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Granular Cell Tumor of Esophagus: A Rare Case Report

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Abstract: Granular cell tumors are uncommon benign tumor of neurogenic origin. They frequently occur in skin and subcutaneous tissue but are found in gastrointestinal tract account to 6-10% of cases Granular cell tumor. Granular cell tumors originating in the esophagus account for 1% of all esophageal tumors. We present a case report of 58 year old male patient with granular cell tumor of esophagus that had dyspepsia. Histopathological examination confirmed the diagnosis of Granular cell tumor, which was further supported on immuno histochemical studies.

Keywords: Granular cell tumor, gastrointestinal tract, Esophagus, Histopathology.

INTRODUCTION

Granular cell tumors were first described in 1926 by Arbrikossoff [1]. They are rare benign soft tissue neoplasms. These rare tumors may arise in different organs but predominantly occur in the oral cavity, skin and breast tissue. Gastrointestinal tract granular cell tumor is very rare which accounts for 8% of all granular cell tumors with common site being esophagus [2]. Granulosa cell tumors accounts for 1% of all esophageal tumors [2, 3]. They originate from Schwann cell. Esophageal Granular cell tumor is common in 4^{th}, 5^{th} and 6^{th} decades with female predominance [3, 4]. Definitive diagnosis of granular cell tumor is by histopathology examination. We report a case of esophageal granular cell tumor in 58 year old male patient who presented with dyspepsia.

CASE REPORT

A 58 year old male patient presented to Medical gastroenterology department with complaints of dyspepsia for the past 2 years. Patient had history of hypertension and epilepsy since 7 years. Basic hematological investigations, liver function tests and renal function tests were normal. Upper esophagogastroduodenoscopy was performed for evaluation of cause of dyspepsia. Endoscopy revealed firm plaque sub mucosal lesion measuring 1cm and located in distal esophagus. Clinical diagnosis was esophageal malignancy with recurrent epilepsy. Patient had undergone endoscopic mucosal resection of the tumor and the specimen was sent to department of pathology for histopathological examination. Macroscopic findings showed two grey white bits altogether measuring 1.5x0.5cm. Microscopic examination showed lesion lined by stratified squamous epithelium. Subepithelium shows lesion composed of sheets and nests of tumor cells (Fig-1). Tumor cells are large polygonal with abundant granular eosinophilic cytoplasm and oval dark nuclei (Fig-2). No pleomorphism and mitosis were identified. Stroma shows congested blood vessels and fibrous tissue. Immunohistochemistry showed tumor cells, diffusely and strongly positive for S-100(Fig-3). Post-operative state was uneventful and 1 month follow up of the case showed good recovery.

DISCUSSION

Granular cell tumors are subcutaneous or sub mucosal tumors which are commonly located in tongue, skin and breast. The first five cases of granular cell tumor of the tongue reported by Abrikosoff designating them as myofibroblastomas [1]. Majority of esophageal granular cell tumors were seen in distal part of esophagus and remaining were fund in proximal part and middle respectively[5]. Majority of them are found incidentally during endoscopy, upper gastrointestinal study or autopsy. Patients with smaller than 1cm are often asymptomatic. Tumors larger than 1cm may present with dyspepsia. The histogenesis of the tumor is uncertain.

Authors suggest the origin of tumor like histiocytic, fibroblastic or primitive mesenchymal origin. Granular cell tumors were strongly positive for S-100 and Neuron Specific Enolase which support the recent evidence of neurogenic origin [6].

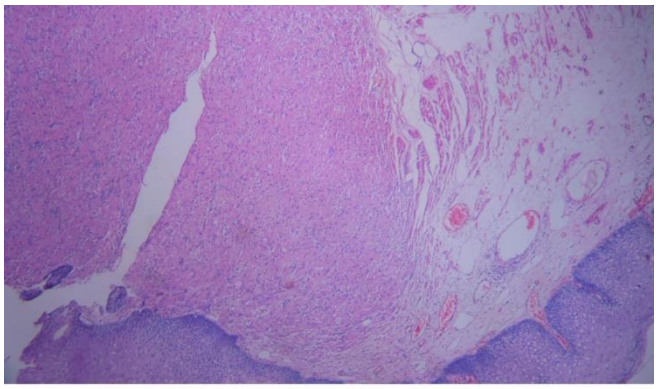

Fig.1: Microscopic examination shows tissue lined by stratified squamous epithelium. Subepithelium shows nests of polygonalcells with granular cytoplasm and round dark-nuclei. stroma shows congestedblood vessels.(scanner view).

