Granular Cell Tumor- Anonymity In Rarity

Dr. Shefali S*, Dr. Samriti G*, Dr. Rattan N**

*Resident,**Professor, Department Of Pathology, MM Institute Of Medical Sciences And Research, Mullana, Ambala-133203.

Abstract: Although an uncommon entity, granular cell tumours have been described in the skin, breast and tongue. The distal oesophagus is the commonest site of involvement by this tumour in the gastrointestinal tract. We report a case of an elderly female presenting with dysphagia who was found to have an esophageal lesion which on histopathology revealed a typical granular cell tumour. [S S Natl J Integr Res Med, 2020; 11(3):94-95]

Key Words: Granular cell tumour, Oesophagus, leiomyoma.

Author for correspondence: Dr. Shefali Setia, Department of Pathology, MM Institute of Medical Sciences and Research, Mullana, Ambala 133203 E-Mail: shefalisetia1992@gmail.com

Introduction: Oesophageal granular cell tumours are rare neoplasms comprising only about 1% of all oesophageal neoplasms[1].They can affect any age group and have a female predilection. Granular cell neoplasms were initially described by Abrikossoff in 1926[2] and were thought to be granular cell myoblastoma. It was in the year 1931 when oesophageal granular cell tumour was first reported. These tumours have a propensity to manifest at any site in the body primarily skin, tongue, breast tissue, oral cavity, respiratory tract and rarely gastrointestinal tract. They are thought to arise from the Schwann cells considering that immunohistochemically these tumours express S-100 protein in both diffuse and intense pattern[3].

Case Report: A 60 year old female patient presented with complaints of sore throat and difficulty in swallowing since 2 months. Onset was gradually progressive and more marked for solids than for liquids. She also complained of dyspepsia, cough with expectoration and vague upper abdominal pain. Patient is a known smoker since last 30 years.

Upper GI endoscopy showed two elevated whitish mucosal lesion at 30 cms from the upper incisors approximately 1cm in length and 4.5 mm in breadth. An endoscopic biopsy was performed and 2 small soft tissue bits collectively measuring 0.4x0.3x0.3 cm were sent for histopathological examination. Histopathological examination revealed an overlying hyperplastic squamous epithelial lining with a tumour in the lamina propria (Figure 1).

Composed of sheets, ribbons and nests of large polygonal spindle cells with intervening delicate fibroconnective tissue septa. The cells show abundant granular eosinophilic cytoplasm with bland, round to ovoid nuclei (Figure 2).

