

Granular Cell Tumor of Vulva: Differentiation from Malignant Lesions

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Abstract

Granular Cell Tumors (GCT) is very uncommon tumors and was first described by Abrikossoff in 1926. They are small, painless, slow-growing subcutaneous nodules most commonly seen in the tongue and oral mucosa. 5-16% of the granular cell tumors have been reported in the vulva. To our knowledge, the imaging findings of GCT of the vulva were not described in the literature before. In this case, we present the imaging findings of a 63-years-old female who had GCT of the vulva. On ultrasonography there was ill-defined, lobulated heterogeneous solid mass with central hypoechoic and peripheral slightly hyperechoic areas in vulvar subcutaneous fat. On MRI, the tumor was hypointense on T1 and T2-weighted images compared to fat tissue, isointense with muscles on T1-weighted images and on T2-weighted images slightly iso-hyperintense with muscles. Dynamic examination showed early diffuse enhancement more peripherally, which were also continuing on late phase images. On CT, the tumor was isodense to adjacent muscles and showed minimal enhancement. There were no regional or distant metastasis and no lymphadenopathy. The tumor was treated by wide surgical excision and pathology was consistent with granular cell tumor.

Keywords: Differentiation • Granular cell tumor • Vulva

Introduction

Granular Cell Tumors (GCT) is very rare tumors and first described by Abrikossoff [1]. These tumors are soft tissue neoplasms of nerve sheath origin [2]. Between 5-16% of the granular cell tumors have been reported in the vulva [3]. Most of the tumors are benign but rarely malignant variants have been reported [2]. Local surgical excision is considered as a curative therapy for benign GCTs. It should be differentiated from soft tissue sarcomas. In this report, we present the cross-sectional imaging findings of a case of GCT of vulva. There are only a few cases in the literature that discussed the imaging findings of GCT occurred in the breast, inferior rectus muscle, the skull base, the respiratory tract and intradural extramedullary region of the spinal canal [4-6]. To our knowledge none of the cases in the literature discussed the imaging findings of GCT of the vulva. Herein, we aimed to report and discuss the imaging findings of the GCT of the vulva.

Case Report

A 63-years-old woman was admitted to our Obstetrics and Gynecology Department with an enlarging mass on her right labium majus. The patient indicated a history of mild swelling on the labium which showed slow increase in size since one and a half year, and had pain while sitting. On physical examination, there was a painful, swollen, large lesion involving the right vulvar subcutaneous tissue. There was no ulceration or discharge on the lesion. Ultrasonography (US) demonstrated an ill-defined, lobulated 36 × 19 mm sized heterogeneous solid mass with central hypoechoic and peripheral slightly

hyperechoic areas (Figure 1). These features can be found in most primary and secondary soft tissue tumors. Magnetic Resonance Imaging (MRI) was performed to further evaluate the lesion. It showed a 39 × 35 × 22 mm sized well-demarcated solid mass of the right posterior vulva in the subcutaneous fat adjacent to obturator internus muscle and the inferior part of the right ischium. The mass lesion had a lobulated, smooth contour. The tumor was hypointense on T1-weighted images compared with adjacent fat tissue and isointense with muscles. On T2-weighted images the tumor was hypointense compared with adjacent fat tissue and iso-slightly hyperintense compared with muscles. After contrast material administration, the tumor showed markedly diffuse enhancement from early arterial phase until late venous phase (Figure 2).

On CT the tumor was diffusely hyperdense compared to fat tissue and isodense to the adjacent muscles (Figure 3). Our differential diagnosis of the underlying tumor was primarily fibroma and also soft tissue sarcomas of fibrous or muscular origin because of the imaging characteristics although there was no necrotic or hemorrhagic component. Chest and abdominal Computed Tomography (CT) were performed to evaluate distant metastasis before the surgical treatment. There was no regional or distant metastasis and no lymphadenopathy on CT scans. The tumor was treated by wide surgical excision. The microscopic examination of the tumor revealed a well circumscribed mass formed by epithelioid-appearing cells with abundant granular cytoplasm. Nuclei of those cells were uniform in size and no mitosis was seen. The tumor cells are immunoreactive for S-100 protein and CD68

