Clinical Findings

A 56-year-old woman presented with an 8-month history of single, asymptomatic nodule over the neck, which was progressively increasing in size. The patient was systemically well. On examination, there was a single firm nodule of size 2.5 × 3.5 cm over the right lateral aspect of the neck. The nodule was dark brown in color and was fixed to the overlying skin. The lesion had a lobulated appearance with a zone of erythema at the periphery [Figure 1]. An excision biopsy was performed to reach a diagnosis.

Histopathologic Findings

Hematoxylin and eosin stain of the tissue showed diffuse infiltration of the dermis with uniform looking cells. The cells had centrally located nuclei and granular eosinophilic cytoplasm. There was no significant nuclear atypia or mitotic activity [Figure 2a and b]. The excised margin was free from tumor. Immunohistochemistry with S100 revealed a strong positive staining [Figure 2c].

Question

What is your diagnosis?
**Answer**

Granular cell tumor.

**Discussion**

Granular cell tumor (GCT) is a rare neoplasm that originates from Schwann cells and is a slowly growing, usually solitary, asymptomatic, smooth-surfaced nodule located in submucosal, intradermal, or subcutaneous region. It is commonly seen during the 3rd to 5th decades of life but can occur at any age with a female preponderance. GCT most commonly affects the head and neck region, which accounts for 45% to 65% of all sites affected by tumor. Of this, 70% are located in the oral cavity, especially the tongue, buccal mucosa and hard palate. Other commonly affected areas are skin and subcutaneous tissues (30%), breast (15%), respiratory (10%) or gastrointestinal tracts, and the biliary system.[1] Multifocal involvement has been reported in 10–25% of the cases.[3] Multiple GCT can occur as an isolated disorder or might be part of the LEOPARD syndrome, Noonan syndrome, neurofibromatosis, and Watson syndrome.

Histologically, it is characterized by large polyhedral cells containing mildly eosinophilic, uniform fine to coarse granules. This granular appearance is the result of dense cytoplasmic lysosomes. These granules stain positively with Periodic acid-Schiff (PAS) and are diastase resistant. Occasionally, spread along the peripheral nerves and vascular channels or infiltration of arrector pili can be seen.[3] On immunohistochemical analysis, GCT shows positivity to a broad panel of antibodies. S100 and CD68 are most commonly used immunohistochemical stains, other positive markers include neuron-specific enolase, nerve growth factor receptor 5, inhibin alpha, protein gene product 9.5, p75, vimentin, calretinin, and microphthalmia-associated transcription factor.[4]

Clinically, various benign tumors including lipoma, dermatofibroma, xanthogranuloma, leiomyoma, neurofibroma, adnexal tumors, and schwannoma can mimic GCT, but they can be differentiated by histopathological and immunohistochemical features.[5]

GCT is usually a benign tumor, occasionally it may be locally aggressive. Less than 2% are malignant and are associated with a poor prognosis.[5] The most common metastatic sites are regional lymph nodes, lungs, liver, and bones. Surgical excision of the tumor remains the mainstay of therapy.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**References**

1. Boulos R, Marsot-Dupuch K, De Saint-Maur P, Meyer B, Tran Ba Huy P. Granular cell tumor of the palate: A case report. AJNR Am J Neuroradiol 2002;23:850-4.
2. Deavers M, Guinee D, Koss MN, Travis WD. Granular cell tumors of the lung. Clinicopathologic study of 20 cases. Am J Surg Pathol 1995;19:627-35.
3. Battistella M, Cribier B, Feugeas JP, Roux J, Le Pelletier F, Pinquier L, et al. Cutaneous Histopathology Section of the French Society of Dermatology. Vascular invasion and other invasive features in granular cell tumours of the skin: A multicentre study of 119 cases. J Clin Pathol 2014;67:19-25.
4. Vered M, Carpenter WM, Buchner A. Granular cell tumor of the oral cavity: Updated immunohistochemical profile. J Oral Pathol Med 2009;38:150-9.
5. Daulatabad D, Grover C, Tanveer N, Bansal D. Granular cell tumor in a child: An uncommon cutaneous presentation. Indian Dermatol Online J 2016;7:390-2.