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GRANULAR CELL TUMOR OF THE MONS PUBIS: A RARE SITE FOR A RARE TUMOR

Pathology

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ABSTRACT

Granular cell tumor (GCT) is an uncommon soft tissue neoplasm of Schwann cell origin, most often arising in the head and neck region. Genital involvement is rare and occurence of the mons pubis is exceptionally uncommon. Owing to its nonspecific clinical appearance, it is often mistaken for more common benign lesions such as sebaceous cysts or adnexal tumors. We report a case of a 51-year-old woman who presented with Granular cell tumor over the mons pubis.

KEYWORDS

Granular Cell Tumor, Mons Pubis

INTRODUCTION

Granular cell tumors (GCTs) are rare, typically benign neoplasms believed to originate from Schwann cells, characterized by large polygonal cells with eosinophilic, granular cytoplasm. Initially described by Abrikossoff in 1926, these tumors occur most frequently in the head and neck region, particularly the tongue, accounting for about 30% of cases[2]. However, GCTs have been reported in virtually every part of the body, including the skin, subcutaneous tissues, and internal organs.

Despite their widespread potential locations, GCTs are exceedingly uncommon in the genital area, with only a handful of cases involving the mons pubis documented in medical literature[1,3]. This rarity contributes to the low clinical suspicion for GCT in this anatomical location, often leading to misdiagnosis. Clinically, GCTs can mimic benign lesions such as sebaceous cysts or lipomas due to their superficial and often unremarkable presentation. This lack of distinct clinical features necessitates histopathological examination for accurate diagnosis. Here, we report a case of a granular cell tumor located in the mons pubis, clinically diagnosed as a sebaceous cyst. This report highlights the diagnostic challenges posed by this unusual presentation and the critical role of histopathology in identifying GCT in atypical locations.

Case Presentation

A 51 year old female presented with a gradually enlarging mass in the mons pubis. The excised specimen was sent to histopathology with a clinical diagnosis of sebaceous cyst. The lesion was skin covered and measured 3.5cm in greatest dimension. Cut section showed a pale white irregular lesion.

Histopathology showed skin with an underlying ill defined neoplasm in the dermis, composed of cells arranged in sheets and trabeculae, separated by thin fibrous bands and thin blood vessels. The cells were polygonal with abundant eosinophilic granular cytoplasm with round to ovoid small vesicular nuclei. Mild anisonucleosis was noted. No areas of necrosis, mitosis or spindling of cells were identified (Figure 1 & 2).

Immunohistochemistry with S100 showed strong nuclear and cytoplasmic positivity in the neoplastic cells (Image 3).

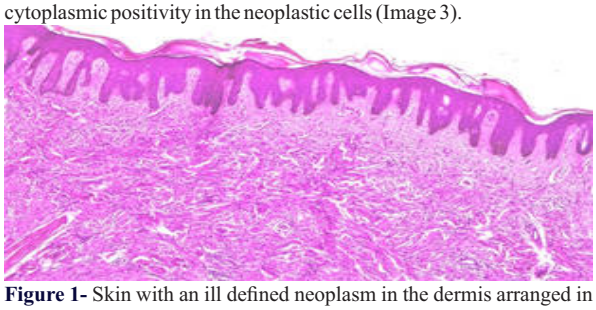

Figure 1- Skin with an ill defined neoplasm in the dermis arranged in sheets and trabeculae.

A diagnosis of Granular cell tumor was made. The patient was kept on follow up and he is asymptomatic until date.

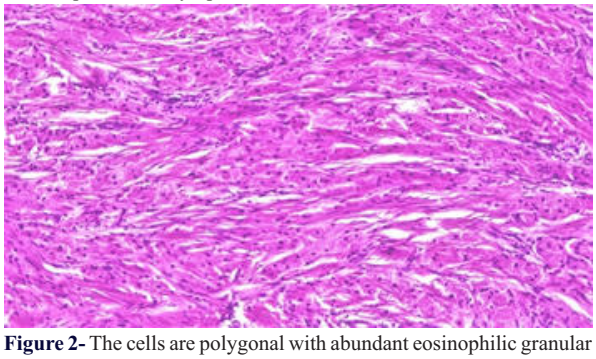

Figure 2- The cells are polygonal with abundant eosinophilic granular cytoplasm with small round to ovoid vesicular nuclei.

