

Leiomyoma of Nipple: A Rare Case Report and Review of Literature

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Abstract

Cutaneous leiomyomas are benign smooth muscle tumors and are rare. They usually occur in genitourinary and gastrointestinal tracts. According to review of the literature, only about 50 cases have been reported until date. They can be solitary and rarely multiple. They are reported predominantly in middle aged females, with an approximate sex ratio (female:male) of 3:1. Nipple leiomyomas tend to be smaller than 2 cm in diameter and must be clinically differentiated from angiomyomas, glomus tumors, eccrine spiradenomas, neurofibromas, nevi, lipomas, Paget's disease, and breast carcinoma. Histopathological examination and immunohistochemistry are necessary to establish the diagnosis and to differentiate it from benign and malignant lesions. Complete excision of the tumor with histologically confirmed tumor free margin is the recommended treatment as it has high risk of recurrence. Herein we report a case of leiomyoma of the nipple of right breast in a 40-year-old woman.

Key words: Benign neoplasm, Leiomyoma, Nipple, Rare

INTRODUCTION

Leiomyoma is a benign smooth muscle neoplasm. It commonly occurs in organs such as uterus, however, can also arise in esophagus and small bowel.¹ Virchow, the eminent 19th century German pathologist stated: "Die mamma ist die amme der geschulste lehre" which meant, "The breast is the wet nurse of the student of tumor, because so many different types of neoplasm develop within it."²

Benign lesions of the breast in total are much more frequent than malignant ones. However, there are no exact epidemiologic data on the prevalence of benign or malignant lesions of the nipple and the bibliography on benign nipple tumors which have been described in the literature includes leiomyoma, milium, florid papillomatosis, syringomatous adenoma, nevoid hyperkeratosis, fibroma,

pseudolymphoma, and haemangioma.³ Leiomyoma of the nipple and areola is one of the most uncommon non-epithelial tumours.¹ Because of its low morbidity most cases go unreported. According to previous literature, only about 50 cases have been reported till date.⁴ It can even occur in breast parenchyma and may mimic malignancy clinically.⁵ In one of the largest review in English literature in the year 1989, only 19 case of leiomyoma involving the nipple or areola were identified, of which four were in men.²

Cutaneous leiomyomas can be divided into five types: (1) Multiple piloleiomyomas, (2) solitary piloleiomyomas, (3) solitary genital leiomyoma, (4) solitary angioleiomyoma, and (5) leiomyomas with additional mesenchymal elements.⁶ They can also be classified into 3 categories according to the muscle fibers from which they originate, namely, piloleiomyomas: Originating from the smooth muscle fibers of the arrector pili muscle; angioleiomyoma, originating from the tunica media of the blood vessels; and dartoic or genital leiomyoma, originating from the smooth muscles of the scrotum, nipple, areola, or vulva. Genital leiomyomas are the least common type of leiomyoma and those located over nipple and areolas are still rarer.⁷ It arises from vascular, arrector pili, genital,

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and areola smooth muscles.⁵ There is a well-developed layer of smooth muscle in the corium of the areola from which the leiomyoma might arise. Contraction of these muscularis mamillae may be seen on stimulation of the nipple.² Nipple leiomyomas can be unilateral or bilateral.⁶ It usually occurs in middle aged females, but few reports of male nipple leiomyomas associated with gynecomastia have also been reported in the literature.⁸ The groin and nipple leiomyomas are usually solitary and are asymptomatic while the cutaneous leiomyomas are sometimes painful, either spontaneously or in response to cold, emotional or tactile stimuli. The pain is thought to occur due to calcium dependent contraction of smooth muscle cells within the tumor.⁶

Other symptoms of nipple tumors include, pruritis, serosanguinous discharge, lichenification, erosion, and nodular enlargement produced by either malignant or benign nipple tumors although neoplasm occurring in the skin and subcutis tissue over breast are benign, they may be misdiagnosed clinically as primary breast carcinoma.³ We report a case of leiomyoma of the nipple in a 40-year-old female. The gross and microscopic features along with various differential diagnoses are discussed.

CASE REPORT

A 40-year-old female patient came to our hospital with 6 months history of enlarging, painful, hard lesion of right nipple. There was no history of nipple discharge or nipple ulceration. No history of breast lumps or axillary lymphadenopathy. There was no history of trauma or intake of drugs. All routine investigations were within normal limits. The lesion was completely excised and was sent for histopathological examination. Following excision, there has not been any history of recurrences during a follow-up period of 4 months.

