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Case Report

Granular Cell Tumour – Esophagus Presented As Gastric Esophgeal Reflex Disease

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Abstract
Granular cell tumor of esophagus belongs to the category of soft tissue neoplasm and originates from Schwann cells. The tumor predominantly occurs in the oral cavity, breast tissue and skin. Esophageal origin is uncommon[1], only 8% cases has been reported; distal two third is the most common site. Endoscopic ultrasound imaging techniques give the important information about the depth of the tumor invasion. However confirmatory diagnosis is always made by histopathological examination of the specimen. A case of esophageal Granular cell tumor that was identified incidentally and removed by endoscopic mucosal resection.

Keywords: *Esophagus, granular cell tumors, Retrosternal pain.*

Introduction

The granular cell tumor was first reported by Abriksoosoff in 1926. It is uncommon and benign in nature[2]. Though skin and soft tissues are commonly involved the tumor may also occur in almost all over the body[2]. It can occur at any site, however more commonly in the skin, oral cavity and breast tissues are the preferred site. The origin of the granular cell tumor is from the neurogenic Schwann cells[1.] Usually the granular cell tumor involves the submucosal layer. Though the diagnosis is straight forward the appropriate treatment is often complex[2]. Initially surgical resection was recommended for the treatment but now a days endoscopic resectioning is recommended for relapse after resection is extremely rare either in situ or other locations[2]. However relapse seldom affects patient's life span.

Clinical History

38 year old male presented with complaint of difficulty in swallowing and retro sternal pain for which he underwent endoscopy examination.

Endoscopy revealed an elevated white coloured patch (leukoplakia) of approximately 1x0.5 cm's, which could be easily removed endoscopically without any untoward effect. The entire stomach from the fundus to the pyloric end as well as rest

of the esophagus are without any lesion on endoscopic examination.

Macroscopy

The sample was 5mm in diameter which was white in colour.

Microscopic Features

An endoscopic biopsy from the lesion was studied which revealed a fragmented biopsy material from the esophagus where squamous mucosa (Figure 1, 2, 3) and a tumor (Figure 1, 4) was identified. The tumor was made up of large polygonal cells with indistinct cytoplasmic border. The cytoplasm has granular appearance. The nuclei are dark hyperchromatic and condensed in nature with very mild degree of pleomorphism (Figure 5).

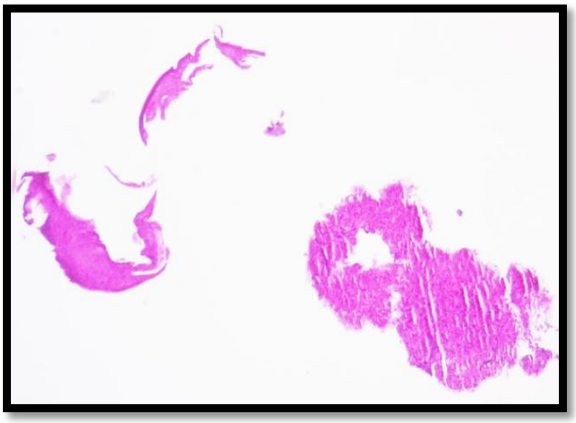

Figure 1: 4x strips of squamous epithelium of esophageal origin with tumor.

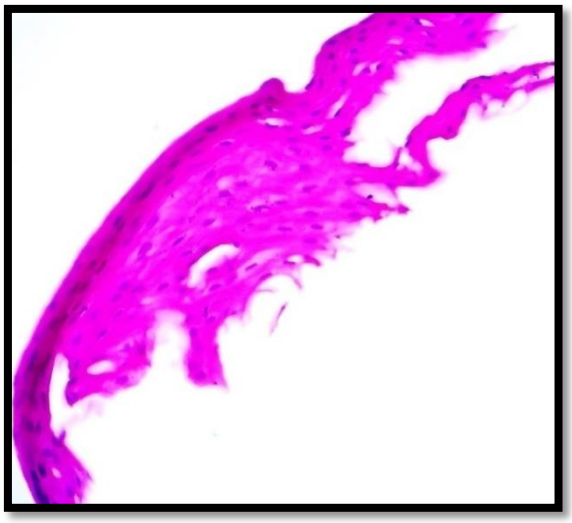

Figure 2: 10x strips of squamous epithelium of esophageal origin where nuclei are bland in appearance. There is an attempt to form keratinization.

