



Abrikossoff's Tumor of the Vulva, Case Report and Literature Review

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Abstract

Abrikossoff's tumor or granular cell tumor is a rare soft and usually benign tissue neoplasm of neural origin. It occurs usually in the neck region or in the head. About 16% occur in the vulva. Some malignant cases have been reported in literature. Granular cell tumor was described first by Abrikossoff in 1926. The histogenesis of granular cell tumors is unknown. The clinical diagnosis of vulvar granular cell tumor is difficult. It may be mistaken for Bartholin gland cyst, lipoma or vulvar carcinoma when ulcerated. Histology is the mainstay of the positive diagnosis. Imagery is not essential for the diagnosis of Abrikossoff's tumor. It is asked when the neoplasm is giant, or when there is a local recurrence or potential metastasis. Surgery is the gold standard treatment of Abrikossoff's tumor. It consists of local excision with wide margins. Prognosis of granular cell tumor is usually good. Authors report a 25-year-old woman presented with an ulcerate tumor on the labium majus. The objective of this paper is to review the clinical and therapeutic aspects of Abrikossoff's tumor located in the vulva from case report and literature review.

Subject Areas

Gynecology & Obstetrics, Oncology, Women's Health

Keywords

Vulva, Vulvar Diseases, Vulvar Neoplasms, Granular Cell Tumor, Abrikossoff's Tumor

1. Introduction

Abrikossoff's tumor also known as granular cell tumor is a rare neoplastic skin and soft tissue neoplasm of neural origin. It usually occurs in the head or the

neck region. Five to sixteen percent occur in the vulva. Histology is important for the diagnosis. This tumor is generally benign, solitary and small nodule palpable on the skin surface. Abrikossoff's tumor in the perineal area is reported at vulva, cervix or ovary level. Only 1 to 3% of granular cell tumor is malignant with local infiltration, recurrences and visceral metastasis. Although uncommon, Abrikossoff's tumor of the vulva should be included in the differential diagnosis of nodular vulvar lesions and malignant neoplasms especially when ulcerated. The treatment of choice is large surgical excision. When the tumor is not completely removed, there is a high risk of local recurrence [1] [2] [3]. We report the case of a 25-year-old patient who presented with an ulcerated tumor of the vulva. The aim of this article is to report medical history and treatment of a granular cell tumor of the vulva in a young woman and literature review.

2. Case Report

A 25-year-old black woman presented with a large and painful tumor on her left labium majus, which appeared two years ago. During her last pregnancy, she noticed an increase in the size of the lesion and a surface ulceration as well as developed three another new lesions, two on the left labium majus and one on the right. Ulceration and pain motivated the young woman to consult the Obstetrics and Gynecology Department six month after delivery. Examination revealed a large ulcerated and indurated tumor of the vulva of 4×3 cm in diameter on the left labium majus. This tumor was associated to three skin colored subcutaneous indurate and painless nodules (**Figure 1**).

Biopsy specimen of the ulcerated lesion established the diagnosis of an Abrikossoff's tumor of the vulva. The histologic appearance showed diffuse large ranges of cells with elongated and polygonal form, granular eosinophilic and abundant cytoplasm and a rounded or oval nucleus with small nucleolus. Cells did not show cytonuclear atypia (**Figure 2**).

The staging of this vulvar tumor by chest X-ray, thoracic and abdominal CT scan did not show any metastasis. The patient undergone a partial vulvectomy

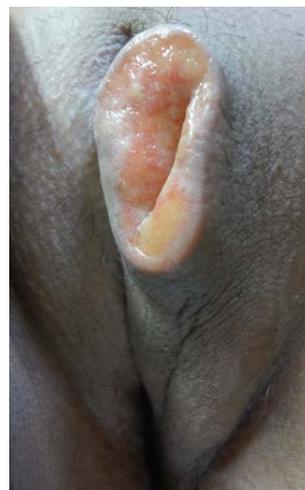


Figure 1. Ulcerated left vulvar tumor.

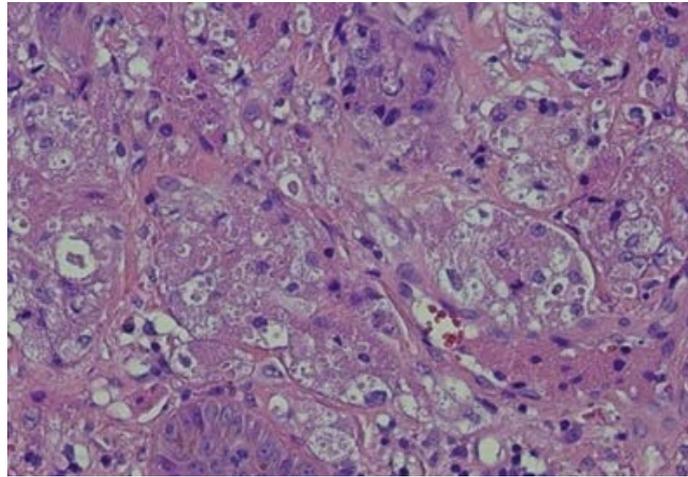


Figure 2. Polygonal cells with granular eosinophilic cytoplasm (HE \times 200).