Gross Examination

The skin covered mass was well-circumscribed, measuring about 3 cm × 2 cm × 1 cm. The cut surface showed homogenous, whitish and whorled appearance (Figure 1a and b).

Microscopic Examination

Sections studied showed raised hyperkeratotic epidermis with minimal papillomatosis with increased basal layer pigmentation. The dermis showed the presence of a well-circumscribed, unencapsulated tumor composed of spindle cells with oval shaped nuclei and eosinophilic cytoplasm arranged in interlacing bundles and short fascicles (Figure 2). The glandular elements were absent. There was no cytological atypia, mitosis or necrosis. Immunostaining



Figure 1: (a) Gross picture of well-circumscribed skin covered mass, (b) Cut surface revealing homogenous whitish surface with whorled out appearance

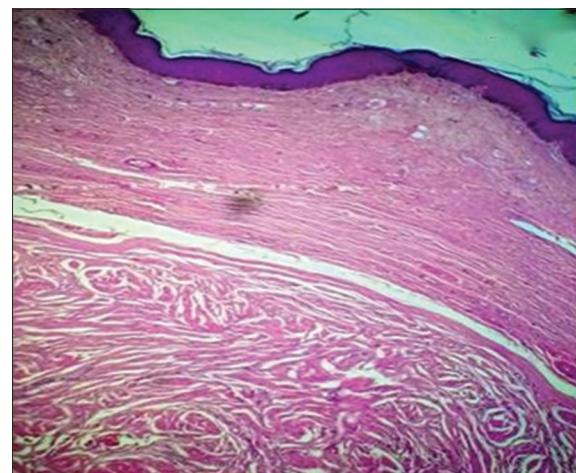


Figure 2: Microphotograph is showing a polypoid, nipple like lesion covered by epidermis with basal hyperpigmentation. The dermis shows a well-circumscribed mass composed of smooth muscle cells (H and E, x40)

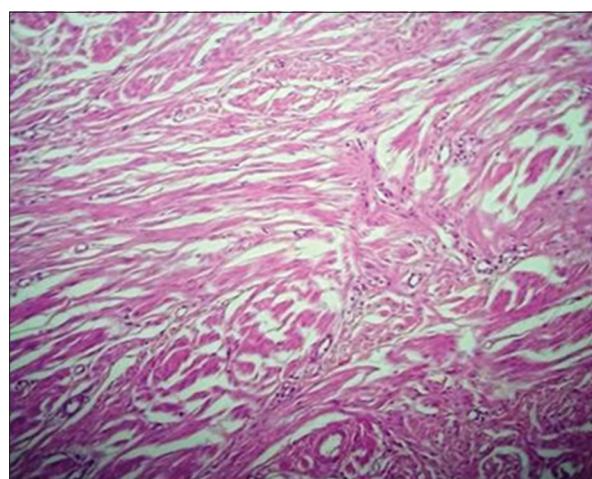


Figure 3: Microphotograph showing typical interlacing fascicles of spindle cells with abundant cytoplasm and oval nuclei with blunt ends (H and E, x100)

for smooth muscle actin was positive (Figure 3). In view of clinical, histopathological and immunohistochemistry findings, the lesion was designated as the nipple leiomyoma.

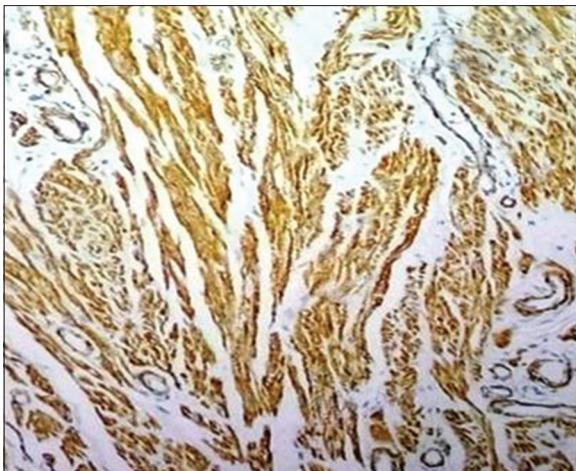


Figure 4: Microphotograph of immunohistochemistry marker (smooth muscle actin [SMA]) showing diffuse positivity (SMA, $\times 100$)

