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Vaginal Neurofibroma: A Rare Cause of Postmenopausal Bleeding Case Report and Review of the Literature

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Abstract: ***Neurofibromatosis (NF) is a dominantly inherited neurocutaneousdisorder .It isa genetic condition that mainly involves the nervous systembones, and skin.Rarely, NF affects the genital tract. Among them, vulva is the most frequent, isolated vaginal localization as a cause of postmenopausal bleedingis extremely rare. Only few cases has been reported in the literature. Case Report: We reported a very rare case of a solitary neurofibroma of the vagina in a 67-year-old woman, menopausal for 12 years, presented with post-menopausal bleeding. She did not have a family history of NF. Examination revealed a vaginal mass of 2 cm. The mass discovered to be pedunculated arising from anterior vaginal wall .The vaginal mass was excised and confirmed to be a vaginal neurofibroma on histopathology. Our patient did not have other manifestations of the disease. Conclusion: Vaginalneurofibromais a very rare cause of postmenopausal bleeding in elderly. This is the second reported case in literature. It should be considered in any painful mass at vagina.In the absence of other easily recognized signs, it may cause difficulty in diagnosis.In elderly, the incidence of malignant as major consequences should be considered.***

Keywords: postmenopausal bleeding, vaginal mass, excision, neurofibroma

1. Introduction

The neurofibromatosis (NF) are genetic disorders that have in common the occurrence of tumors of the nerve system. There are three forms of neurofibromatosis (NF): Neurofibromatosis type-1 (NF1), type two (NF2), and schwannomatosis[1].NF-1 or Von Recklinghausen's disease is the more common type of the disorder, clinically defined by the presence of neurofibromas, multiple “cafe-au-lait" spots, skin-fold freckling,Lisch nodules, skeletal dysplasia with an increased risk of malignancy[1,2].Rarely, NF1 affects the genital tract, and isolated vaginal localization is extremely rare. Only few cases of vaginal neurofibromashas been reported in the literature [3-7].

This paper reports details of an unusual case of localized vaginal neurofibromatosis presenting as a case of postmenopausal bleeding.

2. Case Report

A 67-year-old woman, menopausal for 12 years, presented with a 6-month history of post-menopausal bleeding. There was no history of any medical problem or family history of neurofibromatosis. There was no history of weight loss. On examination, she was of average built and nutrition and weighed 73 kg, pulse was 68 bpm, blood pressure was 140/70 mmHg, there were no café-au-lait spots or nodules in the skin or swelling anywhere else on the body. Heart, chest and breasts were normal. Abdominal examination was normal. Speculum examination revealed a vaginal mass of 2 cm. The mass discovered to be pedunculated arising from anterior vaginal wall away from the ureteral meatus. It was firm, non-tender, opaque, and freely mobile with one cm thick pedicle. Rest of the vagina was normal. Routine examination of blood, urine and Pap smear were normal. On Ultrasonography of abdomen & pelvis, the uterus was found to be small sized with a thick echogenic endometrial echo not corresponding to a postmenopausal uterus. A provisional diagnosis of vaginal polyp was made.

Excision of tumor and endometrial biopsy was done under general anesthesia and sent for histopathology examination. . The report revealed that the growth was composed of fibrous background fusiform cells with scanty cytoplasm and elongated cores, wavy, with fine chromatin, without cytonuclearatypia nor mitoses (Fig 1).Endometrial biopsy revealed a polypoid appearance of the endometrium without histological signs of malignancy.Histopathology diagnose the case as vaginalneurofibroma. The evidence of neurofibroma segmentally distributed without other clinical manifestations or a family history led to the diagnosis of isolated vaginal neurofibroma. The postoperative period was uneventful and the patient was discharged in good condition on the second day. On a follow up visit at 4 weeks, she was asymptomatic and had no more bleeding.

