Türk Kadın Sağlığı ve Neonatoloji Dergisi

To cite this article: Kilic C, Cakir C, Yuksel D, Karalok A, Mesci CG, Turan T. Vulvar granular cell tumor: A rare entity. Turk J Womens Health Neonatol 2021; 3(1): 22-24

Case Report

Vulvar granular cell tumor: A rare entity.

Vulvar granüler hücreli tümör: Nadir bir olgu sunumu

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Abstract

Granular cell tumor (GCT) is a rare soft tissue tumor derived from neural sheath and Schwann cells. They are usually located in skin, viscera, subcutaneous, submucosal tissues of the head and neck, especially in the tongue and mouth. Vulva is involved in 10% of the patients. The majority of GCTs tend to be benign however local recurrences may develop. Local surgical excision is curative for benign lesions. In this article, a benign vulvar granular cell tumor is presented.

Keywords: Vulvar cancer; granular cell tumor; surgery

Öz

Granüler hücreli tümör, nöral kılıf ve Schwann hücrelerinden köken alan nadir bir yumuşak doku tümörüdür. Genellikle deri, iç organlar, baş ve boyunun subkutan ve submukozal dokuları, ağız ve dil yerleşimlidir. Hastaların %10 kadarında vulva tutulumu görülmektedir. Genelde benign karakterde olan bu tümörde lokal rekürrenslere rastlanmaktadır. Benign lezyonlar için lokal cerrahi eksizyon küratiftir. Bu makalede benign bir vulvar granüler hücreli tümör olgusu sunulmuştur.

Anahtar Kelimeler: Vulvar kanser; granüler hücreli tümör; cerrahi

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Received: 30.08.2020 Accepted: 01.01.2021

1. Introduction

A granular cell tumour (GCT) is a rare soft tissue tumour derived from neural sheath and Schwann cells (1,2). It was first described by Weber in 1854 (3). In 1926, Abrikossoff reported five cases and used the term myoblastoma (4). Since then, over 130 cases have been reported in the literature (5). They are usually located in skin, viscera, subcutaneous, submucosal tissues of the head and neck, especially in the tongue and mouth (6, 7). Although there have been reports indicating different localizations in the female genital tract, the vulva is involved in 10% of the patients (5). GCTs occur more commonly in women and the black population (8). They can be seen at all ages, with a peak incidence of 50 years (9). The majority of GCTs tend to be benign; however, local recurrences may develop (10). Less than 2% of these tumours were reported as malignant (11). GCTs generally present as small, slow-growing, solitary and painless subcutaneous nodules (12).

This article presents a case of a benign vulvar granular cell tumour diagnosed and treated in our gynecologic oncology clinic.

2. Case report

The patient aged 50, with gravida 2 and parity 1, applied to our gynecologic oncology clinic with a complaint of vulvar mass located on the right labium majus. The patient had no history of pain, bleeding or discharge from the lesion. Her medical history was unremarkable. Transvaginal ultrasonography showed no abnormal findings in the uterus and adnexa. The result of the cytologic examination of the uterine cervix with Papanicolau smear was within normal limits. A-2 cm nodular, firm and painless vulvar mass was found on the middle region of the right labium majus in the gynaecological examination. Overlying cutaneous tissue was intact. An incisional biopsy was performed under local anaesthesia, and the pathology report of the mass revealed a benign granular cell tumour of the vulva. The tumour involved surgical margins. A positron emission tomography and computed tomography (PET-CT) scan of the whole body was then performed. The metabolic fluorodeoxyglucose (FDG) uptake of vulvar mass was found to be minimally increased. The patient underwent wide local excision with a-1 cm clear margin under general anaesthesia. The final pathology report confirmed the first diagnosis of benign vulvar granular cell tumour **(Figure 1)**. The tumour size was measured as 1.7x1 cm, and surgical margins were free of tumour. No adjuvant therapy was applied. The patient was followed-up for 14 months without any sign of local or distant recurrence. Signed informed consent was obtained from the patient.

