CASE REPORT

Primary Esophageal Adenoid Cystic Carcinoma

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Adenoid cystic carcinoma (ACC) is common in the salivary glands but rare in the esophagus. Routine esophagogastroscope performed in a 54-year-old woman as part of a medical check-up revealed a submucosal tumor (1.5×1.0 cm) at the mid-esophagus. Endoscopic ultrasonography revealed a lesion with mixed echogenicity in the submucosal layer. The submucosal mass was removed by incisional endoscopic enucleation, and pathological analysis revealed epithelial cells with small hyperchromatic angular nuclei in tubular and cribriform patterns. The lesion was pathologically confirmed as an ACC of the esophagus. (*Gut and Liver* 2007;1:178-181)

Key Words: Adenoid cystic carcinoma; Esophagus

INTRODUCTION

Adenoid cystic carcinoma (ACC) is a malignant tumor that is commonly found in the minor salivary glands. Other sites of occurrence include the tracheobronchial tree, breast, skin, lacrimal gland, and the female genital tract. ACC is known to be a slowly growing epithelial tumor. ACC makes up approximately 18.1% of all neoplasm found in salivary glands.¹ Sixty percent of ACC cases occur in the minor salivary glands. Esophageal ACC accounts for 0.1% of esophageal malignancies.² Metastatic regional lymph nodes are uncommon, while distant spread to the lungs and bones is frequent.³ According to the previous studies the prognosis is very poor due to the distant metastases. We report a rare case of primary esophageal ACC that presented as a submucosal mass without metastases.

CASE REPORT

A 54-year-old woman was admitted for the treatment of a submucosal mass detected during a routine endoscopic check-up. The patient was not a smoker and did not drink alcohol. The physical examination did not show any specific findings. The carcinoembryonic antigen (CEA), a marker for squamous cell carcinoma, was normal. The other laboratory findings were normal. An esophagogastroscope examination showed a submucosal tumor of 1.5 × 1.0 cm at the mid-esophagus (Fig. 1). Endoscopic ultrasonography (EUS) showed a lesion with mixed echogenicity in the submucosal layer with intact muscularis propria at the mid-esophagus (Fig. 2). With the agreement of the patient, the submucosal mass was removed successfully by incisional endoscopic enucleation in order

Fig. 1. Endoscopic finding. It shows a 1.5×1.0 cm-sized submucosal tumor at the mid-esophagus.

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for both confirmative diagnosis and treatment.

Histologically, the tumor was a well-circumscribed, non-encapsulated, ovoid, grayish-white, homogeneous, and solid mass found in the submucosa. The tumor was composed of epithelial cells with small hyperchromatic

Fig. 2. EUS finding. It shows a mixed echogenic lesion in the submucosal layer at the mid-esophagus.

angular nuclei in tubular and cribriform patterns. The cells formed ducts, tubules and cylindromatous microscopic spaces, the lumen of which contained eosinophilic mucoid materials. No solid pattern was noted. No squamous differentiation was present. These findings were consistent with an adenoid cystic carcinoma developing from a minor salivary gland of the esophageal mucosa (Fig. 3). A fusion Positron Emission Tomography-Computed Tomography (PET-CT) scan showed neither lymphadenopathy nor evidence of distant metastases. After a 10-month follow-up, the patient remained free from symptoms and metastases.

DISCUSSION

ACC involves the minor salivary glands, submandibular glands and parotid glands.⁴ The clinical behavior of ACC in the head and neck is paradoxical. First, although tumor growth is slow, its clinical course is relentless and progressive. Second, even though an operative intervention is usually feasible, multiple local recurrences are usually followed. Third, although metastatic spread to regional

Fig. 3. Pathologic findings of the resected submucosal tumor. (A) It shows mixed tubular and cribriform features (H&E stain, ×200). (B) It shows numerous 'cylinders' containing eosinophilic material (H&E stain, ×400).

Table 1. Review of Cases of Esophageal Adenoid Cystic Carcinoma Presenting as Submucosal Tumor

	Site	Size	Metastasis	Treatment	Follow up
Kabuto T, et al ⁸	Lower	1.4×1.0 cm	None	Esophageal resection+Esophagogastrostomy	18 months
Blaauwgeers JL, et al ⁹	Middle	1.5×1.0 cm	None	Esophageal resection+Esophagogastrostomy	5 months
Kim JH, et al ¹⁰	Lower	2.3×1.8 cm	None	Esophageal resection+Esophagogastrostomy+ Combination chemotherapy	12 months
Present case	Middle	1.5×1.0 cm	None	Endoscopic enucleation	10 months

lymph nodes is uncommon, distant spread to lungs and bones is frequent.³

The clinical behavior of esophageal ACC is not well known due to the small number of reported cases.⁵⁻⁷ The average age of patients is 65 years, with a male to female ratio of 3.4 to 1. The most common symptom is progressive dysphagia.^{5,7} ACCs have most frequently been reported in the middle third of the esophagus (63%), less often in the lower third (30%) and rarely in the upper third (7%). The most common endoscopic findings include a fungating or polypoid mass rather than an ulcerative or infiltrative growth pattern.⁵ Only three cases have been reported where they were presented as submucosal mass (Table 1).⁸⁻¹⁰ One of the tumors was investigated by endoscopic ultrasonography.⁸ The ultrasonic image in the report showed a thickened hyperechoic submucosal layer and a relatively well-preserved hypoechoic muscularis propria.

Histologically, ACC of the salivary gland may show three different growth patterns: cribriform, tubular and solid. The cribriform pattern, which is most frequently noticed, is characterized by having nests of cells with cylindromatous microcystic spaces. In the tubular form, well-formed ducts and tubules with central lumina, are lined by inner epithelial and outer myoepithelial cells. These are filled with hyaline mucoid material. The solid or basaloid type consists of sheets of uniform basaloid cells lacking tubular or microcystic formation. In the cribriform and solid variants, small true ducts are invariably present, but may not be immediately apparent.^{11,12} The solid pattern appears to be associated with worse prognosis than the other two patterns.^{13,14} In this case, the submucosal lesion was an ACC of the esophagus and there were no solid patterns identified.

The treatment of choice for ACC is radical excision. When the surgical margin is positive, postoperative radiotherapy is recommended. Chemotherapy is not usually chosen due to a poor response rate.¹⁵ The 5-year survival rate is approximately 35%, but the long-term survival is poor. Eighty to 90% of patients die of this disease within 10-15 years.¹⁶ Poor prognostic factors that influence survival include solid histological pattern, advanced clinical stage, and positive surgical margins.¹⁶ Few treatment guidelines are available for esophageal ACC. The treatment of choice for an esophageal ACC is radical excision (i.e., esophagectomy and esophagogastrostomy). The role of chemotherapy, through either adjuvant or primary chemotherapy, is not clear.^{17,18} Postoperative radiotherapy may help improvement of progressive dysphagia.⁹ Until now, three reported cases presenting as a submucosal mass generally had good clinical courses without meta-

stasis after the radical excision (Table 1).⁸⁻¹⁰

Here we report a rare case of esophageal ACC presenting as a submucosal mass detected by routine endoscopic examination and treated by incisional endoscopic enucleation.

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