



Case Report

Malignant hidradenoma of vulva in pregnancy-A rare case

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ARTICLE INFO

Article history:

Received 19-09-2019

Accepted 23-10-2019

Available online 06-12-2019

Keywords:

Malignant
Hidradenoma
Vulva
Metastasis

ABSTRACT

Introduction: Nodular malignant hidradenoma (NH) of vulva is a rare entity. It is a malignant, aggressive tumor of the eccrine glands that was first reported in literature as a clear cell eccrine carcinoma in 1954.

It is a rare tumor with predilection for the head face and extremities. It usually present in sixth decade of life with equal male/female distribution but cases have also been reported in children and neonates. Here we present a case of malignant nodular hidradenoma vulva in a 20yr pregnant woman.

Case Presentation: A 20yrs old primi patient at 38weeks pregnancy presented to our outpatient department with a painless vulval swelling since 3 months which had increased rapidly in size. On examination the swelling was arising from upper one third of right labia minora. The mass was 4 x 4 cm in size and was displacing the urethral meatus towards left and involving the vagina upto the fornices. The patient had history of excision of similar vulvar swelling 7 months back and the histopathology was suggestive of hidradenoma vulva. Patient delivered vaginally after 2 days. She was thoroughly investigated. CT scan lower abdomen and pelvis and chest x-ray revealed that it was a locally invasive tumor with distant metastasis. Patient was given neoadjuvant chemotherapy for 3 cycles after which surgical excision of the residual disease with bilateral inguinal lymphadenectomy done post operatively 3 more cycles of chemotherapy given. Patient was kept in regular follow up.

Conclusion: Malignant hidradenoma of vulva is a locally aggressive tumor with rapid progression, poor prognosis and high recurrence. Early diagnosis and excision with adjuvant chemotherapy is imperative.

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1. Introduction

Malignant Hidradenoma is a rare sweat gland tumor that occurs less commonly in vulval and perianal area. It was first reported in literature in 1954 as “clear cell hidroadenoma or acrospiroma” by Keasbey et al. The tumor arises from the intradermal duct of eccrine sweat glands.¹ It accounts for 6 percent of malignant eccrine tumors. It is a rare oncological entity with less than 100 cases reported till date. Malignant hidradenoma usually arises in the head and neck region and the extremities. It usually presents in the sixth decade of life with equal male/female distribution but cases have also been reported in children and neonates. An unusual case of occurrence of malignant hidradenoma of vulva in 20yrs pregnant woman has been described here.

2. Case Report

A 20 yrs old primi patient at 38 weeks of pregnancy presented to our outpatient department with the chief complaint of a painless vulval swelling since three months. She had noticed the swelling as a small nodule on the right labia minora which had rapidly increased in size in three months.

Patient had a past history of occurrence of a similar swelling 7 months back on the same site which was slow growing painless and reached a size of 3 x 3 cm. Patient had history of excision of that swelling in a private hospital and the excised mass was sent for histopathological examination. HPE report was suggestive of hidradenoma vulva.

On examination, the present swelling was arising from the upper one third of right labia minora. It was 4 x 4 cm in

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size, well circumscribed, nodular and solid in consistency. It was not infiltrating the adjoining vulva. The growth was displacing the urethral meatus to left side and was also involving the vagina upto the right fornix. A solitary enlarged 2 x 1 cm hard right axillary lymph node was also palpable. Patient delivered vaginally after 2 days.

Patient was thoroughly investigated for the type and extent of malignancy. Investigations including ultrasound lower abdomen and pelvis chest x-ray, computed tomographic scans of the chest and abdomen and pelvis were done.

Biopsy of the mass was done. Microscopic section was suggestive of a cellular dermal adnexal tumor with cells disposed in circumscribed lobules separated by fibrous septa. The tumor cells were round to polygonal with micropapillary and ductal differentiation. Individual cells were bland with some areas showing mild nuclear pleomorphism, mitoses and cytoplasmic clearing. The tumor cells at the periphery were seen infiltrating the surrounding muscles and vascular emboli were evident. These findings were suggestive of low grade malignant nodular hidradenoma.

CT Scan abdomen and pelvis revealed a well defined heterogenous enhancing soft tissue attenuation lesion measuring 15 x 15 x 13 cm extending from hypogastrium to introitus. The lesion was seen abutting and compressing the urinary bladder towards left lateral position and abutting and pushing the uterus posteriorly with displacement of bilateral ovaries. Chest x-ray was suggestive of metastasis in right lower lung field.

All these investigations were suggestive of the fact that it was a locally invasive tumor with distant metastasis belonging to T3N2M1 Stage IV disease according to AJCC classification.²

Patient was planned for neo adjuvant chemotherapy. She was subjected to 1st cycle of CAF regime consisting of cyclophosphamide 600mg/m², 5-fluorouracil 600mg/m² and adriamycin 60mg/m². And was advised follow up after 21 days. At the 1st follow up visit there was marked reduction in the size of the lump as evident by a repeat CT scan. She was given 2 more cycles of chemotherapy.

This was followed by resection of the residual vulval growth of size 3 x 3 cm along with bilateral inguinal lymphadenectomy. HPE of lymph nodes was negative for metastasis. Postoperatively she was given 3 more cycles of chemotherapy and further kept in regular follow up.

3. Discussion

Malignant hidradenoma is a rare sweat gland tumor. They can be classified as nodular hidradenoma, hidradenoma papilliferum, solid cystic hidradenoma, clear cell hidradenoma, clear cell acrospiroma and eccrine acrospiroma.

Overall incidence is <1 percent of all skin neoplasms. Malignant nodular hidradenoma is more common in sixth decade of life with no sex predilection.³ Body

involvement is 65 percent soles, 10 percent on palms and 25 percent in other areas of body. Unusual hidradenomas may appear on the genitals of females of labia majora, minora and interlabial sulcus.³ Clinically the tumor may be asymptomatic but may be associated with itching, pain, bleeding or discharge.⁴

The tumor has a high local recurrence rate. Distant metastasis occurs through hematogenous route to para aortic and retroperitoneal lymph nodes, bones, vertebrae, ribs, pelvis, lungs and pleura.⁵

Management involves surgical excision with at least 2 cm free margins. Sentinel lymph node biopsy is suggested to detect regional lymph node metastasis. Positivity of sentinel lymph node is considered as an indication for radical lymphadenectomy.

These tumors are usually radioresistant.⁵ Recommended treatment in advanced disease is chemotherapy followed by surgical excision of the residual disease at the primary site. Chemotherapeutic drugs commonly used are cyclophosphamide, 5-fluorouracil, cisplatin, paclitaxel and adriamycin.

Recurrence rate of malignant hidradenoma is as high as 50 percent. Prognosis is poor and 5 year survival rate is less than 30 percent.⁵

4. Conclusion

Malignant hidradenoma of vulva is a locally aggressive tumor. It has a rapid progression, poor prognosis high rate of metastases and high recurrence rate. Due to rarity of tumor there has yet to be an established standard of care. Radical excision is widely used and lymphadenectomy is frequently reported. Further adjuvant chemotherapy and radiotherapy seems to improve survival in patients with advanced and recurrent cancer. Close follow up for recurrent disease is required.

5. Source of funding

None.

6. Conflict of interest

None.

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Cite this article: Gupta M. Malignant hidradenoma of vulva in pregnancy-A rare case. *Indian J Obstet Gynecol Res* 2019;6(4):557-559.