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Giant Pedunculated Sarcoma Together with Carcinoma In Situ in the Esophagus

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Sarcomas are the least common of all malignant esophageal neoplasms, with an incidence of 0.5%. We describe a case of a giant esophageal pedunculated polyp identified by barium ingestion and endoscopy. Biopsy of the polyp was performed using panendoscopy and the pathology report was of an ulcer with atypical reactive stromal cells. Initially, the patient was thought to have a benign tumor and underwent endoscopic removal of the polyp but this failed because of bleeding from the stalk. After subtotal esophagectomy, the final histological diagnosis was that a sarcoma and a squamous cell carcinoma in situ coexisted in the esophagus. The patient had an uneventful postoperative course and showed no evidence of recurrence in three years of follow-up. We review the literature on esophageal sarcomas and discuss the limitation of panendoscopy in diagnosing unusual esophageal tumors.

Key words: sarcoma, carcinosarcoma, squamous cell carcinoma in situ, esophagus

INTRODUCTION

Squamous cell carcinomas and adenocarcinomas are the major histological types of primary esophageal carcinomas, accounting for more than 95%[1,2]. However, sarcomas of the esophagus are unusual malignant mesenchymal neoplasms, which can arise primarily in the esophagus. The incidence of primary sarcoma of the esophagus is 0.5% of all primary malignant esophageal neoplasmas[1]. Common presenting symptoms are dysphagia, odynophagia and progressive weight loss. In patients with a suspected esophageal neoplasm, barium ingestion esophagraphy and panendoscopy are required for a diagnosis. Accurate histopathology before the operation aids in planning a better strategy for surgical intervention, because surgical treatment varies from simple enucleation or endoscopic resection to partial or total esophagectomy, or even to transcervical, transthoracic or transgastric resections. Here, we report a patient with a giant pedunculated sarcoma and squamous cell carcinoma in situ coexisting in the esophagus but for whom a diagnosis was missed before surgical intervention.

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CASE REPORT

A 49-year-old man was admitted to our hospital because of epigastric pain and dysphagia over five months. He denied any past medical history. He drank heavily but had not smoked for 20 years.

A barium ingestion examination showed a huge mass in the mid-third of the esophagus. Panendoscopy examination showed a giant pedunculated polyp. The top of the tumor was 23 cm from the mouth, the bottom 35 cm and it almost filled the lumen when biopsied. It seemed to be attached to the wall of the esophagus by a narrow stalk (Fig. 1A). A chest computed tomography (CT) scan revealed a well-circumscribed low attenuation mass in the dilated esophagus. The polyp was diagnosed as an ulcer with atypical reactive stromal cells from a biopsy. Chest X-ray and preoperative routine laboratory examinations did not reveal any abnormalities before the patient received surgical removal of the mass.

Because of potential airway obstruction, we decided to try to thrombose the vascular supply to the polyp and separate the mass from the wall by repeated delivery of laser pulses to the stalk using endoscopy with the patient under general anesthesia, but failed because of much bleeding from the stalk. Right thoracotomy was then performed; the esophagus was opened longitudinally and the giant polyp was removed ventrally (Fig. 1B). The polyp was soft and gray with extensive necrosis for a distance of 13 cm. However, frozen section histopathology revealed a high-grade sarcoma. The patient then underwent a subtotal esophagectomy and posterior mediastinal esophagogastrostomy.

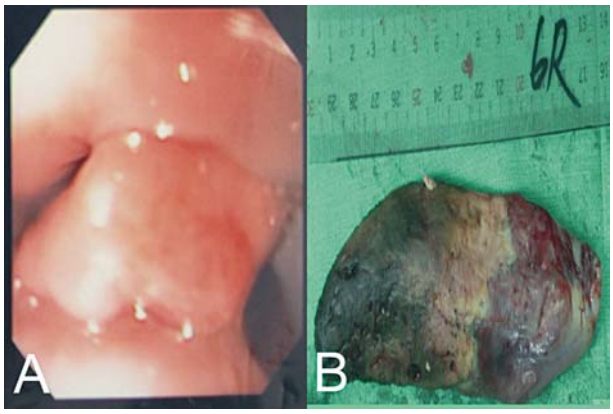

Fig.1 (A) Panendoscopic view of the stalk of the giant tumor, which almost filled the lumen of the oesophagus. (B) Resected a pedunculated polyp from the wall of the esophagus. The maximal extent of this tumor was 13 ×5×3cm.

