MULTIPLE GRANULAR CELL TUMOR
OF THE ESOPHAGUS TREATED ENDOSCOPICALLY

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ABSTRACT
Granular cell tumor (GCT) of esophagus is a rare lesion, usually found incidentally during upper gastrointestinal endoscopic examination undertaken for another reasons. The origin of this neoplasm is still unclear but no organ seems to be immune to this neoplasm. Although most of the lesions are benign, few reports of malignant GCT, synchronous as well as metachronous, are also reported. It can occur at any age, but are most commonly observed in fourth and fifth decade of life. We herein report a case of multiple GCT treated by endoscopic mucosal resection and review the relevant literatures.

Key Words: Granular Cell Tumor, Esophagus, Endoscopic Mucosal Resection.

INTRODUCTION
Granular cell tumor, formerly known as granular cell myoblastoma was, first described in 1926 by Abrikossof as a rare tumor of the tongue.1 It is estimated that about 5-11% of all tumors arise in the gastrointestinal tract2 and about one third of them are localized in the esophagus.3 It is usually an incidental finding, detected during upper gastrointestinal endoscopy. Most of the esophageal GCTs are asymptomatic and the symptoms reported are often nonspecific and are attributed to the large tumors. Single GCT is usual but multifocality is described, either in the esophagus alone or in other sites. Multifocality does not seem to carry an increased risk of malignant behavior. So far less than 40 cases of multiple esophageal GCTs are reported in the English literatures. The sites of distribution of multiple GCT are lower (54%), middle (38%) and upper third (8%) respectively.4 It usually appears as yellowish, polypoid lesion or submucosal tumor of varying size with or without depression in the surface. Histopathological examination of the biopsied material is the mainstay of diagnosis. Although submucosal tumor, these tumors frequently have nests of cells immediately beneath the mucosa that...
allows diagnosis. In some cases the overlying epithelium is
thick due to hyperplasia and the biopsied specimen may not
contain tumor cells. However, multiple biopsies will provide
sufficient specimens for histopathological examination to
make diagnosis. Endoscopic ultrasonography is valuable
to assess the exact location and extent of the tumor and
to determine if the lesion can be resected endoscopically.
Various modalities of treatment are available with
acceptable success rate for each method.

CASE REPORT

A 46-year-old Japanese man was referred to our unit
with increase in size of elevated mucosal lesion of the
esophagus, which was followed up for last two years. He
was admitted for additional evaluation and treatment. On
admission, he was symptom free and physical examination
was normal. Laboratory data were within normal limit
and there was no elevation of tumor markers. Endoscopic
examination revealed three submucosal tumors (SMTs),
one at 28 cm (from incisor) and two at 35 cm of the
esophagus (Fig.1a,b). All SMTs were yellowish in color
and there was umblication of the surface with reddish
discoloration of mucosa. Overlying mucosa was intact
as there was no abnormality on Lugol’s iodine staining.
Biopsy specimens obtained from all three lesions were
diagnosed histologically as GCT. Endoscopic ultrasound of
all SMTs revealed isoechoic lesion confined in submucosa
(Fig.1c). Colonoscopy was negative for mucosal lesions.
All the tumors were resected endoscopically in a single
setting. Postoperative period was uneventful and patient
was discharged after 3 days of hospital stay. Repeat
endoscopy done after 1, 6 and 12 months period showed
no recurrence. Post resection scars are visible in two sites,
but there is no stenosis due to scar.

DISCUSSION

Fig. 1: Esophagoscopy showed one SMT measuring 11mm X 8mm SMT at 28cm (from incisor teeth)
(A) and two SMTs measuring 13mm X 8mm and 12mm X 6mm at 35cm of the esophagus.
(B) All the SMTs were yellowish in color with umblication at the surface with intact mucosa.
Granular cell tumor is an entity more common to the pathologist than to the clinicians. After description of first case of esophageal GCT by Abrikossof himself in 1931, there have been reports of little more than 200 cases. GCTs are usually benign but a few malignant GCTs have been reported, and it is estimated to make up fewer than 2% of all lesions. A few cases of malignant GCT of esophagus have also been reported. There is an interesting yet unexplained association between GCT and other malignant cancers, which seems to be relatively high. Esophageal GCT is also associated with squamous cell carcinoma of the esophagus, although the reported cases are too little to work out its correlation and seems to be coincidental.

As most of the esophageal GCTs are benign, few authors have recommended follow up for small and asymptomatic lesions. Our case was also followed up (although without definitive diagnosis) for two years before he was subjected to the treatment. It seems that esophageal GCTs remain stable for a longer time and change slowly even without therapy. The fact that larger GCTs are associated with symptoms and all the malignant GCTs so far are greater than 4cm, we recommend treatment for larger GCT or those GCT, which shows tendency to enlarge.

Owing to the rarity of the disease no treatment plan has yet been established. In the past, the chosen treatment was surgical excision but the percentage of malignant degeneration is so low that it cannot be justified and it carries several risks of operative mortality and morbidity. Several endoscopic techniques have been introduced for the treatment of esophageal GCT. Laser treatment has been performed successfully, but this is a costly procedure and requires lengthy hospital stay. Due to necrosis of deeper tissue, risk of perforation exists if not done by expert hands. Injection sclerotherapy using dehydrated alcohol and 1% polidocanol has been attempted in its treatment. These are a low cost technique and can be applied in situation where other modalities of treatment are prohibitive due to cost factor. However, it requires multiple endoscopy and complication like esophageal stenosis will add to the morbidity of the patients and demand further treatment. Endoscopic removal is the preferred treatment. Endoscopic mucosal resection is increasingly having patient compliance due to short hospital stay and less morbidity. Definitive diagnosis is possible after obtaining the resected specimen. Microscopic examination of resected margin enables understanding of completion of treatment.

In conclusion, we acknowledge that GCT should be considered as differential diagnosis in the submucosal tumor of the esophagus. It may be multifocal. Malignant degeneration and infiltration is possible. So, we advise endoscopic removal, which is a safe procedure and carries less operative risk.

REFERENCES

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