Isolated PlexiformNeurofibromatosis of Vulva; A Case Report.

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Abstract

Neurofibromas commonly involve peripheral nervous system but rarely involve genital organs such as labia, clitoris, vulva, vagina and cervix.
We presenta case of isolated plexiformneurofibroma of vulva who presented as white lesion of vulva.A tissue biopsy was done and the histopathological analysis-demonstrated a plexiformneurofibroma withspindle shaped cells with wavy nuclei and bland nuclear chromatin. No systemic sign of neurofibromatosis was found. Immunohistochemistry showed strong positivity for S-100 protein which has a strong correlation with neurofibromatosis-1.

Plexiformneurofibroma may present as isolated white lesion of the vulva without any systemic involvement in the form of neurofibromatosis. So a tissue biopsy is mandatory for definitive diagnosis of any white lesion of the vulva.

Key Words: Vaginal Cyst, HistopathologyVulva, Plexiformneurofibroma.

Introduction

Isolated neurofibroma of female genital tract is extremelyunusual and mostly associated with neurofibromatosis (6).Neurofibroma is a disease of peripheral nervous system and occurs most commonly in the extremities. Amongst female genital tract, neurofibromacommonly involvesvulva,clitoris and labia but rarely occurs in vagina, cervixendometrium, myometrium and ovary. Also it may be associated with urinary tract neurofibromatosis.(2,6)

Genitourinary neurofibroma is rare with fewer than 40 pediatric cases of genitourinary neurofibromatosis reported in the literature (3). Among the genitourinary

neurofibromas, vulva is the most commonly affected genital organ (3,4).

Neurofibromas are categorized as cutaneous neurofibromas, intraneuralneurofibromas, massive soft tissue neurofibromas and sporadic neurofibromas or those associated with neurofibromatosis-1. The solitary lesions are rare and usually they are not associated with any systemic symptom.(6)

We present a case of isolated neurofibroma of vaginal wall in 18 year old female presenting as vaginal cyst.

Case Report

An 18-year-old female (secondary school girl) patient reported to our surgical out-patient department with history of overhanging mass affecting her vulva since early childhood. Initially, the mass was small in size and painless, but progressively increased in size during last three years, resulting in discomfort while walking. There was no history of trauma or vaginal bleeding. General physical examination was normaladult female.Local examination revealed a large pedunculated mass arising from the left labiamajora (Figure 1). Mass was thick; 15cm x 9cm in diameter and 5cm at the base, with no involvement of labiaminora and clitoris. There was no extension of the mass to vagina, cervix or uterus and mass was tender.Right labiamajora and minora were normal. MRI of the perineum revealed overhanging mass on the left labia majora. Surgical excision of mass and primary repair of labia majora(Valvoplasty) (Figure2)was performed under general anesthesia. Histopathological exam of excised specimen revealed spindle shaped cells with wavy nuclei arranged in a loose myxomatousstroma.

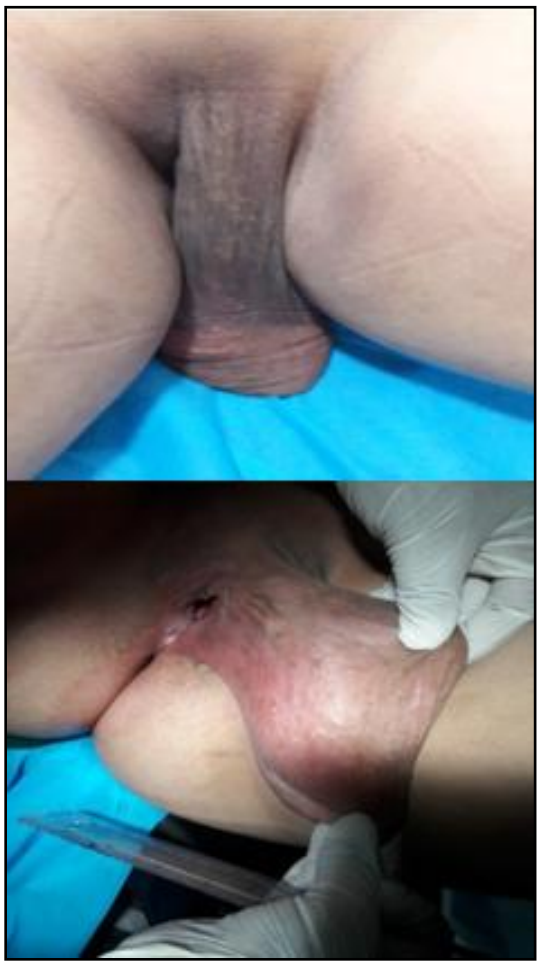

Figure 1: Preoperative photo of neurofibroma of labium majus.

Figure 2: Postoperative photo of the Patient.

Discussion

Neurofibroma is a benign tumour arising from the connective tissue of nerve sheath. Vulvalneurofibroma is very rare. Vulval involvement is found in about 18 percent of woman with Von Recklingausen's disease while approximately half of all vulvalneurofibroma are found in women with neuromatosis. (5)

Neurofibroma of vulva usually small in size (less than 3 cm in diameter) and slow growing but giant rapidly growing solitary one has been reported. Vulvalneurofibroma can be a cause of intractable chronic pelvic pain. Vulvalneurofibroma may be associated with vulval trauma and urinary tract neurofibromatosis. It may occur in any age but mostly seen in adult life. 3-5 percent cases may become malignant and may undergo sarcomatous changes.

Conclusion

Although vulvalneurofibromas are not common, they should be entertained as differential diagnosis of vulvaltumours. This tumour can be a cause of chronic pelvic pain or associated with urinary tract symptoms or dyspareunia. Excision of the tumour considered to be therapeutic. Patients who had excision of such tumor should be followed up closely because of possible recurrence or malignant changes.

Reference

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