Granular Cell Tumor - a Rare Tumor of the Mons Pubis: Case Report and Literature Review

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Abstract
Objective: Granular cell tumor of the mons pubis is rare. A case is reported with literature review.
Material and Method: Study of the management and outcome of a 23 year old Nigerian woman with granular cell tumor in the mons pubis. Literature review was done utilizing a Medline search for the last ten years.
Results: The diagnosis of granular cell tumor is often missed clinically and requires histological evidence. Adequate surgical excision, patient counseling and efficient follow up result in good outcome.
Conclusion: Clinicians and pathologist need to be aware of the clinico-pathological characteristics of granular cell tumors in order to facilitate its appropriate management.
Key Words: Granular Cell Tumor, Mons Pubis

Introduction.
Granular cell tumors of the mons pubis involve the skin and soft tissue and are uncommon, benign, solitary, nodular or ulcerative tumors. They are of neural origin and were first described by Virchow and Weber in 1854. Granular cell tumors are commoner in blacks than Caucasians and rare familial involvement has been reported [1]. The trunk and mucous membrane of the laryngiopharynx especially the tongue are the commonest sites in which granular cell tumors occur. Infrequent sites include the respiratory system, gastro-intestinal tract, brain, heart, and female reproductive [2]. Vulva tumors account for 5 16% of all granular cell tumors in the body and mostly involve the labia majora. Less common female lower genital tract sites include the mons pubis, clitoris, introitus, episiotomy scare and perineal body [3]. Granular cell tumors have been reported in patients between 6 70 years with a mean age of 38.1 years [2].

We report the first documented granular cell tumor in the female genitalia in our hospital which was established as a Nigerian government sponsored tertiary hospital 37 years ago.

Case Report
A 23 year old unemployed secondary school leaver was referred from the dermatological to the oncology clinic of the University of Nigeria Teaching Hospital. She noticed a vulva swelling about one year prior to presentation which persisted despite repeated application of native herbs. Her general physical examination was normal. A 2 × 3 cm firm, non tender, solitary, hyperkeratotic mass was in the right side of her mons pubis. It was not attached to the underlying tissue and the lymph nodes were normal. She defaulted from further evaluation but returned a month later with a shallow ulcer which had elevated edges in place of the solitary mass. The ulcer was non-tender, 2 × 2 cm size and not attached to the underlying structures (Figure 1). Her packed cell volume was 40%, she was negative to human immunodeficiency virus antibodies and her venereal disease research laboratory test was negative.

Extended excision of the ulcer with at least 2 cm of normal skin beyond the edges of the ulcer and involving the full thickness of the subcutaneous was done. Histological sections of the mass revealed a dermal proliferation of large cells with highly granular cytoplasm. The cells were disposed in sheets with poorly defined cytoplasmic borders; the nuclei were small, vesicular and uniform in size, while the cytoplasm was deeply eosinophilic. The

