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Case report

PRIMARY ADENOID CYSTIC CARCINOMA IN THE ESOPHAGUS

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Abstract

Esophageal adenoid cystic carcinoma (EACC) is an exceedingly rare malignant tumor constituting only 0.2% of all esophageal tumors. The tumor exhibits aggressive behavior, composed histologically of ductal and modified myoepithelial cells. We report a case of a 69-year-old female with a diagnosis of an EACC by preoperative endoscopic biopsy. Thoracoscopy esophagectomy was carried out. However, pleural metastasis was found. Therefore, surgical resection of the esophageal tumor was not carried out. The patient underwent an uneventful recovery, followed by palliative treatment and ongoing chemoradiotherapy. EACC is uncommon but exhibits a more aggressive nature compared to its counterparts in the head and neck region. Dysphagia associated with gastroesophageal reflux disease is a common symptom. The duration from symptom onset to diagnosis is typically short. Treatment options include surgical resection, chemotherapy, and radiotherapy, with surgery being the preferred initial approach despite high operative mortality. Prognosis remains inconclusive, with some studies associating poor outcomes with lymph node metastasis and vascular invasion, while others report better survival rates. EACC presents diagnostic and therapeutic challenges due to its rarity and aggressive nature. Prognostic considerations remain unclear, emphasizing the need for further research and accumulated cases to delineate optimal treatment. The presented case demonstrates a 1-year survival with systemic palliative care, contributing to the evolving knowledge surrounding EACC.

Key words: adenoid cystic carcinoma, esophageal cancer, esophageal adenoid cystic carcinoma, surgery

Resumen

Carcinoma adenoide quístico primario en el esófago

El carcinoma adenoide quístico primario de esófago (EACC) es un tumor maligno excepcionalmente raro que constituye solo el 0.2% de todos los tumores esofágicos. El tumor exhibe un comportamiento agresivo, compuesto histológicamente por células ductales y mioepiteliales modificadas.

Presentamos el caso de una mujer de 69 años con diagnóstico de un EACC mediante biopsia endoscópica preoperatoria. Se realizó una esofagectomía por toracoscopia. Sin embargo, se encontró metástasis pleural. Por lo tanto, no se llevó a cabo la resección quirúrgica del tumor esofágico. La paciente tuvo una recuperación sin complicaciones, seguida de tratamiento paliativo y radioquimioterapia continua. El EACC es poco común, pero exhibe una naturaleza más agresiva en comparación con sus contrapartes en la región de la cabeza y el cuello. La disfagia asociada con la enfermedad por reflujo gastroesofágico es un síntoma común. La duración desde el inicio de los síntomas hasta el diagnóstico suele ser corta. Las opciones de tratamiento incluyen la cirugía, quimio y radioterapia, siendo la cirugía la preferida a pesar de la alta mortalidad operatoria. El pronóstico es inconcluso, algunos estudios asocian resultados pobres con metástasis e invasión vascular, mientras que otros informan mejores tasas de supervivencia. El EACC presenta desafíos diagnósticos y terapéuticos debido a su rareza y naturaleza agresiva. El pronóstico sigue siendo poco claro, lo que enfatiza la necesidad de más investigación para delinear el tratamiento óptimo. El caso

presentado demuestra una supervivencia de un año con cuidados paliativos sistémicos, contribuyendo al conocimiento en evolución sobre el EACC.

Palabras clave: carcinoma adenoideo quístico, cáncer esofágico, carcinoma adenoideo quístico, cirugía

Esophageal adenoid cystic carcinoma (EACC) is an extremely rare malignant tumor. It is frequently linked to parotid and salivary glands[1]. However, it occurs occasionally in the esophagus with an incidence only of 0.2% from all the esophageal tumors[2].

EACC behavior is biologically aggressive. Histologically these tumors are known to consist of two main cell types: ductal and modified myoepithelial cells. In addition, this sort of pathology is accompanied by squamous cell and basaloid squamous cell carcinoma components, which can create different malignancies as adenoid cystic carcinoma (ACC)[3]. Previous studies demonstrated inconsistent findings regarding prognosis and treatment. Some authors described that prognosis depends on the histological grade of the tumor and the presence of lymph node metastasis and vascular invasion[3,4]. Treatment options range from radiation and chemotherapy to surgical therapy. However, evaluating the impact of treatment on survival is difficult[4].