Pathologic findings

Microscopically, Haematoxylin and Eosin (HE) stained sections revealed an intact epidermis without any signs of hyperplasia or ulceration. Immunohistochemical analysis was not performed. A tumour composed of sheets of eosinophilic granular cells with bland nuclei intermingled with strands of collagen and occasional chronic inflammatory cells were detected in 100xHE. Necrosis was absent. Microscopic appearance with HE staining was shown in **Figure 1.**

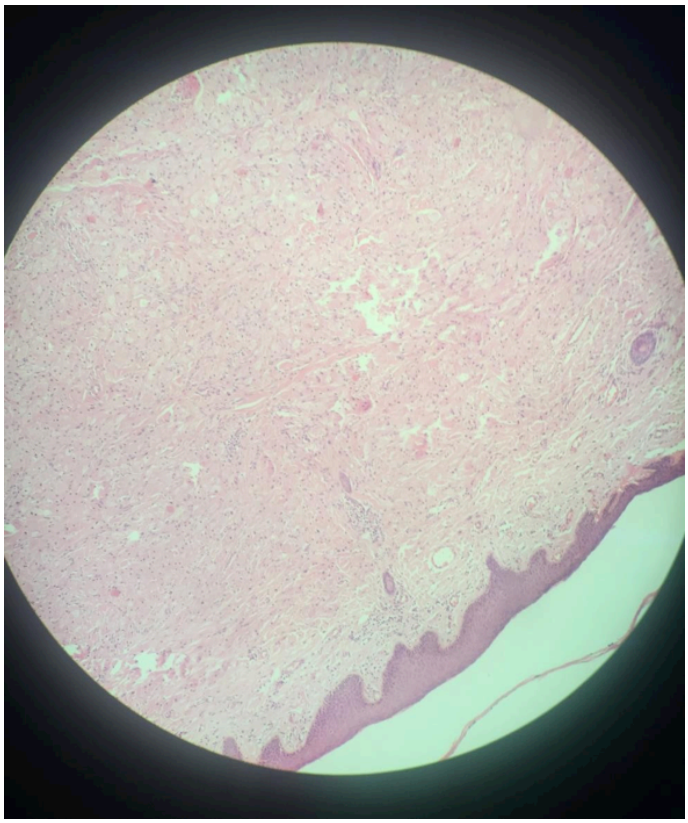

Figure 1. Sheets of eosinophilic granular cells with bland nuclei intermingled with strands of collagen and occasional chronic inflammatory cells, 100xHE

3. Discussion

GCT is a benign soft tissue tumour derived from the neural sheath. They often present as a painless vulvar mass in most cases. The differential diagnosis includes dermatofibroma, hidradenoma papilliferum, epidermal inclusion cyst and Bartholin's gland tumours (13). It can be seen in all parts of the body, especially in the head and the neck region. Also, vital organs can be involved. They are usually seen in the fourth and sixth decades of life (14). However, different studies from all age groups were reported, including childhood and infancy. Our patient has a typical history of benign GCT.

More than 98% of cases have been reported as benign. Malignant

GCT, located in the vulva, is extremely rare, and less than ten patients were reported in the literature (10). In the presence of malignancy, the lesions tend to be multicentric and metastatic. The diagnosis of malignancy can be confirmed with at least three of the following features: necrosis, vesicular nuclei with prominent nucleoli, spindling, mitosis count > 2/ 10 High Power Fields, high nuclear/cytoplasmic ratio and pleomorphism (15). One or two criteria positivity indicates atypical tumours. Also, rapid tumour growth, metastatic lesions, invasion into adjacent structures and tumour size ≥ 5 cm are clinical indicators of malignancy.

GCT can be diagnosed by histopathological assessment. The overlying epidermis usually demonstrates pseudoepitheliomatous hyperplasia (16). Therefore an adequate biopsy, including the dermis, would help differentiate pseudoepitheliomatous hyperplasia from squamous cell carcinoma. The immunohistochemical analysis could be performed for confirmation of diagnosis. GCT cells are positive for S100 proteins, vimentin, inhibin, calretinin, Neuron-specific enolase (NSE) and CD 68 but are negative for Melan-A, gp100, HMB45, keratins, CD31, CD34, smooth-muscle actin and desmin (17). The proliferation index with Ki-67 and immunostaining for p53 overexpression are higher in atypical and malignant forms (15).

Surgery is considered as primary therapy for the treatment of GCT. Local surgical excision is curative for benign lesions. Benign GCTs do not have a capsule, but local infiltration of tumour cells can be detected. If surgical margins are involved, re-excision of the lesion is recommended (18). Malignant forms have an inadequate response to chemotherapy and radiotherapy. For the initial treatment of malignant GCT, radical local excision with regional lymph node dissection should be performed (12). Our patient did not receive adjuvant therapy and did not develop recurrence. However, even benign tumours have a tendency for local recurrence at the excision site (19). Thus GCTs should be followed up for any sign of local or distant recurrence.

Declaration of Interest

The authors report no conflict of interest.

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