CASE REPORT

Granular Cell Tumour of The Vulva: A Case Report

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ABSTRACT

Vulvar granular cell tumours are uncommon neoplasms of neural origin. The clinical presentations, surgical findings and histology of Granular Cell Tumour (GCT) of the vulva in a 50 year old lady is presented. Although rare, this benign lesion must be considered in the differential diagnosis of the labia majora masses, such as Bartholin,s duct cyst, Lipoma, Papilloma, Hydradenoma and Fibroma.

Kye Words: Granular Cell Tumour, Vulva, Rare

Introduction

Granular cell tumour (GCT) is a rare tumour of nerve sheath origin with a predilection for upper aerodigestive tract, skin and soft tissue. The neoplastic cells typically express S100 and CD68. The histogenesis of this tumour is still unknown. Some author however proposed about a Schwann cell derivation¹. Labial granular cell tumours, also known as granular cell myoblastomas, are small firm nodules located on the outside of the labia. They are most common in women aged 30 to 50, and are more prevalent in those with dark skin. The size of a labial granular cell tumour is usually small measuring about a half inch. It may occur on the surface or deep below the skin of the labia. This skin coloured tumour is usually benign and grows very slowly. Complete surgical excision is usually the cure, although tumours may recur.

Report of A Case

A 50 yr. old menopausal female presented a month ago with a small lump in her vulva since last 10 yrs. Since last two months it became itchy. There was no history of any systemic symptoms. Cutaneous examination revealed a skin coloured nodule in her left labium majus. It measured about 1.5 cm. at maximum dimension. The mass is well circumscribed, slightly tender to touch and firm to hard in consistency. An excision biopsy was performed under general anaesthesia. Her postoperative follow up was uneventful. On gross examination the lesion was seen adjacent to overlying skin, measuring 1.5X1X0.8 cm. The cut surface appeared homogenous light tan in colour. (Fig 1). Histopathology revealed irregularly arranged sheets and nests of large polygonal cells with a small hyperchromatic nucleus and abundant coarse eosinophilic granular cytoplasm. The overlying epidermis show pseudoepithelomatous hyperplasia. (Fig. 2).



Fig I: Cut Section of GCT

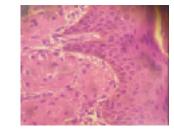


Fig I: Microscopic View (40X;H&E)

Discussion

Granular cell tumour (GCT) of vulva is a rare benign tumour of neurogenic origin resembling Schwann cells. Although it may occur in patients of all age groups, they are most frequently encountered between the third to sixth decades of life. Black females are twice as commonly affected than their Caucasian counterparts. Clinically GCT presents as small, firm, slow growing asymptomatic nodule less than 3 cm. in diameter. They are rarely encapsulated and recurrence is uncommon. In our case also the lesion was solitary and small in size but was slightly itchy and was tender to touch. Pruritus and pain have been reported occasionally with these lesions. Larger lesions may sometimes show surface ulceration, which may clinically give an impression of squamous cell carcinoma. Malignant granular cell tumour is rare, representing 1% to 2% cases¹. This form is slightly aggressive, unresponsive to treatment and ultimately fatal. Apart from histology, tumour size greater than 5 cm, vascular invasion, necrosis, and rapid growth are important indications of malignant behavior². A third type of granular cell tumour has been described which has benign pathologic characters but behaves in a clinically malignant manner. The treatment of choice for all types is wide local surgical excision. The case described here is a slow growing tumour and typifies the benign type granular cell tumour. There is absence of vascular invasion or necrosis. Cutaneous GCT may present with extension into the junctional region of the epidermis and thus may mimic melanocytic neoplasm³. No junctional activity is however seen in our case.

Though the tumour is thought to be of neural origin, a rare subset of distinctive cutaneous non neural granular cell tumours was also described and termed" Primitive polypoid granular cell tumour⁴. To describe the lesion as neural, schwannian or non-neural, immuno histochemistry is necessary which was not performed in our case. The neoplastic cells typically express S-100, CD68, NSE(Neuron specific enolase), Vimentin etc. As described by many authors⁵ distinction between benign and malignant GCT is difficult because of histologic similarity and lack of reliable criteria that can predict clinical behavior. Our patient is however under careful observation for any local recurrence or regional lymphadenopathy.

Clinically the differential diagnosis should include benign cystic lesions such as Bartholin gland tumours, Sebaceous cyst and other nodular benign painless vulvar lesions such as Dermatofibromas, Hidradenomas, Lipomas and Papillomas. GCT in the vulva is uncommon and vulvar involvement has been reported in 5-16% cases⁶. Laksmi et al⁷. reported a case of GCT on the clitoris of an 18 year old female, which was excised subsequently with no recurrence at six months follow up. Some of the authors advise a wide surgical excision of benign granular cell tumour regardless of positive or negative surgical margins⁸. Given the findings of current studies which show that these tumours rarely recurred regardless of surgical margin status⁹. To re excise a tumour even with positive margins should therefore be considered carefully because of the greater morbidity, compared with excision in other areas of the body. The surgical specimen in our case had negative margins and patient was advised for regular follow up only.

References

- 1. Leboit PE, Barr RJ, Burral S, Metcalf JS, Yen TS, Wick MR. Primitive polypoid granular cell tumour and other cutaneous granular cell neoplasms of apparent non neural origin. Am J Surg Pathol 1991 Jan; 15(1): 48-58.
- 2. Nagaraj PB, Ongole R, Bhujanga-Rao BR. Granular cell tumour of the tongue in a 6 year old girl: A case report. Med Oral Pathol Oral Cir Buccal 2006; 11: E 162-164
- Ray S, Jukic DM. Cutaneous granular cell tumour with epidermal involvement: A potential mimic of melanocytic neoplasia. J Cutan Pathol. 2007Feb; 34(2):188-194.

- 4. Lazar AJ, Fletcher CD. Primitive non-neural granular cell tumours of skin: Clinicopathologic analysis of 13 cases. Am J Surg Pathol. 2005 Jul; 29(7): 927-934.
- Le BH, Boyer PJ, Lewis JE, Kapadia SB. Granular cell tumour: immunohistochemical assessment of inhibin-alpha, protein gene product 9.5, S100 protein, CD68, and Ki-67 proliferative index with clinical correlation. Arch Pathol Lab Med. 2004 Jul;128(7): 771-775.
- 6. Cheewakriangkrai C, Sharma S,Deeb G,Lele S. A rare female genital tract tumour : benign granular cell tumour of the vulva: Case report and review of literature. Gynecol Oncol 2005; 97: 656-658.

- Laksmisha C, Thappa DM. Granular cell tumour of the clitoris. J Eur Acad Dermatol Venereol 2007; 21: 392-393.
- 8. Simon J, Schneider GT, Begneaud W, Harms K.Granular cell tumour of the vulva:literature review and case report. J La State Med Soc. 1996 Dec; 148(12): 539-541.
- 9. Barkan GA, Paulino AF. Are epidermal growth factor responsible for pseudoepitheliomatous hyperplasia associated with granular cell tumours ? Ann Diagn Pathol 2003 Apr; 7(2): 73-77.