Due to the uncommonness of EACC, data in the literature are scarce. Therefore, we report a case of an aggressive primary EACC in which we intended to treat with a thoracoscopic esophagectomy in a high-volume center. We review the general aspects of EACC, focusing on its treatment and prognosis as well.

Clinical case

A 69-year-old female with a history of hypothyroidism, rheumatoid arthritis, and breast cancer surgically treated in the past, presented to the office with 6 months of progressive dysphagia and 18 kg weight loss in the last year. Endoscopy was performed and revealed a 5-cm-long, cauliflower-like lesion with central excavation, located at 25 cm from the incisors. Multiple biopsies were taken which showed epithelial cell proliferation arranged in rounded nests with cystic areas of mucinous content, eosinophilic cytoplasm, and rounded hyperchromatic nuclei consistent with ACC.

A chest, abdomen, and pelvis staging computed tomography and a positron emission computed tomography were performed, revealing a hypermetabolic esophageal lesion. Both showed no evidence of metastatic disease.

A thoracoscopic esophagectomy was planned. During thoracoscopic exploration, multiple pleural lesions suggestive of metastasis were found (Fig. 1). Biopsies were taken, and the frozen section confirmed malignant proliferation.

Surgical resection of the esophageal tumor was not carried out. The permanent section described a nodular configuration composed of fibro-myxoid connective stroma and atypical epithelial proliferation with basaloid cells arranged in large nests, with necrosis and mitotic activity consistent with carcinoma (Fig. 2).

The patient had an uneventful recovery and was discharged on postoperative day 2. Two months later, a feeding gastrostomy for nutrition was performed and pallia-

Figure 1 | Thoracoscopy showing a protruding irregular lesion located distally in the esophagus, exhibiting a nodular appearance (star). Multiple pleural lesions suggestive of metastasis were found (arrow)

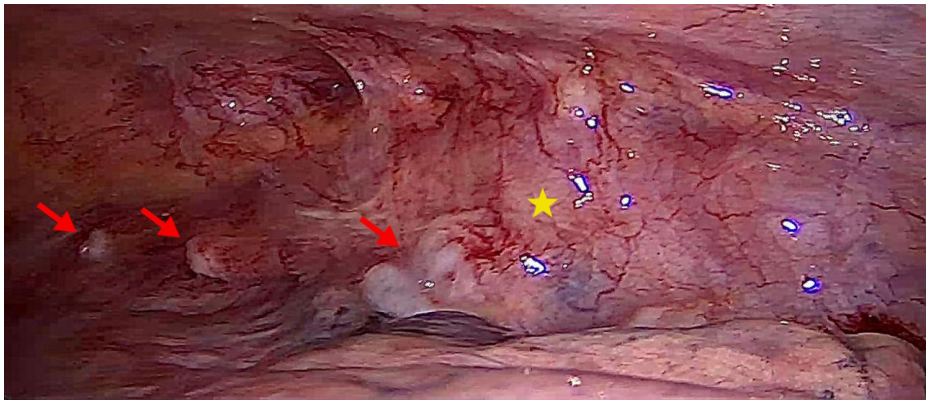
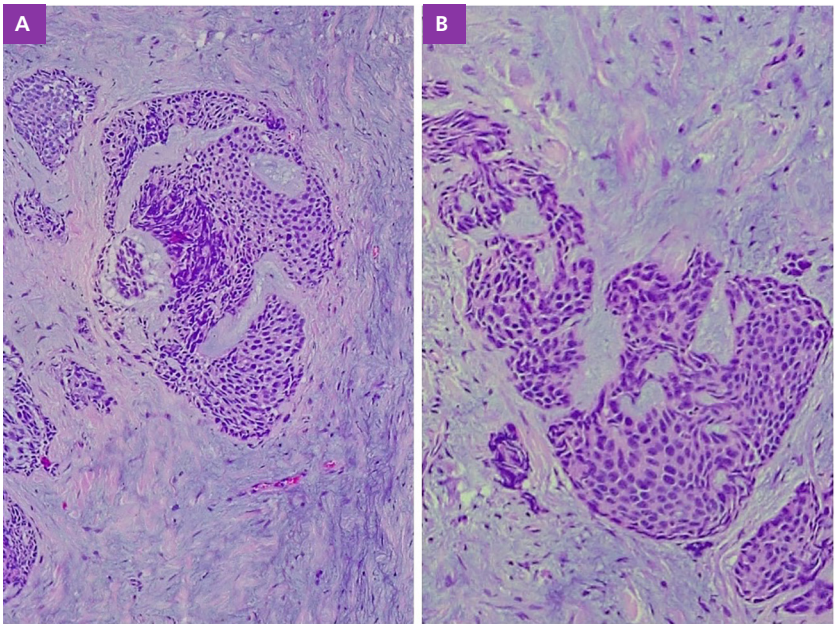


