MALIGNANT HIDRADENOMA OF VULVA IN PREGNANCY- A RARE ENTITY

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Abstract

Malignant nodular hidradenoma (MNH) of vulva is a rare entity. It is a malignant primary skin tumour of eccrine sweat glands and is aggressive in nature. We are reporting this case of 28-year-old primigravida with 38 weeks pregnancy who presented with a vulval swelling and histopathology report of malignant nodular hidradenoma of vulva. Imaging studies showed metastatic disease. Patient was managed with chemotherapy after three weeks of vaginal delivery followed by surgery and chemotherapy, but tumour recurred and is planned for second line chemotherapy. Malignant nodular hidradenoma (acrospiroma) is a rare vulval tumour. Early diagnosis and excision with adjuvant chemotherapy is imperative.

Keywords: Vulva, Hidradenoma, Metastasis, Chemotherapy

Introduction

Malignant nodular hidradenoma of vulva is a rare entity. It is a malignant, aggressive tumour of the eccrine gland that was first reported in the literature as a clear cell eccrine carcinoma (acrospiroma) in 1954. It is a rare tumour with predilection for the head, face and extremities. It generally presents as an ulcerated reddish nodule in elderly (sixth decade of life) with an equal male/female distribution but has also been reported in children and neonates. Here we present a case report of malignant nodular hidradenoma vulva in a 20 year pregnant woman.

Case report

A 20-year old primigravida patient at 38 weeks pregnancy presented to the outpatient Department of Obstetrics & Gynaecology, King George Medical University, Lucknow with a vulval swelling since three months. She first noticed the swelling as a small nodule on the right labia minora which had rapidly increased in size over the past three months. Patient gave history of occurrence of a similar swelling seven months back. The mass was slow growing, non-tender and reached the size of 3x3cm. Patient underwent excision of the swelling at a private hospital and the excised mass was sent for histopathological examination which showed a cellular dermal adnexal tumour with tumour cells showing nuclear pleomorphism, mitosis and cytoplasmic clearing. All features were suggestive of malignant nodular hidradenoma (acrospiroma) of vulva (Figure 1).

The swelling recurred after 5 months. On examination the swelling was present in the upper one third of right labia minora. It was a 4x4cm in size well circumscribed, nodular; firm in consistency, non-infiltrating the adjoining vulval tissue. The growth was displacing the urethral meatus to left side and was also palpable up to the fornix. A solitary enlarged 2x1 cm, hard right axillary lymph node was also palpable. Patient was...
considered for chemotherapy after delivery. Patient delivered vaginally at 38 weeks 2 days period of gestation (POG.) Patient was thoroughly investigated for the type and the stage and extent of malignancy. Investigations included certain blood and imaging investigations – ultrasound lower abdomen and pelvis, chest X-Ray, computed tomographic scans of chest, abdomen and pelvis. CT scan abdomen and chest X-Ray were suggestive of locally invasive tumour along with distant metastasis belonging to T3 N2 M1 TNM scoring system (Tumour size, Lymph Nodes affected, Metastases) stage IV disease according to AJCC (American Joint Committee on Cancer) classification (Figure 2).

Figure 2. CT scan abdomen and pelvis showing a well-defined heterogeneously enhancing soft tissue enhancing lesion (measuring 15x15x13 in cm) extending from hypogastrium to introitus compressing urinary bladder, uterus posteriorly and causing bilateral displacement of ovaries.

Patient was given first cycle of chemotherapy after three weeks of vaginal delivery, CAF (Cyclophosphamide, Adriamycin, Fluorouracil) regimen consisting of cyclophosphamide 600mg/m², 5-fluorouracil 600mg/m² and adriamycin 60mg/m² and discharged in stable condition and advised to follow up after 21 days. At first follow up visit there was marked reduction in the size of the lump as evident by a repeat CT scan and on clinical findings. Patient was given three more cycles of chemotherapy. This was followed by the resection of the local mass of size 3x3 cm along with bilateral inguinal lymphadenectomy. Histopathology of the tumour showed malignant eccrine acrospiroma, lymph nodes were negative for metastasis (Figure 3).

Figure 3. Section shows small neoplasm lying in small nests and islands in a fibrocollagenous stroma. Individual tumour cells are monomorphic round cells, have large nuclei, prominent nucleoli surrounded by scant cytoplasm, surrounding stroma shows muscle bundles, adipose tissue and haemorrhagic foci.

Postoperatively she was given three more cycles of CAF chemotherapy and showed complete regression of disease clinically on imaging investigations and was kept on further follow up.

Discussion

Malignant nodular hidradenoma (MNH) was first reported as clear cell eccrine carcinoma by Keasbey and Hadley in 1954. First line of management is usually surgical excision with at least 2 cm free margins. Some authors recommend a primary resection of 3-5 cm of safe margins. An inadequate excision of the primary lesion appears to be a poor prognostic factor because even lesions classified as benign have a propensity to recur locally if not fully excised. There is not an established consensus on lymph node dissection. The 5-year postsurgical survival rate for malignant nodular hidradenocarcinoma is reported to be less than 30%. As in our case patient had come with the histopathological report of malignant hidradenoma, patient was managed with neo-adjuvant chemotherapy, followed by surgery and adjuvant chemotherapy. Patient had regression of disease completely both clinically and on imaging techniques. Patient remained asymptomatic for 6 months after which there was recurrence of disease. MNH’s pick of incidence occurs in sixth decade of life; with an equal male/female distribution; the tumour has a high local recurrence rate (50%), metastases to lymph nodes, bones or visceral organs (60% of cases) usually recurs within the first two years of diagnosis. As in this patient the disease recurred
after 6 months of treatment. Two cases have been reported recently\textsuperscript{2,4} in which patients were treated with wide local excision followed by adjuvant therapy, but both patients developed local recurrences (14 months and 8 years respectively) along with distant metastasis. In this patient also in spite of chemotherapy and surgery patient had recurrence.

**Conclusion**

Malignant nodular hidradenoma of vulva is rare. The usual sites of occurrence are head, face and extremities and its occurrence in the vulva is extremely rare. It is a locally recurring tumour, hence its primary management is surgical excision followed by adjuvant chemotherapy, and usually the prognosis is poor in these cases.

**Editor’s comment**

The Malignant nodular hidradenoma (MNH) is an uncommon, aggressive malignant cutaneous adnexal tumour that can show differentiation towards various components of eccrine sweat glands. Early treatment should be contemplated in all cases. Wide surgical excision of the tumour with at least 2 cm of clear margins for both primary disease and local recurrences is the treatment of choice. Followed by chemotherapy remains controversial. Radiotherapy may also be considered though its role needs to be substantiated further.

**Conflict of Interest**

The authors declare that there is no conflict of interest and no financial support.

**References**