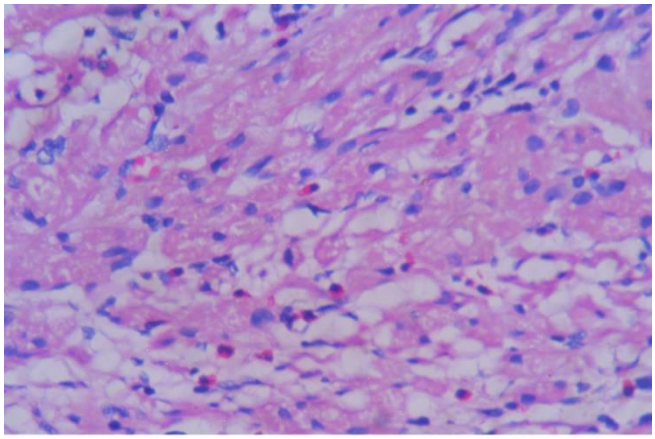

Fig. 2 : sections examined shows nests of polygonal cells with abudant eosinophilic cytoplasm and round nuclei (40X).

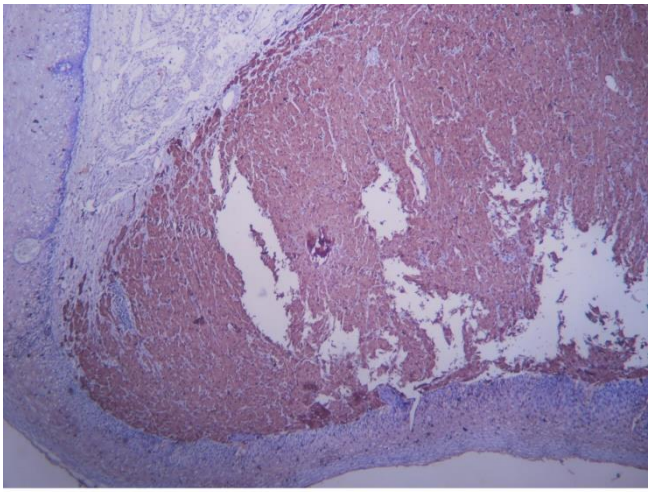

Fig-3: S-100 immunostain is diffusely and strongly positive for tumor cells(10X)

Grossly esophageal granular cell tumor resembles lesions like leiomyoma, lipoma, gastrointestinal stromal tumor and rhabdomyoma. Gastrointestinal stromal tumors are usually located in the submucosa and are rare in the esophagus. Most of the granular cell tumors discovered incidentally at the time of endoscopy, surgery or autopsy. Mostly in esophagus it arises distally. These tumors appear as small localized sessile, yellow white lesions measuring 1 to 4cm in diameter. Histologically tumors show cells arranged in nesting or acinar pattern. Cells were polygonal with dark ovoid nuclei and abundant eosinophilic granular cytoplasm [7].

IHC plays an important role in diagnosis of GCTS. According to literature Granular cell tumors shows immunohistochemical stains positive for S-100 ,CD68, NSE and vimetin whereas Negative for cytokeratin, EMA, Desmin and CD34(7) . Esophageal granular cell tumors are rare with few cases reported in the literature [8].

Esophageal leiomyomas found incidentally. Leiomyomas are typically pale, rubbery, lobulated, round to oval pinkish white lesion. Histologically these lesions are composed of spindle cells are arranged in fascicles and whorling pattern. Cells have elongated bland nuclei and eosinophilic cytoplasm whereas granular cell tumor shows absence of spindle cells with fascicles pattern.

Esophageal gastrointestinal stromal tumors are rare and most of them are malignant they present as intramural tumors or polyps. Microscopic examination of gastrointestinal stromal tumors shows spindle shaped cells arranged in fascicles and whorling pattern. The nuclei of tumor cells are cigar shaped with blunt pointed ends cells having abundant eosinophilic cytoplasm. Gastrointestinal stromal tumors show immunohistochemistry staining positive for CD117 and CD34 whereas granular cell tumors shows immunohistochemistry staining negative for CD117 and CD34.

Lipoma predominantly arises in large intestine and rarely seen in stomach and esophagus. They are sub mucosal lesions covered by intact mucosa. It consists of circumscribed sub mucosal lobular masses of mature adipose tissue with overlying intact mucosa whereas granular cell tumor shows absence of lobular adipose tissue.

Rhabdomyoma appear as intraluminal polyp or infiltrating lesion showing strap cells with esoinophilic cytoplasm along with myxoid matrix whereas granular cell tumor shows absence of strap cells.

CONCLUSION

Granular cell tumors of esophagus are rare benign tumor that have good prognosis. Recurrence metastases are rare. Endoscopic mucosal resection is a safe and accurate procedure. Histopathological examination was required to exclude other possibilities which mimic granular cell tumor by macroscopic appearance. Histopathological examination with Immunuhistochemistry helpful in diagnosis of these cases.

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