Esophageal Granular Cell Tumor Treated by Endoscopic Mucosal Resection

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Dear Sir,

Granular cell tumors, first described in 1926 by Abrikossoff, are relatively rare lesions with a reported incidence of 0.02–0.03% of all tumors in the general population. About 2.7–11% of all granular cell tumors occur in the digestive tract, and approximately 5–12% of these tumors occur multiply. About one third of all digestive tract granular cell tumors arise from the esophagus, 10% from the stomach, and those arising from the colon or rectum are rare.

A 45-year-old man visited our hospital for a health checkup. Endoscopic examination revealed the presence of a submucosal tumor with a yellowish white surface in the lower esophagus. Endoscopic mucosal resection (EMR) was performed using a plastic cap (EMR-C). We used a distal plastic hood at the end of the endoscope to facilitate drawing the lesion up. Thereafter, the elastic was positioned, a diathermy loop was placed around the lesion enclosed by the elastic, and the tumor was resected. Microscopic examination showed a solid tumor with tumor cells arranged in small nests and cords in the submucosal layer (fig. 1). Endoscopy 6 months later showed healing of the esophagus, without any evidence of recurrence.

Macroscopically, granular cell tumors appear as slightly elevated, firm, whitish gray to yellow, smooth nodular tumors. Standard endoscopic biopsy is unlikely to yield a correct diagnosis because most granular cell tumors of the gastrointestinal tract are usually covered by normal mucosa and just a small portion of the tumor can be resected.

Histologically, granular cell tumors were first considered to be of myogenic origin, but electron microscopic and immunohistochemical studies have confirmed the Schwann cell origin of the tumors. The tumor is usually benign and remains biologically quiescent for prolonged periods of time. However, approximately 1.5–2.7% of all cases reported in the literature were considered as malignant variants. Granular cell tumors measuring <20 mm in diameter account for 85% of the cases, although malignant granular cell tumors as small as 10 mm have also been reported. Large

Fig. 1. Microscopic examination showed a solid tumor with the tumor cells arranged in small nests and cords in the submucosal layer. $\times 2$.

size (>4 cm), rapid growth and/or rapid recurrence after previous excision, and invasion of adjacent tissues are reported to be better markers of malignant behavior than the histological features. Malignant granular cell tumors are classified into two types: the 'both histologically and clinically malignant' type, and the 'histologically benign but clinically malignant' type.

EMR is a safe and effective treatment of the tumors, enabling a histopathologic diagnosis. Various EMR techniques, including strip biopsy, endoscopic aspiration mucosectomy using the hood technique (EMR-C), use of a transparent tube and use of a ligating device have been reported in the treatment of granular cell tumors.

EMR is actually considered the therapy of choice for superficial malignant tumors localized in the mucosa and in the upper third of the submucosa. Submucosal lesions of <2 cm in diameter

and separated from the muscularis propria, can also be removed endoscopically. On endoscopic ultrasound, granular cell tumors appear as submucosal homogeneous and hypoechoic masses with a well-defined margin. Endoscopic ultrasound helps to determine whether or not the tumor is separate from the muscularis propria. If the tumor is initially found to be separate from the muscularis propria, the distance between the tumor and the muscularis propria can be increased by injecting an appropriate solution to lift the lesion, after which removal can be carried out more safely and effectively. Massive hemorrhage and perforation can be prevented if the muscularis propria is not injured.

In conclusion, we report the successful removal of an esophageal granular cell tumor by EMR-C. We believe that, given the low risk of EMR, this procedure should be offered as a treatment option for patients with granular cell tumors.