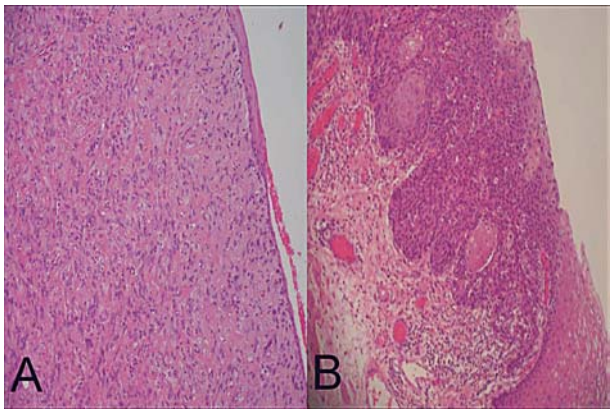

Fig 2 (A) Histological findings of the resected tumor, high grade sarcoma. (B) Histological findings of the resected esophagus, squamous cell carcinoma in situ.

Histopathology of the giant pedunculated polyp revealed a predominantly high-grade sarcoma with a pleiomorphic appearance (Fig. 2A). Some cells had elongated fusiform nuclei arranged in an intertwining and storiform pattern with only little cytoplasm. These cells resembled fibroblasts and small amounts of collagen could be seen between them. In addition, there were some giant cells showing bizarre shaped, large, hyperchromatic nuclei with frequent mitoses. The few tumor cells were positive for vimentin and CD68, but negative for cytokeratin, EMA, S-100, CD34, CD117 (C-kit) and smooth muscle α-actin (SMA). The resected esophagus showed a squamous cell carcinoma in situ and the tumor cells showed enlarged hyperchromatic nuclei through the entire layer of the squamous epithelium but no residual sarcoma (Fig. 2B).

The patient's postoperative course was unremarkable. No evidence of distant metastases was found on subsequent staging. No postoperative adjuvant treatment was given and the patient was alive and free of disease 27 months after surgery.

DISCUSSION

Epithelial tumors of the esophagus (i.e., squamous cell carcinomas and adenocarcinomas) account for more than 95% of all esophageal cancers[1,2]. Malignant mesenchymal sarcomas can arise nearly anywhere in the body, including the esophagus. Esophageal sarcomas are rare; no evidence has suggested that they are increasing in frequency, and they occur most commonly as polypoid intraluminal masses[3]. They may be divided into tumors with mixed epithelial and spindle cell characteristics such as carcinosarcomas, and pure sarcomas of mesenchymal origin such as leiomyosarcomas. These tumors commonly present as three different types: (1) polypoid, which may be either sessile or pedunculated; (2) diffuse, which can simulate a carcinoma; and (3) annular. The polypoid type is said to be noninvasive and to metastasize late, while the diffuse type is invasive and metastasizes early[4].

Esophageal carcinosarcomas are composed of both carcinomatous and sarcomatous elements. The histogenesis of the sarcomatous spindle cell component of this tumor type is a subject of debate. As a result, this tumor has been given a number of different names, including pseuodosarcoma, pseudosarcomatous carcinoma, spindle cell carcinoma, polypoid carcinoma and carcinosarcoma[5]. Studies using electron microscopy and immunostaining showed cytoplasmic organelles specific for squamous cells in the spindle cells of these polypoid tumors. The histological appearances are now considered to represent divergent patterns of differentiation in squamous cell carcinomas. According to the Japanese Society for Esophageal Disease, three major theories have been proposed for the pathogenesis of carcinosarcomas. The first is that the spindle cell component is a reaction to the carcinoma[6]. The second, known as the collision theory, proposes that two individual stem cells might undergo malignant transformation independently and simultaneously, and are actually separate tumors that have merged. The third theory is that individual elements are derived from a single common ances-

tor cell (a so called carcinosarcoma)[6]. This tends to remain localized within the esophageal lumen and not to metastasize. As these are slow-growing tumors, an aggressive surgical approach is recommended. It has been suggested that carcinosarcomas have better prognoses than squamous cell carcinomas but they both occur most commonly in middle aged and elderly males with a history of smoking or alcohol abuse[7-8].