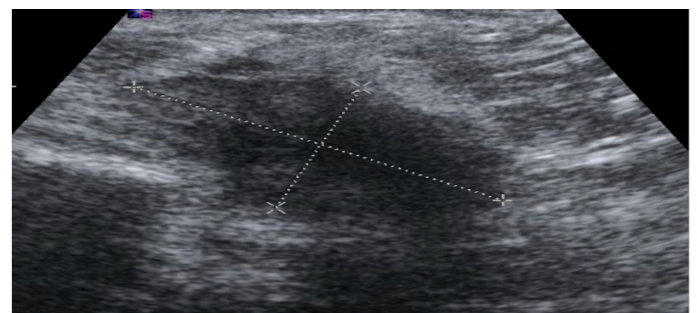


Figure 1. Ultrasonography (US) shows an ill-defined, lobulated heterogeneous solid mass with central hypoechoic and peripheral slightly hyperechoic areas.

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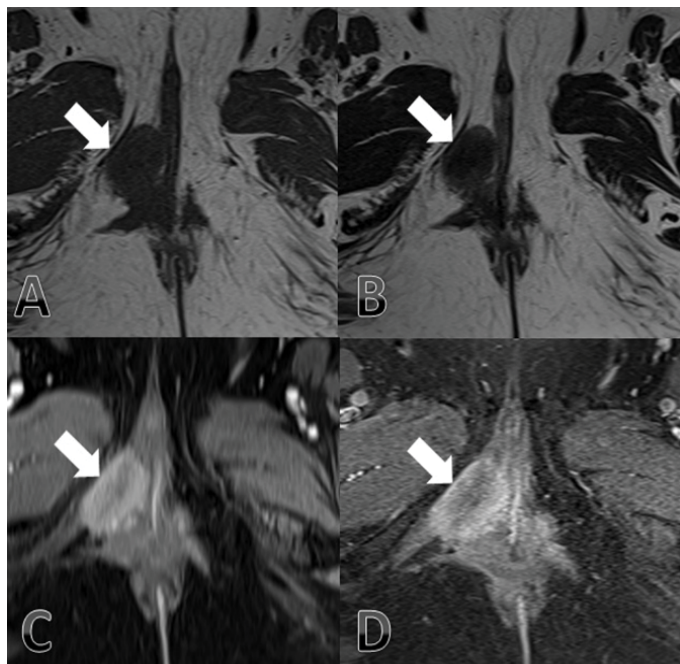


Figure 2. On axial T1-weighted image (A) the tumor is hypointense compared with adjacent fat tissue and isointense with muscles. On axial T2-weighted image (B) the tumor is hypointense compared with adjacent fat tissue and slightly iso-hyperintense compared with muscles. After contrast material administration, the tumor shows markedly diffuse enhancement from early arterial phase (C) until late venous phase (D).

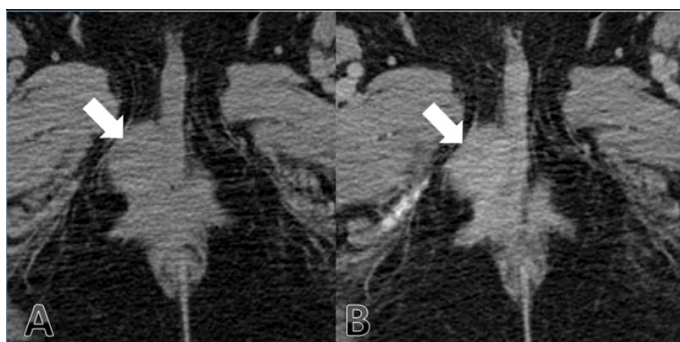


Figure 3. On axial CT image the tumor is hyperdense compared with fat tissue and isodense compared with adjacent muscles (A), after contrast material administration, the tumor shows markedly diffuse enhancement (B).

while EMA positivity was not observed (Figure 4). The findings were consistent with the diagnosis of granular cell tumor of the vulva. The patient has been followed for 6 months without local recurrence or any intra-abdominal lesions..

