

Granular Cell Tumor on Perianal Region: A Case Report

Kesici Ugur¹, Mataraci Emine², Kesici Sevgi³, and Zeren Sezgin⁴

¹ Department of General Surgery, Ministry of Health, Akcaabat Hackali Baba State Hospital, Trabzon, Turkey

² Department of Pathology, Ministry of Health, Akcaabat Hackali Baba State Hospital, Trabzon, Turkey

³ Department of Anaesthesiology and Reanimation, Ministry of Health, Kanuni Training and Research Hospital, Trabzon, Turkey

⁴ Department of General Surgery, Ministry of Health, Numune Hospital, Sivas, Turkey

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Abstract- Granular cell tumor (GCT) was first described by Abrikossoff in 1926. GCT is a rarely seen soft tissue tumor and is generally benign. While the tumor can be seen in all parts of the body it is generally located on the head and neck region, and especially on the tongue. GCT is rarely seen in the anal-perianal region. In accordance with literature this case was reported because it was thought to be the 27th anal-perianal located GCT case. In this case report, approximately 0,5-1 cm pedunculated polypoid lesion was determined in the perianal region during the physical examination of a 23 year old female patient who applied with palpable mass complaint in the perianal region. Lesion in the patient was totally excised with healthy skin-subcutaneous tissue under local anesthesia. A benign granular cell tumor was detected in the histopathological examination. Positive staining was monitored immunohistochemically with S-100 and neuron specific enolase (NSE). GCT is a rarely seen tumor in the anal-perianal region and its malign transformation rate is very low. Even lesions seen in the perianal region have clinically a benign appearance, a histopathological examination should be conducted and also GCT should be kept in mind during diagnosis. Malign-benign separation of these lesions is difficult so histopathological examination should be conducted with great care. Large local excision in the treatment provides curative treatment. But for those presenting malign transformation further examination must be performed for metastasis. After the treatment local recurrence and metastasis should be considered carefully. Prognosis of metastatic disease is very bad.

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Keywords: Granular cell tumor; Soft tissue neoplasms; Perianal region

Introduction

Granular cell tumor (GCT) was first described by Abrikossoff in 1926 (1). GCT is a rarely seen soft tissue tumor and is generally benign (2). While the tumor can be seen in all parts of the body it is generally located on the head and neck region, and especially on the tongue (3). GCT is rarely seen in the anal-perianal region (4). In this study very rarely seen, a perianal located GCT case is discussed with literature.

Case Report

In this case report a 23-year-old female patient who applied with palpable mass complaint in the perianal region was discussed. There wasn't any additional disease in the patient's anamnesis. She stated that the mass in the perianal region existed there for 2 years and

recently caused itching. Approximately 0,5-1 cm sized pedunculated polypoid lesion was detected in the perianal region in the physical examination. Lesion in the patient was totally excised together with the healthy skin-subcutaneous tissue of the base under local anesthesia (Citanest Vial, 2%, 20 mg. AstraZeneca. Istanbul. Turkey). In the histopathological examination 1 cm deep skin-subcutaneous tissue with 0,7×0,3 cm sized skin ellipse on it and 0,2 cm diameter sinus on the skin was macroscopically detected. While no features were monitored microscopically in the epidermis, an infiltration formed by pyknotic, with nucleus, large crude granulated polygonal eosinophilic cytoplasm cells that develop nests in dermis, was observed. While staining was monitored in immunohistochemical examination in cytoplasmic granules with PAS (+), S-100 strong (+), Neuron Specific Enolase (NSE) (+), CD68 strong (+), Vimentin weak (+), staining was not

Corresponding Author: Kesici Ugur

The Ministry of Health Akcaabat Hackali Baba State Hospital, Trabzon, Turkey

Tel: +90 462 2213085, 5062398955, Fax: +90 462 2277786, E-mail: ugurkesici77@mynet.com

Granular cell tumor

monitored with Carcinoembryonic Antigen (CEA), Smooth Muscle Actin (SMA), Cytokeratins (CK). Granular cell tumor was histopathologically detected. Histopathological images are shown in Figure 1 (H&E. $\times 400$), Figure 2 (PAS (+). $\times 400$) and Figure 3 (S-100 (+). $\times 400$).

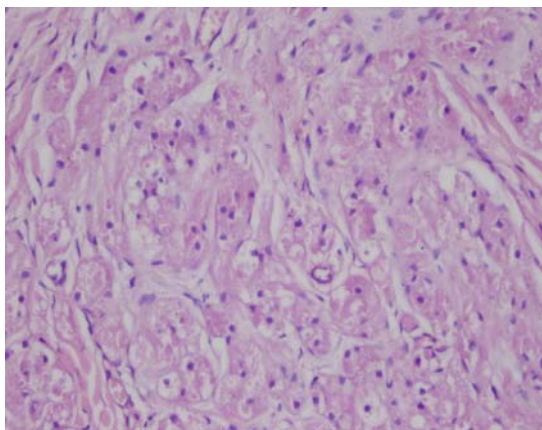


Figure 1. Granular Cell Tumor (H&E. $\times 400$).