Pure sarcomas of the esophagus such as fibrosarcomas, leiomyosarcomas and rhabdomyosarcomas are very rare. The most common histological type of the esophageal sarcoma is a leiomyosarcoma, which constitutes nearly two thirds of these unusual neoplasms[9]. The diagnosis of a malignant soft tissue tumor (sarcoma), especially in the gastrointestinal tract, has a number of problems. Immunocytochemistry is strongly recommended for the precise definition of sarcomas in the gastrointestinal tract[10-11]. Immunopositivity for desmin and SMA are useful for confirming a histological impression of smooth muscle bundles[12]. Stromal tumors are positive for S-100 protein and Schwannomas are positive for NSE[13]. The application of immunohistochemical antiepithelial markers (e.g., for cytokeratin) would be helpful in verifying the presence or absence of an epithelial component in the tumor with carcinosarcoma or sarcoma components[14]. Moreover, vimentin immunoreactivity is indicative of a mesenchymal origin of spindle cells[15]. In the present case, the pedunculated sarcoma was only immunoreactive for vimentin, but was negative for cytokeratin and SMA, which would indicate carcinomatous cells and smooth muscle bundles, respectively.

In this case, the differential diagnosis of a pedunculated polyp of the esophagus included a number of benign and malignant entities. In the benign category, myofibromas, lipomas, fibrovascular polyps and leiomyomas can each present as a pedunculated mass. Among the malignant lesions, adenocarcinomas, squamous carcinomas, leiomyosarcomas, fibrosarcomas and rhabdomyosarcomas are causes of esophageal pedunculated mass lesions. Differentiation between these various lesions can be difficult radiologically or endoscopically, usually requiring histopathology from a biopsy. Surgical resection should be considered first for all lesions if the patient can tolerate an operation. Surgical treatment varies from simple enucleation or endoscopic resection to partial or total esophagectomy, or transcervical, transthoracic or transgastric resections. Chemotherapy and concomitant radiation therapy must also be considered for residual microscopic disease and local control. The prognostic factors for esophageal tumors are histological subtypes, malignancy grade, location and surgical radicality. Polypoid lesions usually tend to have a more favorable prognosis than infiltrating or intramural lesions from five-year survival rates.

Although the preoperative diagnostic biopsy of an esophageal tumor is routinely made via endoscopy, it is important to be aware of the limitations of this approach. Endoscopic biopsy specimens can be helpful if the mucosa overlying the tumor is ulcerated. If the overlying mucosa is intact, a superficial biopsy may yield a false negative result. In a review of 127 carcinosarcomas and 56 pseudosarcomas of the esophagus reported in the literature, Iascone and Barreca found that the correct diagnosis was obtained by means of endoscopic in less than one third of cases[16]. Because accurate histological diagnosis before surgery is important, we suggest that the initial biopsy should obtain an adequate volume of representative tissue for diagnosis. In this patient, we missed the correct diagnosis at first because the biopsies were inadequate and proved negative for malignancy. Because malignancy could not be ruled out, we performed intraoperative frozen section histopathology of the polyp, which proved decisive. We therefore recommend that early surgical removal and histopathology should be used to diagnose an esophageal pedunculated polyp.

In conclusion, there are rare benign polypoid tumors of the esophagus. The esophageal sarcoma polypoid type has a slow and indolent clinical course, followed by late recurrence and eventual death of the patient from the disease. Spread by hematogenous means is the cause of tumor recurrences. Esophagectomy or esophagogastrectomy are the surgical choices. The best therapeutic strategy for a sarcoma remains controversial, but we believe that surgery will remain the best choice for esophageal sarcomas, and patients will need long-term follow up.

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