Discussion

GCTs are usually small, painless, slow-growing subcutaneous nodules. They are usually not encapsulated and recurrence is common. The malignant granular cell tumor is extremely rare, representing 1% to 2% of cases and has high rate of metastases and short survival [2]. GCTs may arise in different part of the body; and more than 40% of the cases are found in the tongue. The other sites are subcutaneous tissue, respiratory tract, gastrointestinal tract, urogenital tract, breast, skull base, central nervous system and orbit. 15% of the cases may simultaneously have several anatomical regions [2-6]. 5-16% of the granular cell tumors have been reported in the vulva [3]. The tumor is more common in Africo-American woman, during the 3rd-6th decades [2]. Benign GCTs generally have slow growth rate and are more common as an asymptomatic mass. Although our case was a slowly growing vulvar mass, the patient had pain as the presenting symptom while sitting. On the imaging techniques (such as thorax and abdominal CT) there were no additional lesions, no regional or distant metastasis and no lymphadenopathy. Clinically,

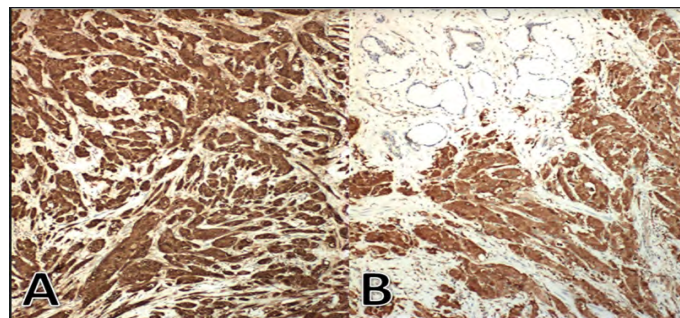


Figure 4. Nuclei of the cells are uniform in size and no mitosis is seen. The tumor cells are immunoreactive for S-100 protein and CD68.

if it was a painless lesion differential diagnosis might be a cystic lesion, sebaceous cyst, papilloma, lipoma and fibroma. But the lesion was painful and the ultrasonographic findings indicated a heterogeneous, lobulated, ill-defined solid mass. We preferred MRI for obtaining more detailed information about the extent of tumor invasion and for making an accurate diagnosis. To our knowledge, there are only a few cases in the literature that discussed the imaging findings of GCT occurred in the breast, inferior rectus muscle, the skull base, the respiratory tract and intradural extramedullary region [4-6]. In these reports, despite these tumors were located in the different anatomical regions of the body, the imaging findings of the GTCs were very similar with each other. Hashimoto et al. had reported the granular cell tumor of the inferior oblique muscle of eye. The GTCs appeared homogeneously isointense with respect to extraocular muscles on T1-weighted images and hypointense relative to the orbital fat on T2-weighted images, enhancing slightly with gadolinium-diethylenetriamine penta-acetic acid [4]. Takayama et al. had reported the granular cell tumor of the intradural extramedullary space of the spine. The tumor was hypointense compared with the spinal cord on T1-weighted images and slightly hyperintense on T2-weighted images, with homogeneous enhancement on post-contrast T1-weighted images. The tumor was hypointense both on T1 and T2-weighted images compared with fat tissue [5]. Maki et al. had reported MRI appearance of granular cell tumor of the breast, the tumor was isointense relative to skeletal muscle on T1-weighted images, hypointense on T2-weighted and IR sequences, and it showed generally homogeneous internal enhancement following gadolinium administration [7].

Similar to all these case reports, in our case the tumor of vulva was hypointense on T1-weighted images compared with fat tissue and after contrast material administration, the tumor showed markedly diffuse enhancement from early arterial phase until late venous phase. The vulvar tumor was isointense with muscles on T1-weighted images similar to Hashimoto et al. and Maki et al reports. On T2-weighted images the tumor was hypointense compared with adjacent fat tissue and slightly iso-hyperintense compared with skeletal muscles, similar to the cases of Hashimoto et al. and Takayama et al. In this age group, primary tumors of the vulvar region are cystic lesions, sebaceous cysts, papilloma, lipoma and fibroma. Since it showed diffuse intense enhancement, malignant fibrous and muscular tumors should also be kept in mind. GCTs are uncommon tumors, may occur at any part of the body including the vulvar region and generally seen hypointense on T1 and T2-weighted images compared with fat tissue, mostly isointense with muscles on T1-weighted images and show marked enhancement after contrast administration.

Conclusion

In conclusion, we report the imaging findings of the GCT of the vulva. Although it is rare, in patients with homogenous nodular subcutaneous masses in and around vulva, these tumors should be considered in the differential diagnosis.

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