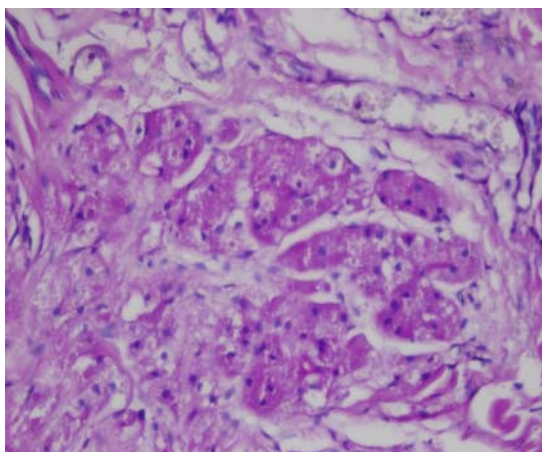


Figure 2. Granular Cell Tumor (PAS (+). $\times 400$).

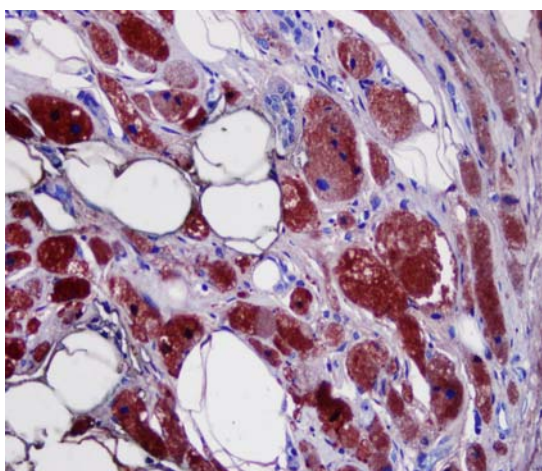


Figure 3. Granular Cell Tumor (S-100 (+). $\times 400$).

Discussion

GCT is generally asymptomatic and is seen between 4th - 6th decades (5). Although GCT was first described as a muscle based tumor, in present it is described as neural based tumor because it is immunohistochemically similar to nerve tissue. Positivity of S-100 protein and neuron specific enolase (NSE) is observed immunohistochemically in high percentages (3). In this case, positivity of NSE and S-100 protein was detected. Non-capsulated, fusiform, firm and dense content granular eosinophilic cytoplasm are histopathologically typical (6). But the tumor histogenesis is not completely known (7).

GCT is a rarely seen soft tissue tumor and is generally localized in the head-neck region. Lesions are generally small (0,5-3 cm) and solitary (4). GCT is seen in the gastrointestinal system and mostly in the esophageal (8). Anal-perianal localized GCT cases are very rarely seen. Case presentation in 2009 by Mistrangelo *et al.* (9) indicates 25 cases located in perianal and anal region in the literature and their case is the 26th case in the literature. In accordance with the information found in literature we consider that this case is the 27th GCT case located in the anal-perianal region. Although GCT is generally benign it has a 1-2% chance to be monitored as malign. Malign lesions mainly develop in lung, liver and bone metastasis with typical lymphatic and hematogenous spread (3,10). Approximately 50 malign GCT cases are reported in the literature (5). There are anal-perianal localized 3 reports with malign characteristics in literature stated by Bouraoui *et al.* (11), Mnasri *et al.* (2) and Francesco *et al.* (3). Because of the histological similarity and the deficiency of reliable clinical criteria, the separation between malign and benign GCT is extremely hard (7). Therefore great attention should be given during the histopathological examination of GCT and immunohistochemical examination should be executed.

Sufficient local excision is effective in all GCTs (malign/benign) for both diagnosis and treatment. In malign lesions a further examination (lymphatic/ hepatic sonography, thorax, abdomen and pelvic computed tomography, thoracic X-ray and bone scintigraphy) should be executed for metastasis (12). It should be monitored closely for lifelong with regard to recurrence in the post treatment period and metastasis in malign ones (5,13). In this case, the lesion was totally excised with healthy skin-subcutaneous tissue under local anesthesia. Further examination was not performed for metastasis because malign transformation wasn't

histopathologically detected. In the post treatment study performed by Fanburg-Smith *et al.* (14) local recurrence was stated to be 32% in approximately 2 years time in malign GCT cases. As for benign GCTs local recurrence rate was stated to be 2-8% if surgical margin is negative and over 20% if it is positive (5). Therefore it is extremely important to perform a large local excision with negative surgical margin during treatment.

The prognosis in malign GCTs is extremely bad. Because when diagnosis is established, lymph node and distant metastasis are already present in most of the malign lesions. Also over 50% metastasis and approximately 30-50% mortality are reported in 3 years in malign lesions (5,14,15). The treatment of metastatic disease isn't promising at all and chemotherapy/radiotherapy doesn't affect the prognosis positively (6). Therefore in cases presenting recurrent and lymph node metastasis, radical surgery (local excision + lymph node excision) may be attempted (12). In conclusion GCT is a tumor rarely seen in the anal-perianal region and malign transformation rate is very low. Even though lesions seen in the perianal region have clinically a benign appearance, a histopathological examination should be performed and also GCT should be kept in mind during the diagnosis. Malign-benign separation of these lesions is difficult so histopathological examination should be conducted with great care. Large local excision in the treatment provides curative treatment. But for those presenting malign transformation, further examination must be performed for metastasis. After the treatment local recurrence and metastasis should be considered carefully. Prognosis of metastatic disease is extremely bad.

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