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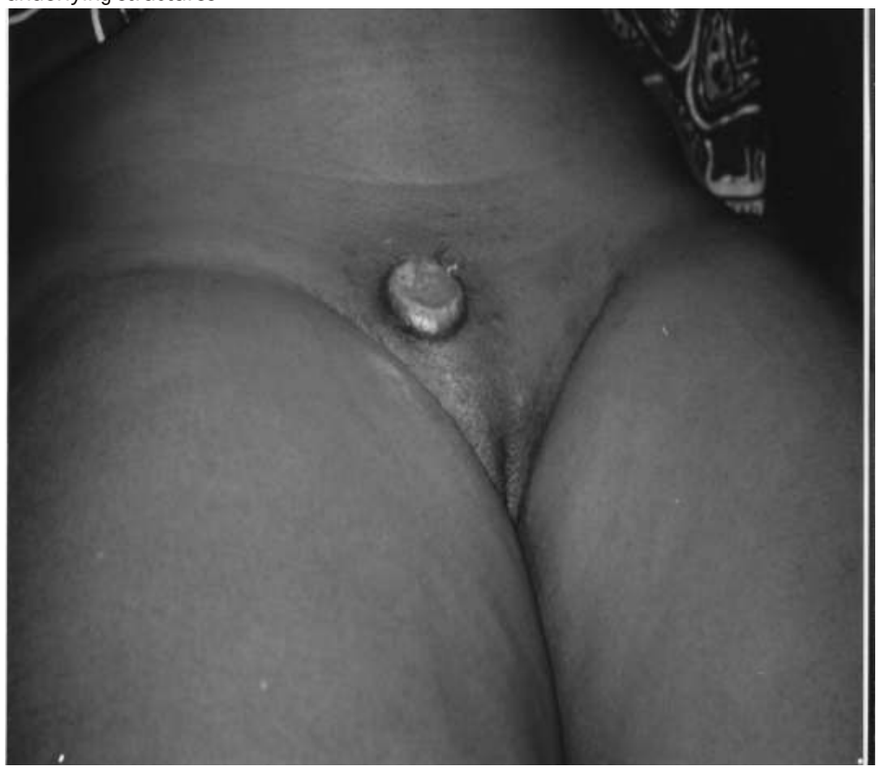
Figure 1. A shallow non-tender, 2 × 2 cm size ulcer with elevated edges not attached to the underlying structures



proliferation reached close to the epidermis which had areas of ulceration with accompanying acute inflammatory neutrophil infiltration and pseudo-epitheliomatous hyperplasia at the edges of the tumor. Necrosis within the tumor was absent, no mitosis was identified in the section and the edges of the sample were tumor free (Figure 2).

Her post operative condition was satisfactory. Subsequent follow up was discussed with her since the tumor was known to show local recurrence. In the past three years she has been well.

Discussion.

Granular cell tumor prior to the introduction of immuno-histochemical staining and electronic microscopy was thought to have originated from skeletal and smooth muscles, fibroblasts, histocytes and neurons at various times. They were consequently termed granular cell myoblastoma, granular cell schwannoma, granular cell neurofibroma, Abrikossoff tumor, Epulus of the new born, myoblastic myoma and granular cell neurogenic tumor. Their ultrastructure and immunochemical staining strongly suggest Schwann cell derivation [4].

Granular cell tumors are seldom diagnosed accurately clinically. The lesion in this case was mistaken for a sebaceous cyst and following ulceration resembled carcinoma of the vulvar. Sexually transmitted diseases and carcinoma of the vulvar are differential diagnosis of granular cell tumor ulcers. The solitary tumor in the mons pubis may be mistaken for sebaceous cysts, lipomas, hydrandenoma, fibroma and papilloma. Histologically granular cell tumors are round, polygonal or spindle shaped large cells with small hyperchromatic, slightly basophilic nuclei arranged centrally or eccentrically. The nucleolus of granular cell tumor is inconspicuous and the cytoplasm is abundant with coarse granules. The granules are probably of lysosmal origin and contain hydrolytic enzymes that stain