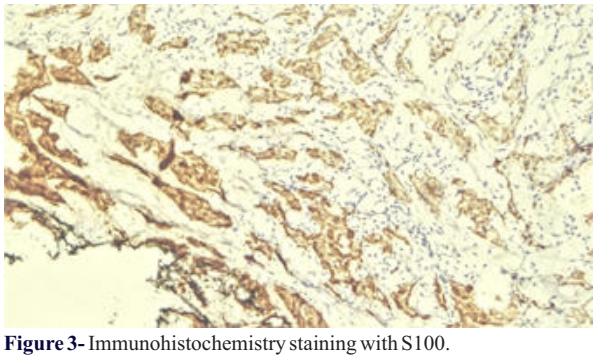

Figure 3- Immunohistochemistry staining with S100.

DISCUSSION

Granular cell tumors are rare soft tissue neoplasms with distinct histological features. Though they may occur almost anywhere in the body, the head and neck region remains the most common site of involvement, with approximately 30% of cases occurring in the oral cavity, particularly the tongue[2]. Cutaneous and subcutaneous granular cell tumors in the trunk, limbs, and extremities follow in frequency. The genital area, however, is an exceptionally rare site for these tumors and involvement of the mons pubis has only been documented in isolated case reports.

The atypical presentation of GCT in the mons pubis adds a diagnostic layer of complexity. Due to its benign clinical appearance, a GCT in this location is often mistaken for a more common superficial lesion such as a sebaceous cyst, lipoma, or other benign subcutaneous nodules like hidradenoma, fibroma and papilloma. In this case, the patient's lesion presented as a gradually enlarging, painless nodule, leading to a preliminary clinical diagnosis of a sebaceous cyst.

Histopathology remains the gold standard for diagnosing GCT.

Typical histological features include polygonal cells with granular eosinophilic cytoplasm, attributed to the abundance of lysosomes, and small, centrally located nuclei. The cells stain with Periodic Acid Schiff but are diastase resistant. Immunohistochemical staining is crucial, with GCTs consistently demonstrating S100 positivity, confirming their Schwann cell origin[4]. Other positive markers are SOX10, inhibin A, CD 68, neuron specific enolase, CD 56, EMA and vimentin.

The suggested malignant features include necrosis, tumor cell spindling, vesicular nuclei with large nucleoli, >2mitoses/ 10hpf, high nuclear to cytoplasmic ratio and pleomorphism.

The differential diagnosis for granular cell tumors includes other benign and malignant entities with granular cytoplasm, such as malignant granular cell tumors, atypical fibroxanthomas, and clear cell sarcomas. This highlights the importance of a thorough histopathological assessment to rule out malignancy, given that up to 2% of GCTs may exhibit malignant behavior. 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.

This case adds to the limited literature on granular cell tumors in the mons pubis[1], reinforcing the importance of awareness among clinicians. Granular cell tumors, while rare, should be considered in the differential diagnosis for nodular lesions in atypical locations, especially when the initial clinical impression is inconclusive or inconsistent with common benign lesions.

CONCLUSION

This case of a granular cell tumor in the mons pubis underscores the diagnostic challenge posed by the rarity of this tumor in the genital area and its benign clinical presentation mimicking a sebaceous cyst. Granular cell tumors should be considered in the differential diagnosis of nodular lesions in atypical locations, particularly when clinical suspicion is low, and routine diagnoses do not align with histopathological findings. This report highlights the critical role of histopathology in identifying unusual presentations of GCT and contributes to the limited body of knowledge regarding Granular cell tumor in the mons pubis.

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