-Month-Old Girl: A Case Report

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Abstract

Connective tissue nevus (CTN) is a benign hamartomatous lesion characterized by the abnormal proliferation or accumulation of dermal connective tissue components such as collagen, elastin, and proteoglycans. Given its heterogeneous clinical presentation, accurate diagnosis remains challenging. Here, we report a rare case of a large, broad-based, exophytic lesion in the groin of a pediatric patient. The lesion featured a dense papillary surface covered by a mildly thickened epidermis. Scattered blood vessels were observed in the dermis of the exophytic component, along with an increase in collagen fibers. These findings were consistent with the diagnosis of CTN. This case represents an uncommon clinical variant of CTN, characterized by clinical and histopathological features resembling those of adnexal and other hamartomatous lesions. Awareness of such presentations is important for accurate diagnosis and to ensure appropriate management. Further histopathological and immunohistochemical evaluations may aid in differentiating CTN from mimics and assessing potential syndromic associations.

Categories: Plastic Surgery, Pathology
Keywords: collagen nevus, connective tissue nevus (ctn), hamartoma, pediatric groin, pediatric patient

Introduction

Connective tissue nevus (CTN) is a benign hamartomatous lesion characterized by abnormal proliferation or accumulation of dermal connective tissue components such as collagen, elastin, or glycosaminoglycans [1,2]. CTN is histologically classified based on the predominant component: collagenoma, elastoma, or nevus mucinosis [1]. Clinically, CTN may appear as firm, asymptomatic, skin-colored to yellowish dermal plaques or nodules, occurring sporadically or as cutaneous manifestations of genetic disorders such as tuberous sclerosis complex or Buschke-Ollendorff syndrome [1-3]. CTN is commonly located on the trunk and extremities; in many cases, the lesions are solitary and localized [2]. Disseminated forms have also been reported [4].

The population incidence of CTN is unknown in both adults and children, and the condition is likely underrecognized because many asymptomatic lesions are not biopsied. In a pediatric institutional series, diagnosis was frequently established during childhood, but the histological and clinical spectrum was broad [2].

Here, we report a unique case of a large, broad-based, exophytic lesion with a dense papillomatous surface in a pediatric patient.

Case Presentation

A 14-month-old female patient presented with erythematous swelling in the right inguinal area, which was first noted at the time of her 1-month health checkup. As the lesion gradually increased in size, she was evaluated at a local hospital and subsequently referred to our institution with a presumptive diagnosis of right vulvar lymphangioma.

At presentation, physical examination revealed a well-demarcated, reddish-brown, mulberry-like exophytic lesion extending from the right inguinal region to the labia majora. Routine laboratory examinations revealed no clinically significant abnormalities (table). A skin biopsy was performed, and the differential diagnoses included fibroepithelial polyps and epidermal nevus. As the histopathological findings did not support a diagnosis of lymphangioma, conservative management with close clinical observation was adopted.

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Parameter	Result	Reference range*
White blood cell count	13.7 ×10³/μL	3.5–9.8 ×10³/μL
Hemoglobin	11.9 g/dL	11.3–15.2 g/dL
Platelet count	539 ×10³/μL	130–369 ×10³/μL
C-reactive protein	0.07 mg/dL	<0.35 mg/dL
Total protein	6.6 g/dL	6.7–8.2 g/dL
Albumin	4.5 g/dL	4.0–4.8 g/dL
Aspartate aminotransferase (AST)	34 U/L	10–33 U/L
Alanine aminotransferase (ALT)	17 U/L	6–35 U/L
Lactate dehydrogenase (LDH)	240 U/L	124–222 U/L
Blood urea nitrogen	14.0 mg/dL	8.4–20.4 mg/dL
Creatinine	0.17 mg/dL	0.40–0.74 mg/dL

*Reference ranges are those used by our institution.

TABLE 1: Laboratory findings at presentation

However, the lesion rapidly enlarged, leading to functional difficulties, including fecal contamination (Figure *1*). Given the progressive enlargement, associated functional impairment, and the inability to exclude other pathological entities based on the biopsy findings alone, treatment options, including continued observation and surgical excision, were discussed. Surgical excision was ultimately selected for definitive diagnosis and treatment. The lesion was removed without a uniform surgical margin, with the excision primarily following the contour of the lesion. Where necessary, the excision design was modified to facilitate primary closure and minimize dog-ear deformity. The resected specimen measured 54 mm × 62 mm.



FIGURE 1: A well-demarcated, reddish-brown lesion is located in the right inguinal region.

Histopathological examination of the excised tissue initially confirmed the diagnosis of a fibroepithelial polyp, which was consistent with the initial biopsy findings. However, the unusually rapid growth led to concerns of underlying malignancy. Therefore, a second pathological review was performed, which ultimately resulted in a revised diagnosis of CTN.