DISCUSSION

Leiomyoma of the nipple is a very rare benign neoplasm, which was first described by Virchow in 1854.⁴ These tumors are most common seen in middle aged women with an approximate sex ratio (female: male) of 3:1.⁶ In females, it has been speculated to be related to estrogen and progesterone.⁸ Other causes include trauma and certain drugs like oral contraceptives. Similar nipple leiomyomas are also seen in males, associated with gynecomastia, or idiopathic.³ Duration of these lesions ranges from 1-month to several years, and it usually involves the right breast, as in our case. No history of nipple discharge or retraction has been noted in all cases. It is usually single but occasionally can involve both breasts. Sometimes it can be a part of Reed's syndrome which is an autosomal dominant genetic condition, characterized by multiple cutaneous and uterine leiomyomatosis.⁶

The origin of leiomyoma in nipple and parenchyma has been proposed by Kaufman *et al.* in his theory that these neoplasm arise from smooth muscle cells that surround capillaries in the subcutaneous tissue of the breast. Diaz-Arias *et al.* suggested that the origin of these tumors may be from, (a) Teratoid origin with extensive overgrowth of the myomatous elements, (b) embryologically displaced smooth muscle from nipple, (c) angiomas smooth muscle, (d) multipotent mesenchymal cells, and (e) myoepithelial cells.⁵

Hereditary multiple cutaneous leiomyomatosis is a tumor predisposition syndrome characterized by multiple cutaneous and uterine leiomyomas and increased risk of developing renal cancer up to 1-7%. Hence, abdominal imaging should be done in patients with family history and multiple leiomyomas. Our patient did not have uterine leiomyomas, family history of cutaneous leiomyomas or renal mass.

Nipple leiomyomas are usually <2 cm, and it should be clinically differentiated from angioliomas, glomus tumor, eccrine spiradenomas, neurofibroma, nevi, lipomas, Paget's disease, and breast carcinoma. Histopathological examination is necessary to establish the diagnosis. Piloleiomyomas are smooth muscle tumors and are usually well differentiated. They occur mainly in the reticular dermis and are not encapsulated. The smooth muscle bundles are interlaced with varying amount of collagen. Special stains like von gieson can be used to distinguish smooth muscle from collagen. Immunohistochemical stains for desmin, S100, smooth muscle actin marker for smooth muscle differentiation may be performed to detect leiomyoma.⁶ The electron microscopy details are round to fusiform nuclei with frequent folds and notches. Moderately dispersed chromatin and surrounded by a compact nucleolus. The cytoplasm shows prominent parallel arrays of thin filaments with focal densities, numerous pinocytic vesicles, and focal aggregates of glycogen. Focal intercellular desmosome like structures can be seen.⁵

The various differential diagnosis include adenoleiomyoma, cystosarcoma phyllodes, fibroadenoma with prominent smooth muscle, fibromatosis, benign spindle cell tumor of the breast, fibrous histiocytoma, myoid hamartoma, myoepithelioma, and leiomyosarcoma. The first three lesions contain ductal elements, hence can be ruled out. Fibromatosis, benign spindle cell tumor, fibrous histiocytoma, and myoepithelioma are composed of fibroblasts, myofibroblasts, and myoepithelial cells than purely smooth muscle bundles. Myoid hamartoma is composed of scattered glandular elements between fibrous tissue and smooth muscle. Leiomyosarcoma is a malignant lesion that presents in the middle aged and elderly female patients. It is more cellular with pleomorphism, mitotic activity and sometimes coagulative type of necrosis.¹ Rarely leiomyomas show cytological atypia and mitosis.⁹ However, our case did not reveal increased cellularity, mitosis or cytological atypia.

Medical treatment includes calcium channel blocker and alpha adrenergic blockers to reduce the pain. Gabapentin an anticonvulsant drug has also been used to control piloleiomyomas related pain. The usual treatment is complete excision with tumor free margins as there is a very high chance of recurrences. Even carbon dioxide laser is also used as a treatment modality.^{6,7}

CONCLUSION

Leiomyoma of the nipple is rare benign smooth muscle tumors. Careful history taking and complete excision of the tumor with histologically tumor free margins is the recommended treatment as it has a high rate of local

recurrence up to 50%. Re-excision should be performed in the case of positive margin. Sometimes, cutaneous leiomyomas may be a sign of underlying systemic disorders. If multiple cutaneous leiomyomas are present, then detailed family history and abdominal imaging should be performed to rule out malignancy. Histopathological examination and immunohistochemistry stains help to distinguish leiomyoma from other benign and malignant breast lesions.

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