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Research Article

GRANULAR CELL TUMOR OF APPENDIX: A 15 YEAR STUDY

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A R T I C L E I N F O

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A B S T R A C T

Background: Granular cell tumour (GrCTs) is a rare, benign soft tissue tumour that likely arises from Schwann cells. They are typically found in the tongue. Appendix is rarely affected, and it is most often found in middle-aged women, with a higher incidence among women of black ethnicity.

Material and methods: This study was a retrospective review of all appendectomy cases to identify granular cell tumors of appendix during a period of 15 years who presented to the department of pathology government medical college Srinagar from November 2004–October 2018.

Result: During a period of 15 years, three cases of granular cell tumor of appendix were seen. Two female and one was male patient was identified.

Conclusion: GrCT of appendix is a rare tumour and should be kept in differential diagnosis of sub-mucosal appendicular nodules.

INTRODUCTION

Granular cell tumour (GrCTs) is a rare, benign soft tissue tumour that likely arises from Schwann cells (1). Although it was initially classified as a myoblastoma, recent studies have shown that it is more likely to be neural in origin (2). GrCTs are typically found in the tongue or the dermal and subcutaneous regions (1). The appendix is rarely affected, and it is most often found in middle-aged women, with a higher incidence among women of black ethnicity (3). Approximately 0.5%–2.0% of GrCTs are reported as malignant (4). A malignant GrCT is aggressive and has a poor prognosis (5) whereas a benign lesion has excellent outcome after surgical resection (4). The presence of GrCT in appendix, manifests as a circumscribed submucosal nodule, and is often detected incidentally during endoscopy or surgical resection. Many of the GrCTs do not exceed 2 cm in diameter and the tumor does not infiltrate the muscularis propria (6).

MATERIAL AND METHODS

This study was a retrospective review of all appendectomy cases to identify granular cell tumors of appendix during a period of 15 years who presented to the department of pathology government medical college Srinagar from November 2004–October 2018. The cases were identified from the departmental data base and the corresponding medical records were traced. The histopathology slides were also traced and reviewed.

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RESULTS

During a period of 15 years, three cases of granular cell tumor of appendix were identified. Two were female and one was male patient (table-1).

Table 1 Clinical and Demographic details of patients

Patient demographics	Number of patients (n)
No. of patients	3
Gender	
Females	2
Males	1
Clinical presentation	
Acute appendicitis	2
Incidental mass	1
Size of tumor	
≤3cm	2
>3cm	1
Age	
42 yrs	1
51yrs	1
18 yrs	1
Gross finding	
Submucosal nodule	3

Two patients presented as acute appendicitis while as one patient had incidental finding of mass in right iliac fossa while being evaluated for some other cause.

On gross examination of the appendix, tumor size was upto 3cm in two cases while as one patient had less than 3cm sub-mucosal nodule in appendix. Microscopically the tumor was composed of large cells arranged in sheets having round to oval nucleus and abundant granular cytoplasm (fig-1). On IHC tumor cells were diffusely positive for S-100 (fig-2).

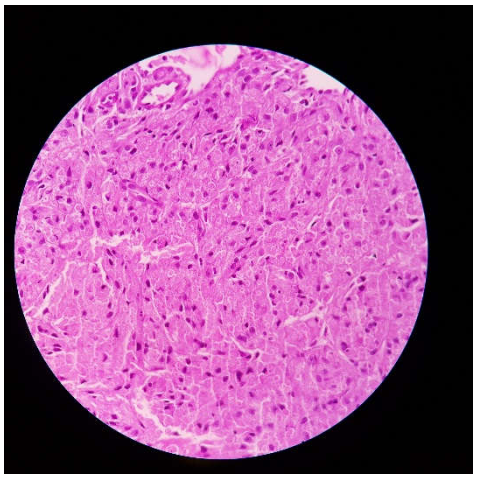

Fig 1 Tumor cells in syncytial pattern having granular cytoplasm.

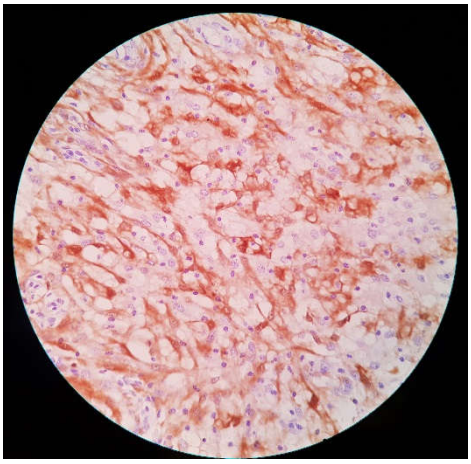

Fig 2 Granular cells revealing S- 100 positivity

DISCUSSION

GrCT was first described in 1854 by Weber (Weber & Virchow 1854) as a cluster of large cells featuring granular eosinophilic cytoplasm (7). Abrikosoff named these lesions as granular cell myoblastomas and he assumed, observing a group of five GrCTs located on the tongue, that GrCT had a striated muscle origin (8). In 1952,Feyrter renamed this form of tumors 'granular cell neuromas', pointing out the tendency of such lesions to affect peripheral nerves and presuming a perineural origin (9). The cell borders may appear indistinct, making it resemble a syncytium. Its benign status is further attested by the fact that no recurrences are reported, not even in lesions whose excision is incomplete (10). Histomorphologically granular cells may also be round, oval, polygonal or spindle-shaped and that the nuclei may be dark or vesicular,

being located in variable positions within the cell. The eosinophilic cytoplasm presents fine-to-coarse granularity. The phagolysosome granules contain large amounts of hydrolytic enzymes (such as acid phosphatase) and are strongly PAS-positive, diastase resistant and consistently positive for Luxol fast blue and myelin basic protein. The tumour cells are immunoreactive for S-100 protein, calretinin, NSE, laminin and CD68 (11), but they do not react with antibodies for neurofilaments or glial fibrillary acidic protein (3). GrCTs are commonly found in female patients aged 20–40 years (12). Embryologically, the appendix develops as part of the mid gut. In humans, it is a vestigial organ with glands forming simple tubes, which are often forked and secrete from 1 to 2 ml of mucinous fluid daily. The amount of lymphoid tissue varies with age: there is little in the foetal appendix, while it increases after birth and attains its maximum at puberty, thereafter gradually declining. During late middle life, little lymphoid tissue remains. This sequence of growth and atrophy of the appendiceal lymphoid tissue correlates with the age incidence of acute appendicitis, which is prevalent in young patients, and with the site of GCT occurrence (13).

CONCLUSION

GrCT of appendix is a rare tumour and should be kept in differential diagnosis if submucosal nodule is identified during gross examination of appendectomy specimen**.**

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