Figure 3. Surgical tracing for partial vulvectomy.



Figure 4. Partial vulvectomy, final appearance.

(**Figure 3**) carrying all the lesions (**Figure 4**). After a follow-up of more than three years, the patient is doing well (**Figure 5**).



Figure 5. Appearance six months after surgery.

3. Discussion

Abrikossoff's tumor is usually solitary, subcutaneous or soft tissue lesion of various sizes appearing most frequently on the head, the neck, or the tongue, but it is also described anywhere on the body. Clinical presentation is usually of firm nodule. It occurs more frequently in adult black female [1] [2] [3] [4]. The size ranges frequently from 0.5 cm to 3 cm in diameter. Lesions may be multiple in 10%. Abrikossoff's tumor is generally slow growing and takes years to increase by 1 cm. As reported in our patient, it may increase rapidly in size during pregnancy. Granular cell tumor may occur in children or adult men. It is most common in the third and fourth decade of life. Benign forms of Abrikossoff's tumor are more common in young patients. This is particularly the case with our patient. Malignant forms are more common in elderly patients [1] [4] [5].

Histogenesis of Abrikossoff's tumor remains uncertain. Most authors emphasize the theory of neural origin. This hypothesis is strengthened by the immune-histochemical expression of S-100 protein, neuron-specific enolase, Leu7 (CD57) and myelin basic protein in granular cells [1].

Granular cell tumor presents with dermal or subcutaneous nodule that is firm, asymptomatic or little painful or pruritic and slow growing. It may be normochromic or reddish-brown, occasionally exhibiting ulceration or verrucous surface. Our patient had an ulcerated lesion and three subcutaneous nodules that appeared successively. The lesion is clinically unspecific. Diagnosis is based on histology [5]. Abrikossoff's tumor can occur anywhere on the body. More than half are found on the head and the neck area especially on the tongue. The most common perineal tumor occurs on the vulvar area. Between five and 16% of Abrikossoff's tumor occur in the vulva. Vulvar lesions appear usually on the labium, but other sites as perianal, perineal, clitoral or mons pubis have been reported [1] [3] [5]. The tumor usually presents as non-tender nodule and is typically smaller than 3 cm. It occurs mostly as a solitary mass, but in 3% to 16% lesions are numerous and synchronous.

Malignancy is rare. It occurs in 1% to 3% of cases. Metastases are reported in lung and brain. The predictive clinical features of malignant behavior are increased size tumor more than 5 cm, advanced age at diagnosis and local recurrence. The predictive histological features of high risk of malignancy are increased mitotic activity, tumor necrosis, large nucleoli, prominent spindling, high nuclear/cytoplasmic ratio and pleomorphism. The immune-histological criteria of malignant behavior are p53 expression and Ki67 positivity [1] [3] [4]. Our patient had no clinical or histological signs of malignant behavior.

Differential diagnosis in the vulvar region includes Bartholin gland cyst, lipoma, papilloma, hidradenoma, fibroma and vulvar carcinoma for ulcerated forms [6]. In our patient we suspected vulvar cancer.

Imagery is not necessary for the diagnosis of granular cell tumor. It is asked when tumor is giant or when there is a local recurrence or potential metastasis. Ultrasound shows a homogeneous mass often well limited. The mass is hypoechogenic or heterogeneous. CT scan showed a homogeneous and well-limited mass isodense to muscle, located in subcutaneous area [7].

Macroscopically, Abrikossoff's tumor of vulva presented with a growth in the perineal area varying in size from few millimeters to several centimeters, rarely larger than 4 cm in diameter. It develops in the dermis or subcutaneous tissue as a slow growing, non-tender, lump over months or years. The nodule has a color that varies from brownish to red, or it is covered by normal skin. The mass is mobile and the overlying skin may be depigmented, ulcerated or thickened with a "cobblestone" appearance. The tumor is poorly circumscribed with irregular margins and is yellow-gray and fleshy on cross section [1] [6]. Microscopically, granular cell tumors display oval or polygonal cells arranged in nests separated by bands of dense fibrous tissue. The tumor consists of irregularly clustered groups of large epithelioid-appearing cells separated by hyalinized stroma. The cytoplasm has a distinct granular appearance due to accumulation of lysosomes. Most lesions are well circumscribed. About 50% show poorly defined margins [5] [6]. The cells have small nuclei with abundant eosinophilic and coarse cytoplasm, containing Periodic Acid Schiff positive, diastase negative, resistant granules. These nuclei rarely contain mitotic figures [1]. The presence of necrosis, high mitotic activity, cellular pleomorphism with evident vesicular nuclei and nucleoli are indicators of aggressive behavior [4] [5].