The lesion presented as a pedunculated, elevated mass with an irregular surface, covered by a mildly thickened epidermis. Within the dermis, there were scattered vellus hair follicles, sebaceous glands, and eccrine ducts, along with dilated blood vessels. Beneath the epidermis, small blood vessels are seen in the proliferating fibrous connective tissue. No skin appendages are observed (Figure *2A*). The dermal stroma was relatively hypocellular, without significant cytologic atypia or mitotic figures. In the dermis, thick collagen fibers were predominantly arranged in haphazardly interlacing bundles. On EVG (Elastica van Gieson) staining, these thick collagen fiber bundles were highlighted (Figure *2E*). The spindle cells in the stroma were negative for desmin and αSMA (Figures *2B*, *2C*). The endothelial cells of the intervening small vessels were positive for CD34 (Figure *2D*). In addition, no smooth muscle tissue was identified in the vascular walls by αSMA or desmin immunostaining. No findings suggestive of an artery were observed. Collectively, these pathological features are consistent with collagen-predominant CTN.

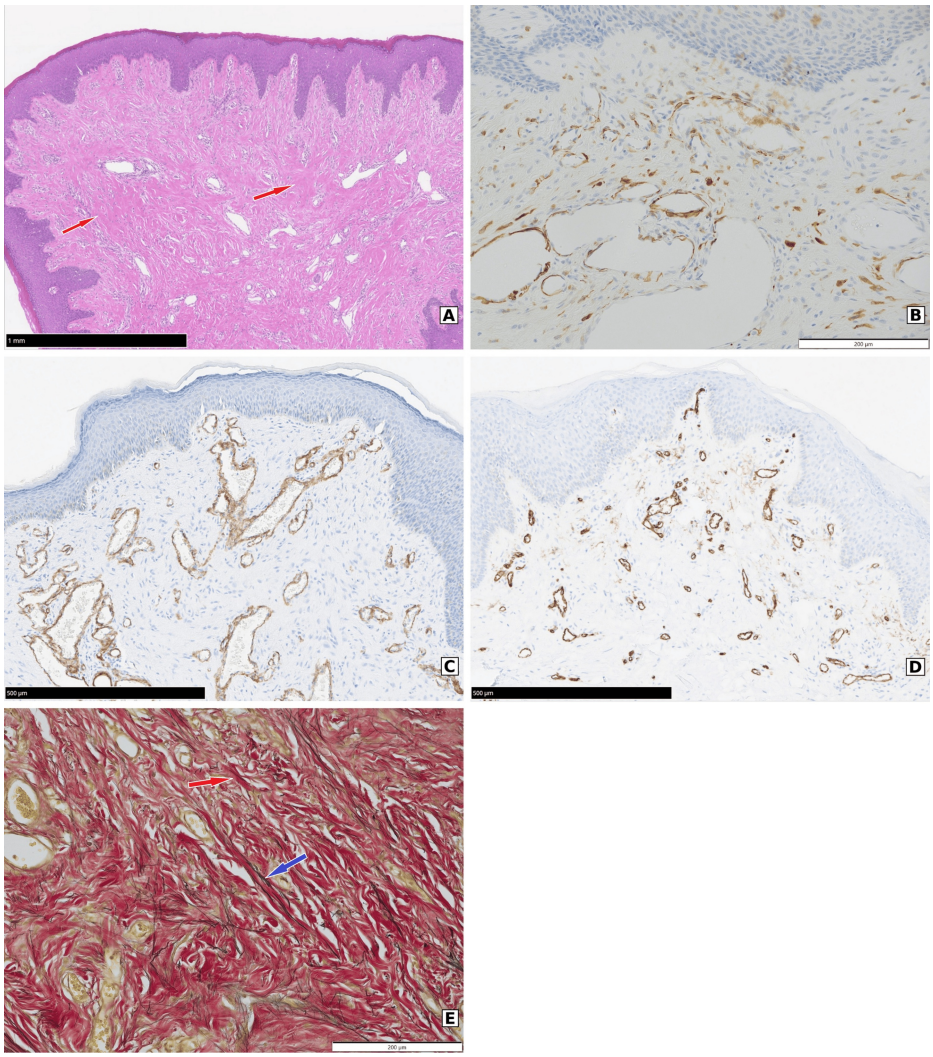


FIGURE 2: Histopathological examination of skin biopsy specimen

Multiple papillary projections of varying sizes extend outward, covered by mildly thickened epidermis. The dermis of the exophytic areas shows scattered blood vessels, accompanied by an increased number of fibroblasts and collagen fibers. Vellus hair follicles and sebaceous glands are scattered throughout, as well as eccrine ducts. No cytologic atypia, increased mitotic activity, or necrosis was identified. (A) Hematoxylin-eosin stain demonstrates thickened collagen fibers arranged in haphazardly interlacing bundles within the dermis (red arrows); (B) Desmin stain shows no significant staining of stromal spindle cells; (C) α-Smooth Muscle Actin stain shows no significant staining of stromal spindle cells; (D) Cluster of Differentiation 34 stain highlights vascular endothelial cells within the dermis; (E) Elastica van Gieson stain demonstrates thickened collagen bundles (red arrow) with preserved elastic fibers (blue arrow).

