CASE REPORT

Vulval Granular cell tumor: A rare entity

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ABSTRACT

Granular cell tumors are rare neoplasms of neural origin. The most common sites for granular cell tumors are the head and neck region; however, up to 15% occur in the vulvar area. The tumors are usually benign; however, malignant tumors have been reported. Although uncommon, ulcerative granular cell tumors should be included in the differential diagnosis of ulcerative vulvar lesions. We present a case of granular cell tumor in female patient who presented with ulcerative lesion in vulva

INTRODUCTION

Granular cell tumors are uncommon neoplasms of neural origin.[1] They occur throughout the body; only about 10% are found in the vulvar region. Granular cell tumors are most frequent in the third and fourth decade of life, the African-American race, and the female.[2-4] Generally, they appear as small, firm, painless, slow-growing subcutaneous nodules.[2] They are rarely encapsulated and recurrence is common.[5] The malignant granular cell tumor is rare, representing 1% to 2% of cases. This form is highly aggressive, unresponsive to treatment, and ultimately fatal. A third type of granular cell tumor has been described which has benign pathologic characteristics but behaves in a clinically malignant manner.[6,7] The treatment of choice for all types is wide, local surgical excision.8 We report a case of granular cell tumor with a clinically malignant behavior but with benign pathological features.

CASE REPORT

A 68-year-old female patient referred from gynecology clinic to our dermatology clinic in Forwaniya hospital for ulcerative lesion in the vulva since 8 months 4-year. The lesion was not painful, but the patient complained that her underwear would stick to the area. The patient gave a history of diabetes and hypertension that were controlled with oral hypoglycemic and antihypertensive (calcium channel blocker). Uterine hysterectomy was done for the patient 15 years ago for management of large fibroid. She had a history of genital Chlamydia and candidal infection. Laboratory tests were negative for gonorrhea, rapid plasma reagin, and HIV, and positive for Chlamydia and Candida, for which she was treated with azithromycin and floucanozole.

Clinical examination revealed solitary, painless, none tender ulcerative mass 4x4 cm in the vulva. The ulcer was covered with drayed pus and exudates (Fig. 1). There was no enlargement and tenderness of the inguinal lymph nodes. Differential diagnoses of squames cell carcinoma and chancriod was kept in mind for the lesion. Routine laboratory investigations including complete blood picture, lipids, electrolytes hepatic and renal profile as well as blood sugar revealed no abnormal

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Fig. 1 Solitary erythematous mass 3x3cm with ulceration on the vulva.

findings except mild hyperglycemia.

Punch biopsy from the edge of the lesion was done and histopathological examination showed ill defined dermal mass composed of polygonal cells with granular Eosinophilic cytoplasm and centrally rounded nuclei. The overlying epidermis revealed parakeratosis, acanthosis with elongation of rete ridges, also there are multiple squames eddies with central keratinization. No cells deformity or mitotic figures were detected. PAS stain revealed positive result for cytoplasmic granules of polygonal cells. Immunohistochemistry stains revealed positive results with S100 and vimentine but were negative for CD68 and chromogranine (Fig. 2 A,B,C,D). These histopathologic features were consistent with the diagnoses of granular cell tumor.

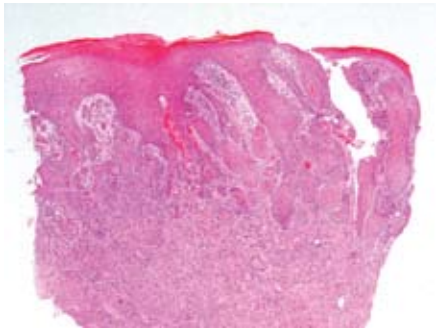

Fig. 2A Ill defined dermal infiltrate associated with epidermal hyperplasia.

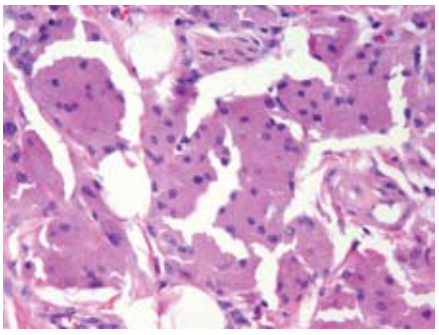

Fig. 2B The infiltrate composed of large polygonal cells with granular cytoplasm.

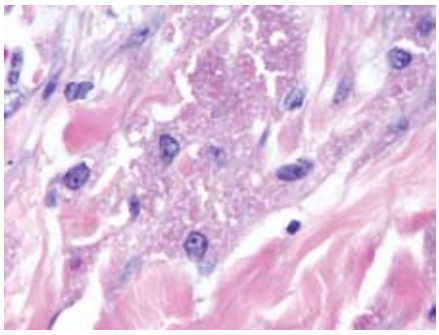

Fig. 2C The cells are PAS positive.