Before planning treatment, clinicians should take a detailed history and do physical and other examinations especially chest X-ray, vaginal and abdominal sonography to exclude multicentric lesions [7]. For benign tumors, management consists of local excision with wide margins. In the event of positive margins, re-excision is recommended [1] [6]. In malignant granular cell tumors, histological margins should be carefully evaluated and repeated surgery with regional lymphadenectomy. Postoperative radiotherapy should be considered in cases where complete excision can't be achieved. However the effectiveness of radiotherapy or chemotherapy has not yet been established in malignant form of Abrikossoff's tumor [1] [4].

Once diagnosed with an Abrikossoff's tumor, patient must be counseled to follow up regularly with physical exams. She should alert her clinician if any growth recurs at the excision site or if any nodular growth develops elsewhere on the body [7]. Prognosis of vulvar granular cell tumor is usually good. Cases of recurrence are correlated to incomplete removal of the lesion [6]. Recurrence rates of granular cell tumors vary from 2% to 8% in benign lesions with clear margins and reach up to 20% with positive margins. Lung or brain metastases are reported in malignant forms [7].

Finally, Abrikossoff's tumor of the vulva is usually benign if reported in young female and if the lesion is smaller than 5 cm. When lesion is ulcerated, it is necessary to evoke the diagnosis of vulvar cancer and to do biopsies. Diagnosis is rectified by histological examination, which seeks signs of aggressive behavior. The treatment consists of performing surgical excision with wide margins. Follow-up must be extended because recurrences are possible also in benign forms.

4. Conclusion

Abrikossoff's tumor or granular cell tumor is a rare soft tissue neoplasm of neural origin. It usually occurs in the neck region. Up to 16% of cases occur in the vulva. This tumor is usually benign and asymptomatic. The clinical diagnosis of vulvar granular cell tumor is difficult. It may be mistaken for Bartholin gland cyst, lipoma or vulvar carcinoma. Abrikossoff's tumor should be included in the differential diagnosis of nodular vulvar lesions and neoplasms. Surgery is the gold standard treatment of granular cell tumors. It consists to do local excision with wide margins. The effectiveness of radiotherapy or chemotherapy for malignant forms has not yet been established. After surgery patient must follow up regularly with physical exams. The prognosis of granular cell tumors is mostly good. Recurrence is correlated to incomplete removal of tumor.

Informed Consent Was Obtained from Patient

The author reports no actual or potential conflict of interest in relation to this article.

References

- [1] Levavi, H., Gad Sabah, G., Kaplan, B., Yehezkel Tytiun, Y., Braslavsky, D. and Gutman, H. (2006) Granular Cell Tumor of the Vulva: Six New Cases. *Archives Gynecology Obstetrics*, **273**, 246-249. <https://doi.org/10.1007/s00404-005-0054-6>
- [2] Schmidt, O., Fleckenstein, G.H., Gunawan, B., Fuzesi, L. and Emons, G. (2003) Recurrent and Rapid Metastasis Formation of a granular Cell Tumor of the Vulva. *European Journal of Obstetrics Gynecology and Reproductive Biology*, **106**, 219-221. [https://doi.org/10.1016/S0301-2115\(02\)00165-3](https://doi.org/10.1016/S0301-2115(02)00165-3)
- [3] Horowitz, I.R., Copas, P. and Majmurdar, B. (1995) Granular Cell Tumors of the Vulva. *American Journal of Obstetrics and Gynecology*, **173**, 1710-1714. [https://doi.org/10.1016/0002-9378\(95\)90414-X](https://doi.org/10.1016/0002-9378(95)90414-X)
- [4] Ramos, P.C., Kapp, D.S., Longacre, T.A. and Teng, N.N.H. (2000) Malignant Granular Cell Tumor of the Vulva in a 17-Year-Old: Case Report and Literature Re-

view. *International Journal of Gynecological Cancer*, **10**, 429-434.

<https://doi.org/10.1046/j.1525-1438.2000.010005429.x>

- [5] Crivelin, L.L. and Milanez Morgado de Abreu, M.A. (2014) Abrikossoff's Tumor: Report of a Rare Tumor and Its Unusual Vulvar Location. *Jornal Brasileiro de Patologia e Medicina Laboratorial*, **50**, 371-373.
<https://doi.org/10.5935/1676-2444.20140043>
- [6] Rivlin, M.E., Meeks, G.R., Ghafar, M.A. and Lewin, J.R. (2013) Vulvar Granular Cell Tumor. *World Journal of Clinical Cases*, **16**, 149-151.
<https://doi.org/10.12998/wjcc.v1.i4.149>
- [7] Tawfiq, N., Sabri, S., Saiss, K., Bouchbika, Z., Benchakroun, N., Jouhadi, H., Sahrhoui, S. and Benider, A. (2013) Granular Cell Tumor: Report of a Complicated Vulvar Localization of Pulmonary Metastases. *Cancer Radiothérapie*, **17**, 671-674.
<https://doi.org/10.1016/j.canrad.2013.05.012>



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