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Case Report

Malignant granular cell tumor presenting at unusual site with rare histomorphology

Shruti Mahawar[1]*, Anoja Aparajita[1], Kanhu Charan Das[1]

[1]*Dept. of Histopathology, Apollo Hospital, Bhubaneswar, Odisha, India*

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ABSTRACT

The granular cell tumor (GCT) is are benign soft tissue tumor of neuroectodermal differentiation. Its likelihood of malignant transformation is bare minimal, and it seldom involves the anal canal. Herein, we report a case of anal canal with unusual histomorphology too. This report highlights the anomalous nature and site of the tumor, along with its peculiar histology. It also focuses on enhancing diagnostic awareness regarding the entity.

1. Introduction

The granular cell tumor are mostly benign soft tissue neoplasms of nerve sheath origin and are composed of polygonal cells with eosinophilic, granular cytoplasm. It can present at any site throughout the body; however, the gastrointestinal tract is infrequently involved, the anal canal being an exceedingly unusual site. Furthermore, only 0.5–2% of them undergo malignant transformation. In the present case, the cells resembled histiocytes, having clear foamy cytoplasm morphologically, posing a diagnostic challenge to both pathologists and surgeons. However, positive expression for S100 protein and neuron-specific enolase antigen by immunohistochemistry in tumor cells confirmed the diagnosis. Till now, only two cases of malignant granular cell tumor of the anal canal have been reported. But this is the first reported case, wherein the histomorphology was unusual too.

2. Case Summary

We report the case of a 53-year-old female who presented in the gastroenterology department with complaints of bleeding P/R since 1 month. Per rectal examination and sigmoidoscopy revealed a polypoidal lesion in the anal canal. (Figure 1). An excision biopsy of the polyp was sent to the histopathology department with a suspicion of squamous cell carcinoma. Histopathology revealed diffuse proliferation of polygonal cells having clear to granular cytoplasm, nuclear pleomorphism, a high nucleo-cytoplasmic ratio, vesicular nuclei, and prominent nucleoli. (Figure 2) Areas of sarcomatous differentiation and necrosis are also noted. (Figures 3 and 4) Immunohistochemically, tumor cells were positive for S-100 protein and neuron-specific enolase. (Figure 5 A,B) Mib count was approximately 30% in the hotspot areas. (Figure 6) On the basis of physical examination, histopathological, and immunohistochemical findings, a final diagnosis of malignant granular cell tumor was made. Although the neoplasm was excised completely, the patient didn't turn up for chemotherapy thereafter and was lost to follow-up.

* *Corresponding author*.
E-mail address: shrutivimi@gmail.com (S. Mahawar).

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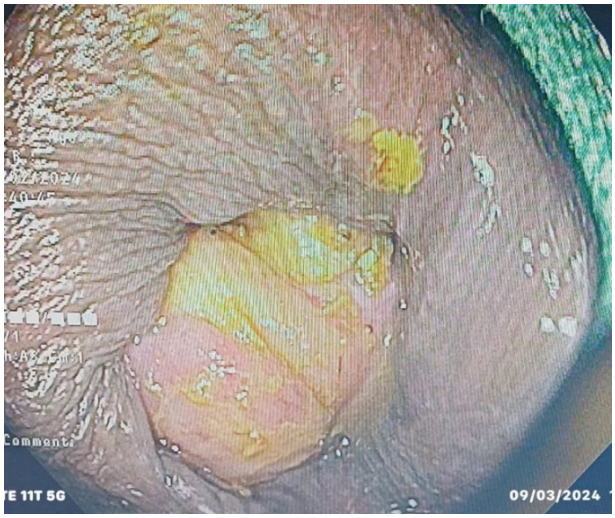

Figure 1: Anal polyp

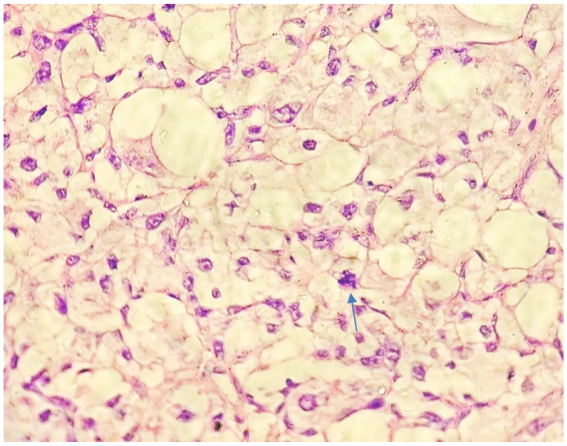

Figure 2: Sheets of granular cells displaying nuclear pleomorphism, prominent nucleoli and atypical mitosis (Green arrow). (H&E, 400x)