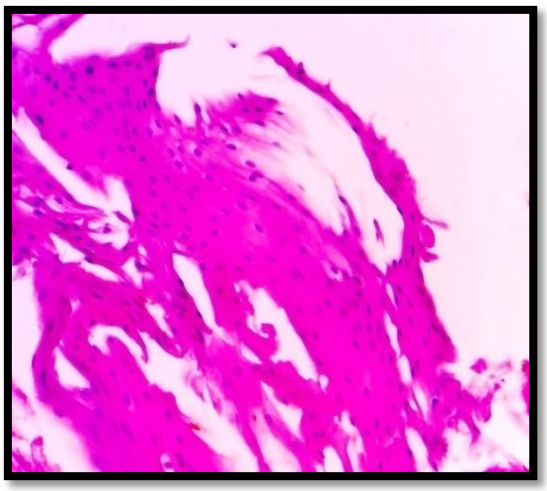

Figure 3: 10x Strips of squamous epithelium

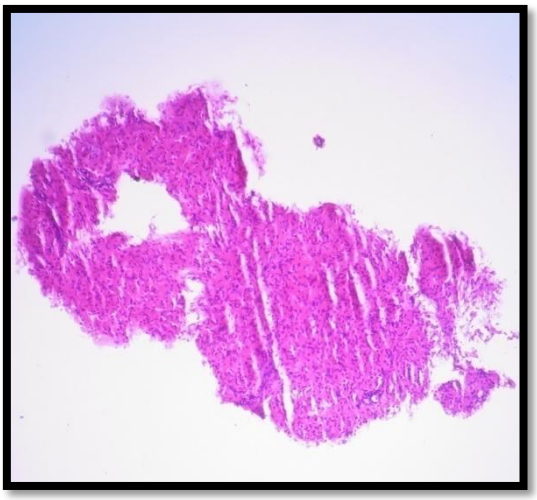

Figure 4: 10x Tumor with pinpoint nuclei and abundant cytoplasm

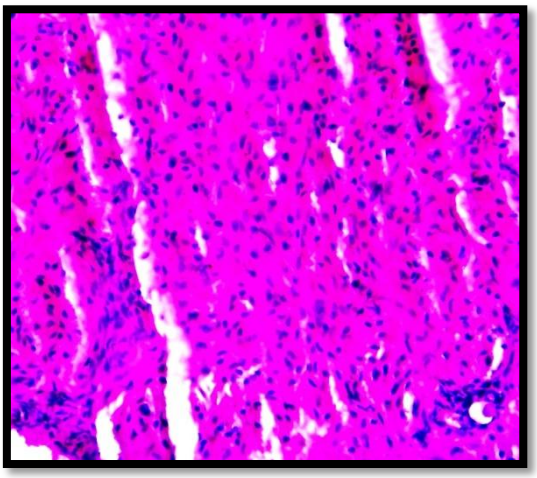

Figure 5: 20x Tumor with hyper chromatic pinpoint nuclei with abundant cytoplasm

Discussion

Granular cell tumor is a relatively uncommon one which originates from the Schwann cell of the neural tissue[1]. Esophageal granular cell tumor is

rare and with the introduction of EUS (esophageal ultra sound) and advances in endoscopic resection, these tumors are presently diagnosed and managed more easily[1]. The tumor can occur at any age group but more common at 40-60years; females are predominantly affected than males[2]. The histogenesis of granular cell tumor were debated for a long period of time whether it is neoplastic or degenerative origin. At present the tumor is widely accepted to be of neurogenic (Schwann cell) origin based on immunohistochemical (S-100 positive) and ultrastructural findings[3]. Accumulation of numerous lysosomes are the reason for granular nature of the cytoplasm of the tumor cells[3]. Usually it present as asymptomatic lesion and it is an incidental finding during the investigation of other diseases[1]. The symptoms such as dysphagia and acid reflux are present when the tumor size >1cm and multifocal in nature. Typically the lesions are sessile, solitary nodular white to gray elevated lesion which is morula like appearance, surface appears smooth, rarely the lesion were ulcerated and necrosis[1]. Though the lesion commonly involved in the submucosa it can also involve in the mucosa and muscularis propria. The lesion commonly occurs in the distal esophagus and rarely multiple lesions also identified in some patients. If the lesion involves muscularis propria it is difficult to differentiate from the condition called leiomyoma[3]. Tumor size usually range from few millimeter to 2 cm and 75% are presented with <2cm in diameter[3]. Endoscopic ultrasound is the only diagnostic tool which is available now a days because it diagnose the depth of the invasion and also assist the sampling for fine needle aspiration procedure[1]. Though endoscopic ultrasound is useful to differentiate the sub epithelial lesions, microscopic examination is definitive for the confirmatory diagnosis. The cells are usually round to polygonal in nature with round nuclei with granular eosinophilic cytoplasm and small pyknotic nuclei and pseudoepithelomatous hyperplasia of overlying squamous epithelium are the characteristic histopathological features. Immunohistochemistry analysis is very helpful when the histopathological diagnosis is difficult. Positive staining with s 100, PAS, nestin, vimentin and negative staining with markers like desmin, CD117, CD34, fibronectin, and carcinoembryonic antigen will assist the diagnosis. Rarely the lesion gets misdiagnosed as esophageal squamous cell carcinoma[1], when a squamous epithelium overlying the deep granular cells exhibit pseudoepithelomatous hyperplasia; granular cell tumor usually a benign nature, 2% of malignant cases were also reported[1]. Usually the malignant lesions are more than 15mm in length and has the features of nuclear fission, necrosis, a high mitotic index, and nuclear-cytoplasmic Ratio. Tumors fulfilling at least 3 of these criteria are classified as malignant according to Fanburg-Smith et al. The treatment modalities includes Yttrium-aluminum-garnet laser ablation, dehydrated alcohol injection, endoscopic resection, and surgical treatment[3]. Endoscopic resection with biopsy forceps may be effective only for tumors measuring < 2cm, as there is increased risk of an incomplete resection. Transthoracic excision of Granular cell tumor is the most definitive treatment and is indicated for tumors amenable to endoscopic resection. Recent advances in minimally invasive surgery allows for decreased peri-operative morbidity and hospital stay.[3]

Conclusions

Esophageal granular cell tumor is considered as a sub epithelial lesion when it is asymptomatic one, especially in the distal part of the esophagus. Esophageal ultrasound is the most beneficial diagnostic tool which helps in planning the appropriate treatment based on the depth of invasion. Complete resection by EMR (endoscopic mucosal resection) or ESD (endoscopic submucosal dissection) is recommended. Long term follow up needed because of the high recurrence rate. Patient is symptom less for the past 10 months of follow up period.

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