The clinical course, including the initial presentation, treatment, and postoperative follow-up, is summarized in Figure 3.

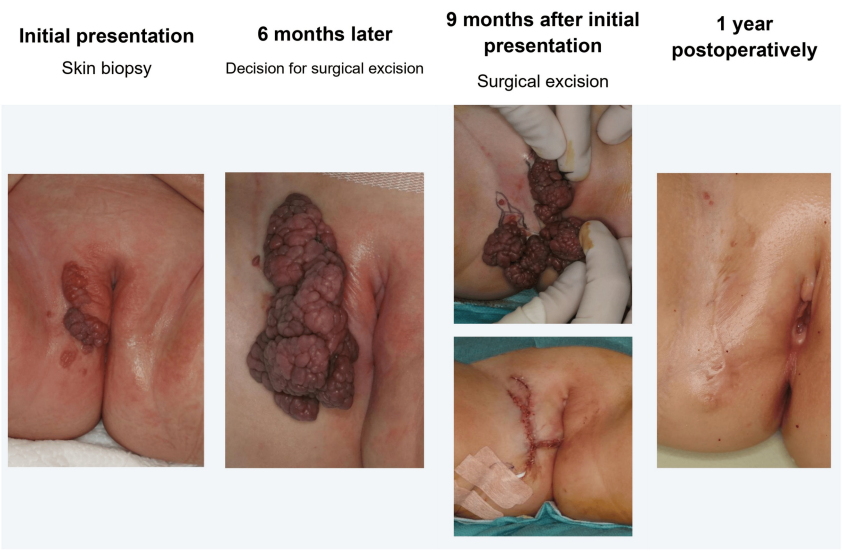


FIGURE 3: Clinical timeline from initial presentation to postoperative follow-up

Discussion

CTN is a rare benign cutaneous hamartoma characterized by an uneven distribution of extracellular matrix components within the dermis, including increased or decreased collagen fibers, elastic fibers, and glycosaminoglycans [1]. In recent years, various subtypes of CTN have been documented. The most widely accepted classification, proposed by Uitto et al. (1980) [1], categorizes CTN into collagenous, mucinous, and elastin types based on predominant histologic components, inheritance patterns, and associated systemic abnormalities. Clinically, CTN often presents as solitary or multiple firm papules or nodules that occasionally coalesce into plaques. Since lesions are generally asymptomatic, many patients do not seek medical attention, thereby making it difficult to determine their true incidence and prevalence [1,2,5]. Although CTN is generally considered to have no significant impact on overall health [5], the presence of associated systemic disorders in pediatric patients underscores the importance of early recognition and intervention. Timely diagnosis and appropriate management may prevent serious complications, improve disease prognosis, and contribute to maintaining a normal quality of life. However, it should be noted that CTN in pediatric patients may present with atypical features, potentially complicating the diagnosis.

CTN can occur sporadically or as part of genetic syndromes, such as Buschke-Ollendorff syndrome (associated with LEMD3 mutations and is characterized by connective tissue nevi and osteopoikilosis) [6], and Proteus syndrome, a complex overgrowth disorder involving multiple tissues, including the skin, bone, and fat [7,8]. The clinical appearance of CTN is highly variable, and its resemblance to other cutaneous hamartomas, such as epidermal nevus, fibroepithelial polyp, or nevus lipomatosus cutaneous superficialis, can make the diagnosis challenging.

Fibroepithelial polyps are typically small pedunculated nodules that are not usually observed as large, broad-based, exophytic lesions with a dense papillomatous surface, as noted in the present case. Furthermore, fibroepithelial polyps rarely contain cutaneous adnexal structures such as hair follicles or sebaceous glands. Fibroepithelial polyps typically contain only sparsely distributed structures [8]. In contrast, the present case exhibited numerous hair follicles and eccrine structures, which deviate from the usual histopathological profile of these polyps.

Multiple papular lesions resembling those in the present case have been described in association with nevus lipomatosus cutaneous superficialis [9]; however, we observed no ectopic adipose tissue in the dermis. Instead, the lesion demonstrated a marked increase in dermal connective tissue, predominantly characterized by proliferation and thickening of collagen bundles, which is consistent with the diagnosis of CTN.

