Solitary Collagenoma of the Labium Majus: A Rare Occurrence

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Abstract

Collagenomas are connective tissue nevi representing hamartomatous proliferation of collagen. Isolated collagenomas are of rare occurrence. They are sporadically acquired, and lesions are often localized to a single body region. We described a case of solitary collagenoma in a 23-year-old woman, confirmed on histopathology and special stains for collagen, involving an uncommon site such as labium majus. A high index of suspicion for a skin-colored circumscribed solitary growth and a low threshold for biopsy often help to arrive at a final diagnosis.

Key Words: Collagenoma, isolated collagenoma, labium majus, vulvar collagenoma

Introduction

Isolated collagenomas are rare, sporadic, and acquired skin lesions with unclear pathogenesis. The lesions are often localized to a single body region arranged in a linear/segmental or irregular distribution. Majority of the cases in the literature have been shown to involve palm, soles, or scalp. We report a case of solitary acquired collagenoma involving labium majus that has been described previously only once in the English literature.

Case Report

A 23-year-old married woman presented to the dermatology outpatient with complaints of an asymptomatic, progressively increasing swelling involving left side of vulva of 13 years duration. Patient denied any history of preceding trauma. The lesion began spontaneously as a pea-sized soft growth over the lower part of left labium majus and increased progressively over a period of 3–4 years to involve its whole length. There was no significant past medical, surgical, or family history. Her physical examination revealed an average built woman with unremarkable systemic examination. Cutaneous examination revealed a circumscribed skin-colored swelling with a cerebriform surface having sulci and gyri, involving the whole length of left labium majus. There was complete sparing of bilateral labia minora and right labium majus. The medial side of the swelling had a smooth surface [Figure 1a and b]. It was nontender, noncompressible, and soft in consistency. The temperature of overlying skin was normal, and there was no associated thrill or bruit. There was no significant regional lymphadenopathy. Multiple, small, skin-colored soft papules and plaques could be appreciated over the mons pubis. Rest of the mucocutaneous examination did not show any abnormality. Baseline hematological and biochemical investigations were within normal limits.

We considered the differential diagnoses of connective tissue nevus, smooth muscle hamartoma, and organized vulvar lymphedema. However, the absence of overlying hair, skin-colored lesion without hyperpigmentation, and negative pseudo-Darier’s sign ruled out smooth muscle hamartoma. There was no oozing of clear fluid on multiple pinpricks making diagnosis of lymphedema unlikely. A skin biopsy from the lesion, on hematoxylin- and eosin-stained sections, demonstrated markedly increased density of collagen fibers in superficial and deep dermis as well as subcutaneous tissue that were arranged in a linear array.
a haphazard manner [Figure 2a]. These findings were corroborated on Masson’s trichrome stain that showed increased collagen bundles in green color and decreased elastin fibers [Figure 2b]. On the basis of clinical and histopathological features, a diagnosis of sporadic type of isolated collagenoma of the vulva was made. The patient was advised treatment in the form of intralesional steroids, followed by excision and vulvoplasty, if required. However, she refused any treatment in view of asymptomatic and benign nature of the lesion.

Discussion

Connective tissue nevi are benign, circumscribed hamartomas of dermal connective tissue which are classified according to the predominant component of the extracellular matrix involved, namely, collagenomas (collagen), elastomas (elastin), or nevus mucinosis (proteoglycans). Collagenomas are connective tissue nevi that represent hamartomatous proliferation of collagen and have been defined based on their pattern of distribution (localized or generalized) and mode of inheritance (acquired or inherited). The inherited autosomal dominant group comprises dermatofibrosis lenticularis disseminata in Buschke–Ollendorff syndrome, familial cutaneous collagenomas (FCC), and Shagreen patches seen in tuberous sclerosis. The acquired group includes eruptive collagenomas and isolated collagenomas. Collagenomas have also been reported to be associated with pseudohypoparathyroidism, Down syndrome, and hypogonadism.

Clinically, collagenomas manifest as asymptomatic skin-colored papules, plaques, nodules, or swelling/tumors of variable size either solitary or multiple arranged in a linear/segmental or irregular distribution usually involving the upper trunk, arms, back, thighs, and soles. Occasionally, head and neck region may also be involved. Eruptive collagenomas and FCC may be indistinguishable as both are characterized by multiple asymptomatic skin-colored papules and nodules distributed symmetrically on trunk (mostly the upper back) and upper limbs. However, FCC has a positive family history and may have associated cardiac disorders such as cardiomyopathy and conduction abnormalities. On the other hand, Shagreen patch of tuberous sclerosis present as asymmetrically distributed skin-colored plaques predominantly in the lumbosacral region, which may be single or few.

Isolated collagenomas are rare, sporadic, and acquired ones with unclear pathogenesis. The lesions are often localized to a single body region and consist of plantar cerebriform collagenoma occurring either in isolation or in association with Proteus syndrome, linear or zosteriform collagenoma, knuckle pads collagenoma, and papulolinar type of collagenoma. Majority of the cases in the literature have been shown to involve palm, soles, or scalp. Localization of lesions to labium majus as seen in our case is of a rare occurrence. To the best of our knowledge, there is only a single case report of solitary collagenoma involving the labium majus in a 9-year-old girl child, published about two decades ago. Both cases had similarities in the onset of a lesion around 10 years of age, no family history, and absence of other cutaneous or systemic complaints.

Histopathology is the gold standard for the diagnosis of collagenoma, which shows a thickened reticular dermis with haphazardly arranged thickened collagen bundles. There is increased amount of collagen fibers, with either normal or decreased elastic tissue which can be demonstrated with special stains such as Masson’s trichrome for collagen and Verhoeff–van Gieson for elastic fibers. Our case had similar histological findings.

A further workup is necessary to rule out any underlying systemic disorder, particularly in a proven case. The lesion is usually persistent with no malignant potential; therefore, active intervention is generally not advocated. Solitary/isolated lesions have been shown to respond to intralesional steroid therapy, resulting in marked effacement of lesions. Surgical excision with
primary or secondary closure may be done in patients demanding treatment.

**Conclusion**

Ours is a unique case of solitary acquired collagenoma, in a young woman with no family history and involving an uncommon site such as labium majus. To the best of our knowledge, this seems to be the first case report from the Indian subcontinent. A high index of suspicion and a low threshold for biopsy are required to arrive at a final diagnosis.

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Nil.

**Conflicts of interest**

There are no conflicts of interest.

**What is new?**

We report possibly the first case of solitary acquired collagenoma involving the labium majus, from the Indian subcontinent as despite our best search efforts, we could trace only single previous case report published so far in the English literature.

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