Figure 2 | Histopathological examination showing a nodular proliferation of atypical epithelial cells, primarily composed of basaloid cells arranged in nests. Additionally, evidence of necrosis and mitotic activity is observed. A: Hematoxylin and eosin stain, ×10 magnification. B: Hematoxylin and eosin stain, ×40 magnification



tive treatment was planned. Oncology consultation was indicated and she is currently undergoing her second chemotherapy and radiotherapy session.

Written consent was obtained from the patient to use medical records and images in the publication of this manuscript.

Discussion

ACC was named as such in 1953 by Foote and Frazell[5]. They are considered to arise from myoepithelial cells and intercalated ducts of the submucosal esophageal glands[6]. These tumors contain two main types of cells: epithelial and myoepithelial[4]. Histologically they exhibit small cells with large nuclei and marked hyperchromasia. Furthermore, they demonstrate immunoreactivity to the S100 protein. These tumors originate from mucous glands, particularly the minor salivary glands[2], with the palate, parotid gland, and submaxillary glands being the most common locations. EACC is a rare form of presentation, constituting 0.1% of malignant esophageal tumors[4]. Its most frequent location is in the middle third of the esophagus, followed by the lower third, and lastly the upper third. It is more common in men (4:1) with an average age of 60 years. In contrast to head and neck tumors, ACC and its esophageal presentation appear to exhibit a more aggressive clinical and biological behavior[7]. Bone and lung are the primary locations where metastasis most frequently occurs.

Patients with ACC may experience persistent dysphagia[4]. It may also be associated with gastroesophageal reflux disease as a result of eosinophil infiltration that could cause eosinophilic esophagitis[8]. The duration of these symptoms from their onset to diagnosis is typically short (average time of 3 months)[2]. Consistently with the literature, our patient presented dysphagia as an initial symptom associated with weight loss. Six months went by from the onset of symptoms until diagnosis.

Treatment for these tumors can vary from surgical resection to systemic options like chemotherapy and radiotherapy. Surgical excision is the preferred initial treatment. Previous studies considered surgery as the treatment of choice for localized primary EACC due to the extensive variability in which local recurrences and metastases occur[4-9,10]. However, the operative mor-

tality is fairly high at 15%. Therefore, different treatment options have been described. A previous case report described that combination chemotherapy with doxorubicin, mitomycin C, and 5-fluorouracil is an effective treatment modality in this disease[9]. Nonetheless, it is widely believed that chemotherapy may not be effective[11]. In addition, Yoshikawa et al. reported the first case of an EACC treated with endoscopic submucosal dissection. Despite their results, they concluded that further accumulation and examination of additional cases are required to clarify the prognosis and most appropriate treatment for these patients[3].

Regarding the prognosis, the available information is inconclusive. Previous studies explained that lymph node metastasis and vascular invasion are associated with a poor prognosis[3]. Additionally, other authors reported EACC prognosis as poor with more frequent organ metastasis than in other carcinomas, with a one-year survival rate of 23% and a median survival rate of 7 months[2]. Dutta et al., conversely, reported an overall 5-year survival rate of 47%[5]. In our present case, we performed systemic palliative care treatment with chemoradiotherapy over a 4-month period in the setting of the advanced stage of the disease with distant metastasis. The patient remains alive with an overall survival of 1 year from the time of diagnosis.

In conclusion, ACC is a distinct malignancy originating from myoepithelial cells and intercalated ducts of the submucosal esophageal glands. Its diagnosis is challenging due to its limited occurrence and aggressive clinical behavior. Treatment is still unclear, and literature on the subject is scarce. Surgical resection represents the preferred initial option, with alternatives including chemotherapy and radiotherapy. Prognostic considerations for EACC remain inconclusive. In our specific case, however, we report a one-year survival following a systemic palliative care approach. Further research and accumulated cases are crucial for a clearer understanding of prognosis and optimal treatment modalities in this rare and challenging malignancy.

Conflict of interest: None to declare

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