Granular cell tumor of the esophagus and its endoscopic treatment

Özofagusun granuler-hücreli tümörü ve endoskopik tedavisi

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To the Editor

The most often encountered benign esophageal tumors are submucosal leiomyoma, squamous papilloma and inflammatory fibroid polyps. Some lesions, such as granular cell tumor (GCT), leiomyomas and gastrointestinal stromal tumors (GIST) may be difficult to diagnose by biopsy because they are mostly submucosal. Although GCTs are almost exclusively benign lesions, determination of their malignant potential and appropriate management are as yet unresolved problems.

A 40-year-old man with a 4-year history of mild sensation of a retrosternal "sticking" of foodstuffs was hospitalized. His medical history was unremarkable. Laboratory parameters including tumor markers were all normal. Endoscopy revealed a smooth sessile polyp (1 cm) with overlying normal mucosa in the distal esophagus. Computed tomography showed no evidence of nodal or visceral metastases. Initial endoscopic biopsies revealed a diagnosis of squamous cell papilloma. Later, the solitary lesion was successfully treated by endoscopic polypectomy, and the specimen of the polyp, which was confined to the submucosa, revealed GCT (Figure 1a). The tumor was composed of irregular bands of fibroblastic cells and the lamina propria contained sheets of enlarged polygonal cells with granular cytoplasm. The cytoplasmic granules were positively immunostained with S-100 (Figure 1b) and neuron specific enolase (NSE). The tumor was negative with C-kit (CD117), smooth muscle actin, desmin and myoglobin. An endoscopic ultrasound scan showed normal findings in the resected area.

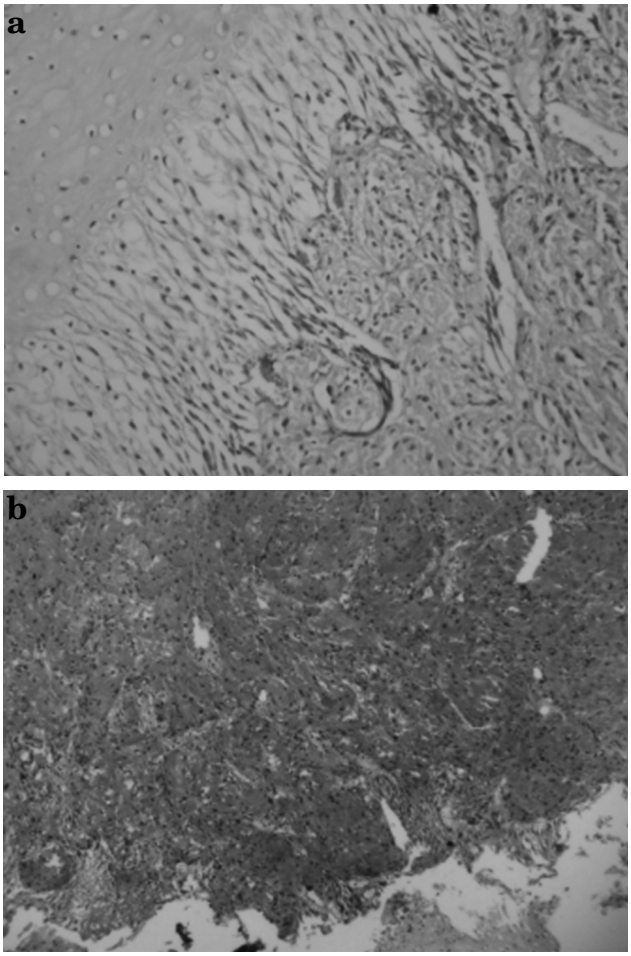

Figure 1. a) The microscopic findings of the polyp: confined to the submucosa and composed of sheets of enlarged polygonal cells with granular cytoplasm, **b)** Cytoplasmic granules of the tumor which positively immunostained with S-100

Approximately 10% of GCTs involve the gastrointestinal tract; the esophagus is the most frequent site (1). GCTs were first reported by Abrikossoff in the early 1900s (2). It is believed that these tumors are derived from neural or Schwann cell elements, as evidenced by positive staining for S-100 protein and NSE. It is often incidentally diagno-

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Manuscript received: 04.12.2006 **Accepted:** 15.03.2007

sed at endoscopy. The present case showed a confusion in diagnosis when biopsy was taken with endoscopic forceps and was mistaken for squamous cell papilloma. Furthermore, GISTs are important submucosal lesions due to their malignant potential, and up to 90% of GISTs are strongly positive for CD117. It is reported that a GCT might mimic a GIST or spindle-cell squamous cell carcinoma (3). Hence, pathologists should be aware of these difficulties.

Observation is suggested in small asymptomatic GCTs. The management options include endoscopic polypectomy and surgery. The GCTs are usually benign neoplasms; malignant GCTs constitute fewer than 2% of all tumors (4). Endoscopic or surgical excision should be restricted to symptomatic patients with tumors larger than 1 cm or with histologic features of malignancy (1, 5). This paper draws attention to considerations in granular cell tumor and potential differential diagnosis.

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