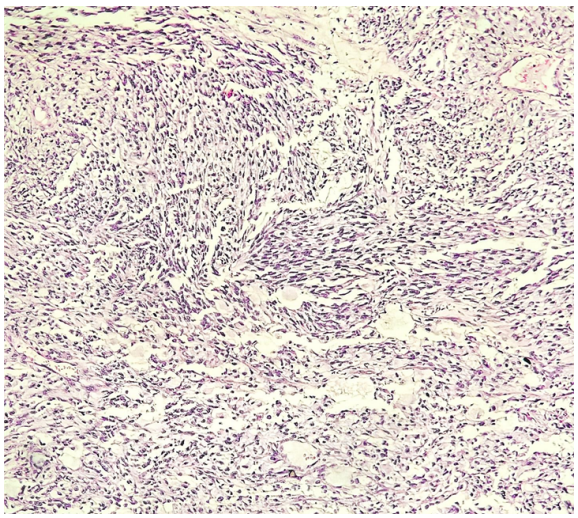

Figure 3: Areas of spindling (H&E, 40x)

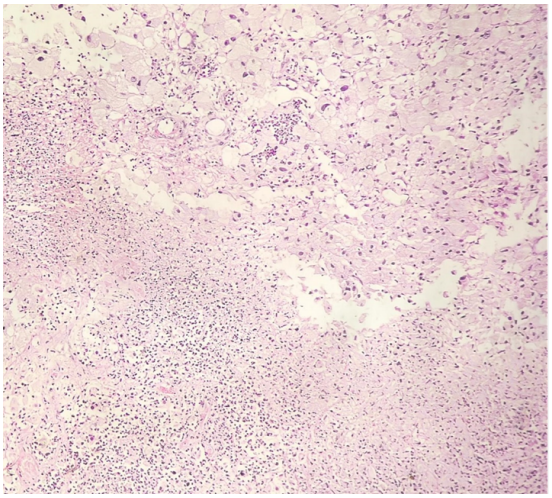

Figure 4: Areas of necrosis (H&E, 40x)

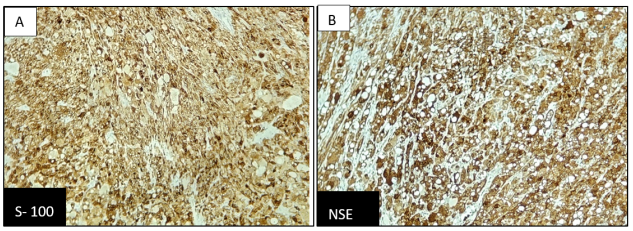

Figure 5: A, B): Strong granular positive (IHC, 100x)

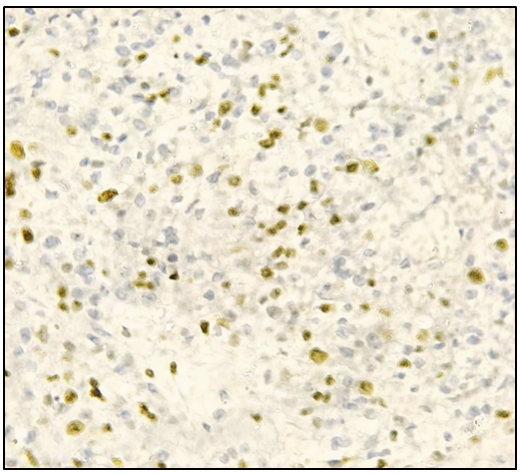

Figure 6: High Mib-1 index (IHC, 400x)

3. Discussion

Granular cell tumor is benign neoplasm of the neuroectodermal origin and comprises a large polygonal cell having coarse cytoplasmic granules within the cytoplasm. Ultrastructural studies demonstrate that these granules are lysosomes, which are found analogous to the lysosomes found within the Schwann cells that have ingested myelin.[1] It has no predilection for particular age or gender; however, females in their 4th to 6th decades are more predisposed,[2–5] which is consistent with our case. It can occur at any location with predilection for the head and neck region.[2,3] Its involvement of the gastrointestinal tract is infrequent, the anal canal being the least reported site. Bouraoui et al. al.[4] and Mnasri and Bouchoucha[5] were the first to describe a case of malignant GCT of the first to describe a case of malignant GCT of the anal region. After extensive research, no such case with a peculiar histomorphology has been found.