The mulberry-like appearance of the present lesion resembles both the botryoid CTN [10] and botryoid rhabdomyosarcoma [11]. However, the present case was histologically distinct from both. Unlike the botryoid CTN reported by Huard et al. [10], which was characterized by an increased benign striated muscle component, the present lesion predominantly showed an increase in thickened dermal collagen bundles. Furthermore, in contrast to botryoid rhabdomyosarcoma, no malignant rhabdomyoblastic proliferation, significant cytologic atypia, or increased mitotic activity was identified. Thus, despite the similar gross morphology, the present case represents a collagen-predominant CTN with a botryoid clinical appearance.

Although the clinical presentation in our case differed somewhat, the occurrence of multiple similar lesions should prompt consideration of syndromic associations in the differential diagnosis [7,12]. These include Buschke-Ollendorff syndrome and, particularly for plantar lesions, Proteus syndrome.

In the present case, the lesion developed in early childhood, initially presenting as a small erythematous swelling in the inguinal region. Over time, the lesion rapidly progressed into a large mass. Subsequently, based on clinical history, physical findings, histopathology, and immunohistochemistry, a diagnosis of CTN was established. CTN is a benign cutaneous lesion, and to our knowledge, there have been no reported recurrences of solitary connective tissue nevi, particularly after complete surgical excision. In the present case, no recurrence was observed during the 1-year postoperative follow-up period.

Conclusions

We report a rare case of CTN extending from the inguinal region to the labium majus in a pediatric patient. In this case, complete surgical removal was necessitated by the rapid development and initial diagnostic uncertainty. Although histopathological examination confirmed the diagnosis of CTN, we present this case report given the rarity of its clinical presentation. Because long-term recurrence data are limited, continued clinical follow-up is appropriate.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Meishan Cui, Megumi Kawai, Hisashi Hikasa

Acquisition, analysis, or interpretation of data: Meishan Cui, Tateki Kubo

Drafting of the manuscript: Meishan Cui

Critical review of the manuscript for important intellectual content: Meishan Cui, Megumi Kawai, Tateki Kubo, Hisashi Hikasa

Supervision: Hisashi Hikasa

Disclosures

Human subjects: Informed consent for treatment and open access publication was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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References

1. Uitto J, Santa Cruz DJ, Eisen AZ: Connective tissue nevi of the skin: clinical, genetic, and histopathologic classification of hamartomas of the collagen, elastin, and proteoglycan type. J. Am Acad Dermatol. 1980, 3:441-61.
2. Saussine A, Marrou K, Delanoé P, et al.: Connective tissue nevi: an entity revisited . J Am Acad Dermatol. 2012, 67:233-9. 10.1016/j.jaad.2011.08.008
3. Pierard GE, Lapiere CM: Nevi of connective tissue. A reappraisal of their classification . Am J Dermatopathol.

1985, 7:325-33. 10.1097/00000372-198508000-00003
4. Xiao M, Yang L, Dong L, Wang Y, Sun X, Tao J: Three cases of eruptive collagenoma and a literature review, 1970-2012. Eur J Dermatol. 2014, 24:384-5. 10.1684/ejd.2014.2317
5. McCuaig CC, Vera C, Kokta V, et al.: Connective tissue nevi in children: institutional experience and review . J Am Acad Dermatol. 2012, 67:890-7. 10.1016/j.jaad.2012.01.036
6. Hellemans J, Preobrazhenska O, Willaert A, et al.: Loss-of-function mutations in LEMD3 result in osteopoikilosis, Buschke-Ollendorff syndrome and melorheostosis. Nat Genet. 2004, 36:1213-8. 10.1038/ng1453
7. Biesecker LG: The multifaceted challenges of Proteus syndrome . JAMA. 2001, 285:2240-3. 10.1001/jama.285.17.2240
8. McKee's Pathology of the Skin: With Clinical Correlations . Calonje E, Brenn T, Lazar A, McKee PH (ed): W.B. Saunders Company, Philadelphia, Pennsylvania; 2012.
9. Lane JE, Clark E, Marzec T: Nevus lipomatosus cutaneus superficialis. Pediatr Dermatol. 2003, 20:313-4. 10.1046/j.1525-1470.2003.20407.x
10. Huard E, Raynal M, Fraitag S, Angot E, Raymond S, Hebert V, Janela R: Botryoid connective tissue nevi: an uncommon presentation of hamartoma of the skin. JAAD Case Rep. 2022, 25:53-7. 10.1016/j.jdcr.2022.05.020
11. van Sambeeck SJ, Mavinkurve-Groothuis AM, Flucke U, Dors N: Sarcoma botryoides in an infant . BMJ Case Rep. 2014, 2014:bcr2013202080. 10.1136/bcr-2013-202080
12. Pope V, Dupuis L, Kannu P, et al.: Buschke-Ollendorff syndrome: a novel case series and systematic review . Br J Dermatol. 2016, 174:723-9. 10.1111/bjd.14366