Figure 1: H & E (4X) Showing Hyperplastic Squamous Epithelial Lining

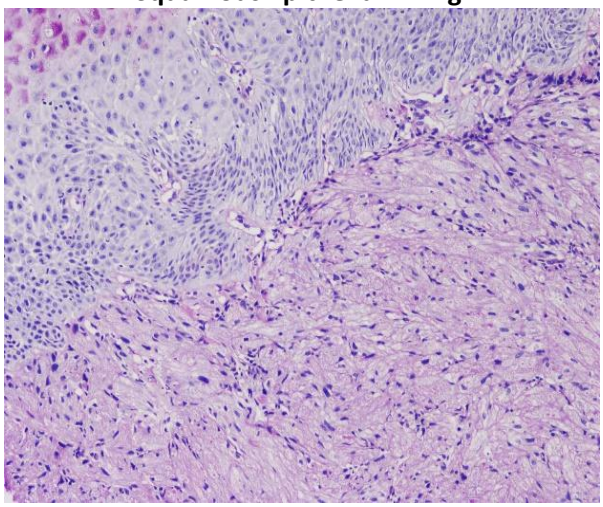
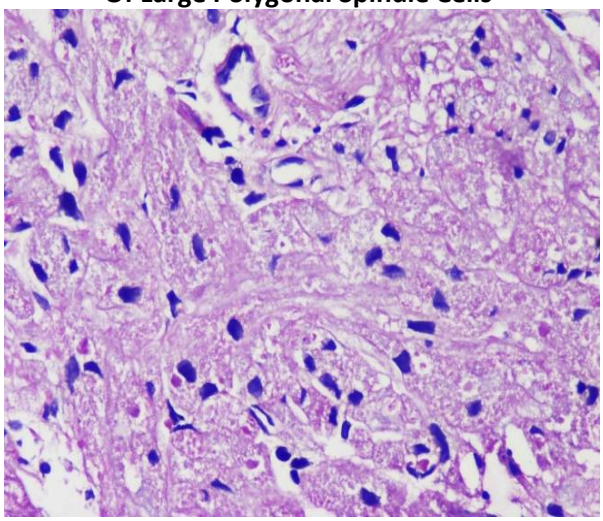
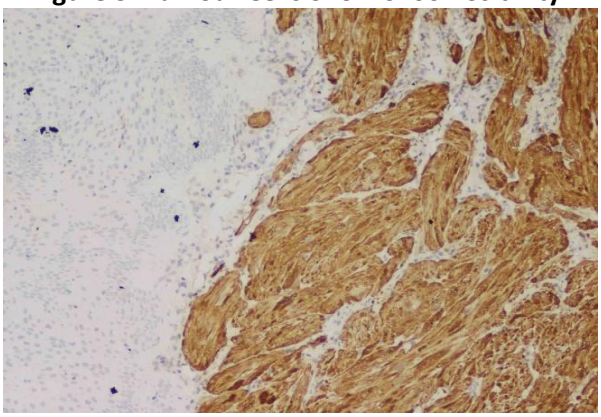


Figure 2: H & E (10X) Showing Sheets, Ribbons Of Large Polygonal Spindle Cells



The granules were fine to coarse which were PAS positive and diastase resistant and S100 positive (Figure 3). No infiltrative or pseudo invasive pattern was noted. No areas of necrosis, high mitotic activity, prominent nucleoli or spindling of nuclei were noted. Based on these microscopic findings a diagnosis of granular cell tumour of oesophagus was offered.

Figure 3: Tumour Cells Show S100 Positivity



Discussion: Granular cell tumours of the oesophagus are generally solitary and benign[4]. Besides the tongue, skin, breast, respiratory epithelium, biliary system approximately 1-8% of granular cell tumour occur in gastrointestinal tract[5]. Oesophageal granular cell tumour is the second most common oesophageal stromal tumour after leiomyoma[6]. These tumours are generally seen in the middle and lower third of the oesophagus. Smaller granular cell tumours are generally asymptomatic whereas larger tumours >1cm usually present with dysphagia and vague symptoms in the form of ill defined abdominal pain and dyspepsia.

The classification of oesophageal GCTs into benign and malignant has remained an enigma. Fanburg Smith[7] defined histological criteria for the distinction between benign and atypical malignant tumours[8] and they defined 6 histologic criteria for this purpose. The criteria adopted included increased nuclear cytoplasmic ratio, nuclear pleomorphism, vesicular nuclei and prominent nucleoli, tumour necrosis, spindling and increased mitotic activity (>2/HPF)[9]. Malignant neoplasms were those with >3 of these criteria whereas benign lesions had 0 criteria. Those in between meeting 1-2 criteria were classified as atypical[9]. Chen WS recommended surgical or endoscopic removal of those lesions >10mm,symptomatic and exhibiting rapid growth[10] **.**

Conclusion: Granular cell tumours are rare neoplasms of the oesophagus. Most granular cell tumours are in distal oesophagus. Histopathology is considered essential to see the extent of invasion and to rule out malignancy. Although, an endoscopic ultrasound and endoscopy aids in differential diagnosis, the final diagnosis of granular cell tumour rests on histopathological confirmation. While most granular cell tumour are regarded as benign tumours, some 1-3% show malignant features and therein lies the importance of subjecting the tissue for histopathological diagnosis.

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Conflict of interest: None
Funding: None
Cite this Article as: Setia S, G S, N R. Granular Cell Tumor-Anonymity In Rarity. Natl J Integr Res Med 2020; Vol.11(3): 94-95