GCTs are usually benign tumor. Its malignant counterpart accounts for only 1–2% of GCT.[2] Presence of characteristic features like necrosis, spindling, high nucleo-cytoplasmic ratio, nuclear pleomorphism, vesicular nuclei with prominent nucleoli, and increased mitotic activity (>2 mitoses/10 HPF) indicates its malignant transformation.[6,7] Any three or more of these histopathological findings is regarded as a malignant GCT. Histomorphology of GCT shows the presence of large cells in sheets having granular cytoplasm, but in the present case, most of the cells show foamy to clear cytoplasm, mimicking histiocytes, making it even more challenging for the pathologist. Various histologic differentials with similar morphology, which include atypical fibroxanthoma, malignant fibrous histiocytoma, and malignant fibrous histiocytoma and malignant melanoma enters the scenario. These need to be excluded by immunohistochemical analysis. GCT shows diffuse granular positivity for S100 and NSE antigens immunohistochemically. The present case was strong granular positive for Vimentin, S-100, and NSE, whereas it was negative for Pan CK, p40, CD68, SMA, CD117, and Desmin ruling out other differentials.

A benign granular cell tumor has an excellent prognosis; as complete surgical resection is considered curative. On the other hand, malignant granular cell tumor tends to have poor prognosis, with 74% and 65% survival rates at 5 years and 10 years, respectively. They show a 32% to 41% recurrence rate and an 11 to 62% metastatic tendency between 3 and 37 months after diagnosis.[8,9] Complete excision with negative margins along with close colonoscopy surveillance and follow-up is recommended as standard protocol for both benign and malignant granular cell tumor.[10,11]

Henceforth, cognizance of the various differential diagnosis of malignant granular cell tumor and vigilant microscopic examination becomes the key factor for its accurate diagnosis and management.

4. Conclusion

Malignant transformation of granular cell tumor is rare and can occur at anal canal too. Its atypical morphology is a diagnostic challenge, and pathologists should be well aware of the entity and its differentials. A careful histopathological analysis and prompt communication with the clinician can render its recurrence or metastasis, hence increasing the disease-free survival rate.

5. Source of Funding

None

6. Conflict of Interest

None

References

1. Ordóñez NG. Granular cell tumor: a review and update. *Adv Anat Pathol*. 1999;6(4):186–203.
2. Lack EE, Worsham GF, Callihan MD, Crawford BE, Klappenbach S, Rowden G, et al. Granular cell tumor: a clinicopathologic study of 110 patients. *J Surg Oncol*. 1980;13(4):301–16.
3. Suchitra G, Tambekar KN. Gopal Abrikossoff's tumor of the tongue: Report of an uncommon lesion. *J Oral Maxillofac Pathol*. 2014;18(1):134–6.
4. Bouraoui S, Letaief H, Mestieri H, Chadly-Debbiche A, Zineb SB, Haouet S, et al. Malignant granular cell tumors. Report of a case of anal localization. *Ann Pathol*. 1999;19(2):151–2.
5. Mnasri H, Bouchoucha S. Granular cell tumor of the perianal region: Which therapeutic attitude? *Acta Chir Belg*. 2005;105(1):112–3.
6. Machado I, Cruz J, Lavernia J, Llombart-Bosch A. Solitary, multiple, benign, atypical, or malignant: the "Granular Cell Tumor" puzzle. *Virchows Arch*. 2016;468(5):527–38.
7. Collins BM, Jones AC. Multiple granular cell tumors of the oral cavity: report of a case and review of the literature. *J Oral Maxillofac Surg*. 1995;53(6):707–11.
8. Thacker MM, Humble SD, Mounasamy V, Temple HT. Case report. Granular cell tumors of extremities: comparison of benign and malignant variants. *Clin Orthop Relat Res*. 2007;455:267–73.
9. Moten AS, Zhao H, Wu H, Farma JM. Malignant granular cell tumor: Clinical features and long-term survival. *J Surg Oncol*. 2018;118(6):891–7.
10. Chilukuri S, Peterson SR. Granular cell tumor of the heel treated with Mohs technique. *Dermatol Surg*. 2004;30(7):1046–9.
11. Brown AC, Audisio RA. Regitnig Granular Cell tumour of the Breast. *Surg Oncol*. 2011;20(2):97–105.

Author's biography

Shruti Mahawar, Consultant

Anoja Aparajita, Senior Consultant

Kanhu Charan Das, Senior Consultant

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