3. Discussion

Neurofibromatosis is a hereditary neurologic disorder firstlydescribed by the German pathologist Frederich von Recklinghausen in 1882 [3].This phacomatosisusually diagnosed in childhood or early adulthood, is classified into three genetically distinct subtypes characterized by multiple cutaneous lesions and tumors of the peripheral and central nervous system. Neurofibromatosis type 1 (NF1), or Von Recklinghausen's disease affecting approximately one in 3500 people. , and presents as skin lesions in the form of cafe-au-lait spots and neurofibromas at various sites like the skin, brain and spinal cord [1-4]. Neurofibromatosis type 2

(NF2), is a disease much more rare characterized by bilateral vestibular schwannomas and meningiomas[1-4]. Schwannomatosis is the most recent addition to the list of NF. It is characterized by the occurrence of multiple schwannomas [1,2].

Neurofibromas, the hallmark of NF-1, are benign tumors that arise from Schwann cells. They may occur anywhere in the body including gastro-intestinal tract, urinary bladder, heart and larynx. However, female genital tract involvement is uncommon[1,2]. The vulva is the most frequent genital organ to be affected, followed by clitoris and labia, whereas vaginal, cervical, uterine, and ovarian neurofibromas have been reported rarely [5,6]. Less than 20 cases of vaginal neurofibroma have been reported, most of them have been in association with the uterine cervix, ovary, endometrium, myometrium and the vulva [5,6].Single solitary vaginal neurofibroma is extremely rare, first described by Norris et al, there are then 12 cases described [6-15].We found only one case in elderly patient with post menopausal bleeding reported in Tunisia by Mourali et al [9]. In this case, They describeda 70-year-old woman, multiparous admitted for postmenopausal bleeding with no history of any other symptoms related to NF-1. On speculum examination, the cervix is healthy with a 1.5 cm polyp on its lower lip and the presence on the anterior side of the vagina of a polypoid mass of 2cm, pedunculated, smooth, not bleeding on contact. It was firm and not painful consistency. Excision of tumor and endometrial biopsy showed an isolated vaginal neurfibroma. Three years later, she developed a juvenile granulosa cell tumor of the ovary. On the other hand, Gomez-laencina et al described anothercase of 71-year old patient with pelvic pain and uterine mass who underwent a hysterectomy and diagnosed as a localized neurofibromatosis of the entire uterus corpus. The patient had an unknown NF-1[5].

The present case had no family history of neurofibromatosis and there was no lesions of the disease elsewhere in the body. It also represents the second case of localized vaginal neurofibroma in elderly women, presented as postmenopausal bleeding [5, 6]. Initially it was clinically and ultrasonically diagnose as vaginal polypoid. This condition should be kept in mind as a rare cause of vaginal swelling and post-menopausal bleeding. The comparison with other similar cases reported in the literature is difficult due to the paucity of series and the wide heterogeneity of cases relative to the original definition of histology and their presentations.

The major consequenses of neurofibromas is the risk of malignant transformation which increases dramatically in elderly patients. Evans et al reported 8-13 % lifetime risk of malignant peripheral nerve sheath tumor [16]. Other tumors can be associated with NF-1 include malignant gliomas, pheochromocytomas, leukemia, gastrointestinal tumors [4]. Mourali et al reported an association with a juvenilegranulosa cell tumor of the ovary in elderly women three year after the diagnosis of vaginal neurofibroma [9].Our patient has done well and is asymptomatic at present.

NF-1In elderly population, may double the risk of memory loss and cognitive impairment [4]. In this context, cognitive function should be assessed with the Mini-Mental State Examination. It is also important to educate about the possibility of the cognitive impairment and to provide neuropsychological assessment as needed.

4. Conclusion

There are few reports in the literature describing vaginal neurofibroma in association with NF-1 in elderly woman.Our case illustrates this unusual mode of presentation. The condition should be kept in mind as a rare cause of post-menopausal bleeding. The tumorshould be removed surgically to avoid the risk of degeneration. The possibility of recurrence and transformation justifies a closely follow up.

5. Consent

The patient has given written consent for publication of the current case report and accompanying pictures.

6. Conflicts of Interest

The authors report no conflicts of interest.

7. Author Contributions

All authors were involved in the patient's care. OM, and SH drafted the manuscript. All authors participated in the design. All authors read and approved the final manuscript.

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