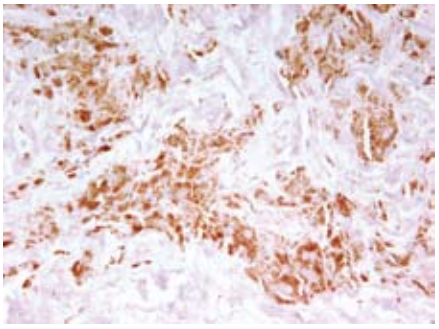

Fig. 2D The cells show strong positive staining for S100.

The patient was then scheduled for surgery, and a wide local excision of the tumor was performed and examined histopathologically. The area healed, and she is doing well postoperatively. The pathology report confirmed the granular cell tumor. Follow up the patient for one year revealed no recurrence or any changes at the site of the lesion.

DISCUSSION

Granular cell tumors are rare benign neoplasm nerve sheath origin. The tumors usually occur in third and fourth decade of life and may occur in children or adults.[1] They are also seen more often in females than males and more frequently in African Americans than Caucasians.[2] The female-to-male ratio is approximately 3 to 2. Although they can occur anywhere on the body, more than half are found in the head and neck area, especially the tongue. The tongue is involved in 25% of cases.[3] Our case is a female patient in seventh decade presented with granular cell tumor in vulva.

There are many sites was reported in literatures for Granular cell tumors such as the breast, gastrointestinal tract, cervix, lungs, parotid, penis, scrotum, peripheral nerves, skin, and vulva.[4] They have also been reported in the eyelid and appendix.[5] It is estimated that between 5% and 16% occur in the vulva.[6] Five percent of granular cell tumors are found in the gastrointestinal tract; if they are large enough there, they may cause symptoms of pressure or obstruction.[3]

Granular cell tumors usually present as nontender nodules and are typically smaller than 3cm. A nodular mass may be seen in the labium, clitoris, or mons pubis. Most of the time a solitary mass is seen, but in 10% to 15% of cases multiple tumors are noted.[2] our case was presented with ulcerative mass in the vulva.

There are case reports of one patient with 26 lesions and another with 52 lesions.[3] While the tumors may increase in size rapidly during pregnancy, they are generally slow growing and usually take years to increase by 1cm.[1] While most are asymptomatic, some symptoms may occur, including pain and pruritis.[7] Fibroma, sebaceous cyst, xanthogranuloma, basal and squames cell carcinoma are usually inter in the differential diagnosis of granular cell tumors.[2,4]

The granular cell of diagnostic necessity is a large polygonal, oval or bipolar cell with abundant, fine or coarsely granular eosinophilic cytoplasm, and a small, pale-staining or vesicular nucleus a centrically located in the cell. The cell membrane is moderately distinct, and some cells may contain large clumps of the granular cytoplasmic material, perhaps with clear haloes surrounding the clumps. Ultrastructural studies have described the cytoplasmic granules as autophagic vacuoles containing cellular debris, including mitochondria and fragmented endoplasmic reticulum, as well as myelin. Granular cells often occur in ribbons separated by fibrous septa, giving the appearance of infiltrating or "invading" into underlying tissues, especially muscle, with the bipolar shape being more frequently noted at the leading edge. The cells may also appear to be streaming off from or metaplastically arising from underlying muscle fibers. Older lesions tend to become desmoplastic with a few scattered nests of granular cells in a densely fibrotic background. Granular cells demonstrating nuclear enlargement, hyperchromatism and pleomorphism, or with mitotic activity or increased cellularity, are elements of the malignant variant of this tumor.[2]

The more oval granular cells near the surface tend to occur in broad sheets with minimal background stroma. They have a remarkable resemblance to macrophages and are S-100 protein positive with immunostaining. Immunohistochemistry will also be reactive for neuron-specific enolase (NSE), laminin and various myelin proteins. Staining is negative for neurofilament proteins and glial fibrillary acidic protein (GFAP). The interstitial cells stain for myelin protein.[7]

The malignant granular cell tumor has two distinct variations or subtypes. The first variant has a benign histopathology, not different from a typical granular cell tumor except for increased mitotic activity and mild nuclear pleomorphism. The clinical features of large size, rapid growth and surface ulceration must, therefore, be used to arrive at a malignant diagnosis, and the pathologist should carefully evaluate the lesional periphery for signs of true invasion. The second variant shows transition from typical benign granular cells to pleomorphic granular cells to pleomorphic nongranular spindle cells and giant cells with numerous mitotic figures. Malignant granular cell tumors are often negative for immunoreactivity for S-100 protein, NSE and vimentin.[3,7]

Granular cell tumors should be treated with wide local excision.[8] Recurrence rates are 2% to 8% in benign lesions with clear margins and 20% with positive margins.[3] Malignant tumors are seen in 1% to 2% of cases and are very aggressive; they may not be diagnosed until regional or distant metastasis occurs.[1-3] They may show necrosis, nuclear pleomorphism, spindling, and increased mitotic activity, or they may appear morphologically benign. The malignant tumors do not respond to radiation or chemotherapy; treatment is surgical.[3] The average age of malignant granular cell tumor patients is 50 year.[8]

All patients with granular cell tumors should be counseled to follow up regularly with physical examinations. They should inform the physicians if any growth recurs at the excision site or if any nodular growth develops elsewhere on the body.

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