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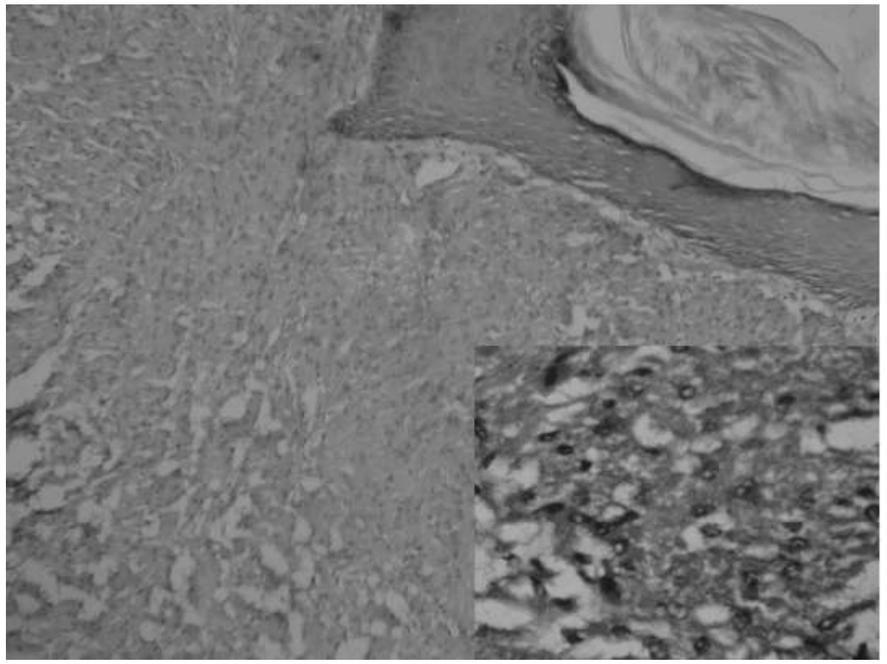
Figure 2 . Histological Section of the Granular Cell Tumor at Low and High Magnification



positive for luxol fast blue and periodic acid Schiff [3]. Similar granules are seen in amelioblastic fibroma, Wallerian degeration in severed nerves, traumatized muscles, lieomyosarcoma, angiosarcoma and appendiceal granular cell lesion [5]. Variants of granular cell tumor show mild pleomorphism, vesicular nuclei and distinctive nucleoli. The tumor is not encapsulated and the cells are arranged in irregular bundles with the tendency to infiltrate surrounding tissues. Mitosis is rare except when malignant. Fibrosis or desmoplasia is varied and pseudoepitheliomatosus hyperplasia in overlying squamous epithelia appears like squamous cell carcinoma [8].

The histological difference between malignant and benign granular cell tumor is not clearly distinct. The diagnostic criteria for malignancy include clinical and pathological features. Rapid growth of tumor, tumor size of 4cm or more, older patients and short excision recurrence interval are important clinical features suggestive of malignancy and potentially malignant disease (aggressive behavior) [3,6,7]. Histologically malignant tumors and those with aggressive behavior show increased mitotic activity with more than two mitotic figures per high power field, increased nucleo-cytoplasmic ratio, vesicular nuclear, large nucleoli, marker ki 67 (clone ki-67 poly, Dako) values of more than 10% and presence of p 53 on immuno-staining [6].

The excised tumor margins need to be carefully examined to ensure that they are not infiltrated and where infiltrated a wider re-excision is necessitated. Local recurrence within 2 years in benign granular cell tumors often results from inadequate excision and tumors with cells arranged as ill defined advancing edges with marked infiltration rather than nodular, distinct edges [2,8,9]. Wide excision done in this case is adequate for diagnosis and treatment of benign granular cell tumors. The use of pre-operative needle aspiration to define the tumor histologically has previously been suggested [10]. This procedure is probably limited by the inadequacy of such biopsy samples to clearly discriminate between benign and malignant tumors.

Adjuvant chemotherapy is ineffective in granular

cell tumor but radiotherapy is being tried for malignant granular cell tumors. Benign granular cell tumors of the mons pubis have good prognosis except for their tendency to local recurrence. Mortality results from multicentric disease or recurrence affecting vital organ and malignancy. Multicentric tumors and malignancy are reported in 3 20% of cases [3]. When local recurrence occurs in benign tumor, sonography is essential to exclude unsuspected upper genital tract, lymphatic and hepatic deposits. Chest X-ray, CT scan and bone scintigraphy are also done to exclude chest, abdominal and bone lesions.

The awareness of the clinico- pathologic features of granular cell tumors of genital organs enhances prompt diagnosis, early treatment and effective follow up. These reduce the risk of morbidity and mortality in granular cell tumors.

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