

Epithelial-myoeplithelial carcinoma of the supraclavicular region: An unusual cause of dyspnea

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ABSTRACT

Epithelial-myoeplithelial carcinoma (EMC) is an extremely rare disease and usually develops in major salivary glands, such as the parotid gland. EMC is regarded as a low grade-malignancy tumor, and the treatment protocol involves wide surgical excision with secure clear margins although postoperative radiotherapy is generally performed to reduce local recurrence. The present study aims to report a case of EMC with a supraclavicular location due to its rare occurrence and atypical location.

Keywords: Dyspnea; epithelial-myoeplithelial carcinoma; supraclavicular region.

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Epithelial-myoeplithelial carcinoma (EMC) is an extremely rare disease, which was first reported by Donath et al. in 1972 [1]. EMC includes two cell groups with distinct features, which are the key to diagnosis. EMC is a rare tumor representing approximately 1% of the salivary gland tumors [2]. EMC usually develops in major salivary glands, such as the parotid gland, but infrequently occurs in the maxillary sinus, trachea, tongue, lung and larynx [3–5].

CASE REPORT

A 58-year-old woman presented with dyspnea and painless mass on the left supraclavicular area that had been present for a year. The patient's history included Diabetes Mellitus (DM) for two years and right hemithyroidectomy 15 years ago, which was the cause of benign thyroid nodule. There is mild fullness on the examination of the nasopharynx. Left vocal cord was observed paramedian

fixation. The result of the nasopharynx biopsy of the patient was lymphoid hyperplasia. Neck ultrasonography (USG) showed a mass of 42x26 mm in the left supraclavicular region that was seen to be blooded with Doppler USG. Fine needle aspiration biopsy was reported as suspicious cytology. Ca, PTH, anti-TPO values were normal. Upper gastrointestinal system endoscopy was carried out. The result of the biopsy was superficial gastritis. Neck computed tomography (CT) and magnetic resonance imaging (MRI) were performed (Fig. 1, 2).

An excisional biopsy was planned for the patient. As an intraoperative problem, it was seen that the left recurrent nerve was completely swollen by the tumor. Approximately 6x4x3 cm mass was totally excised. The pathological result was reported epithelial-myoeplithelial carcinoma; the surgical margin was positive. Chemoradiotherapy was planned because of positive margin and big tumor size. No recurrence was detected in nearly

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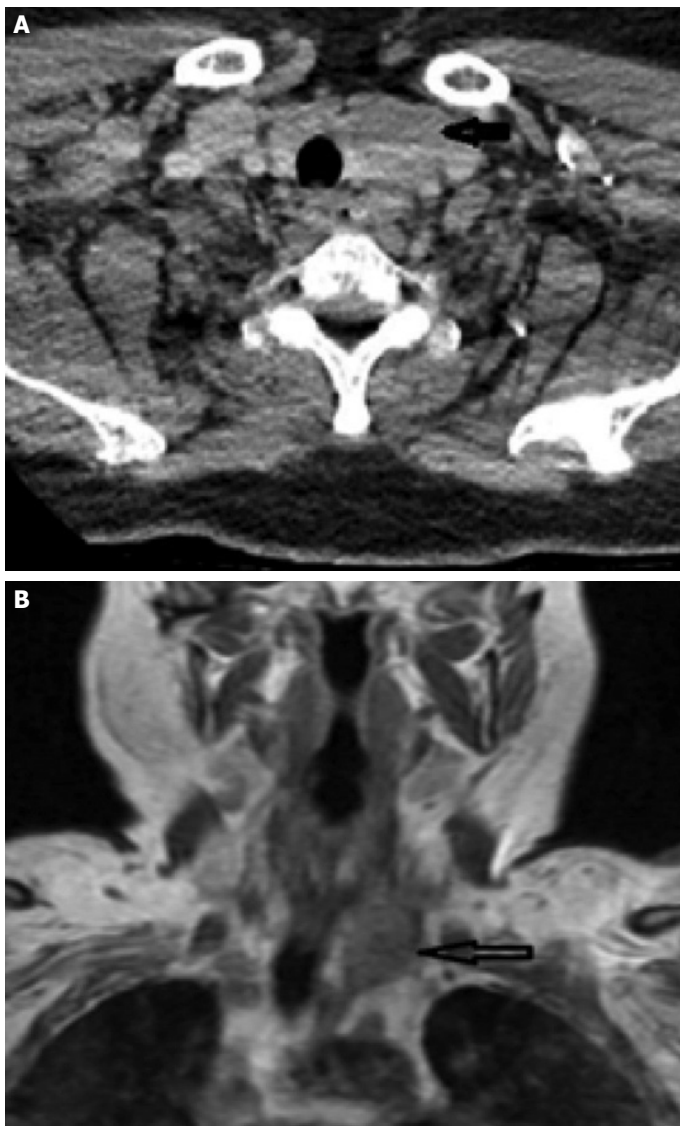


FIGURE 1. (A) Neck computed tomography (CT), axial plan, 4*2,5*3,5 cm size retrosternal mass, trachea pushed right side (arrow). **(B)** neck magnetic resonance imaging (MRI) coronal plan, contrast +, multilobular mass extending to the upper mediastinum (arrow).

18-month follow-ups. Written informed consent was obtained from the patient for this study.

DISCUSSION

Although EMC mostly develops in the parotid gland, some reports claim that it also occurs in the nasal cavity, paranasal sinus, nasopharynx, bronchus, lung, lacrimal gland, submandibular gland, and base of the tongue [2, 4, 5]. To our knowledge, no case of EMC in the supraclavicular region has been reported previously.

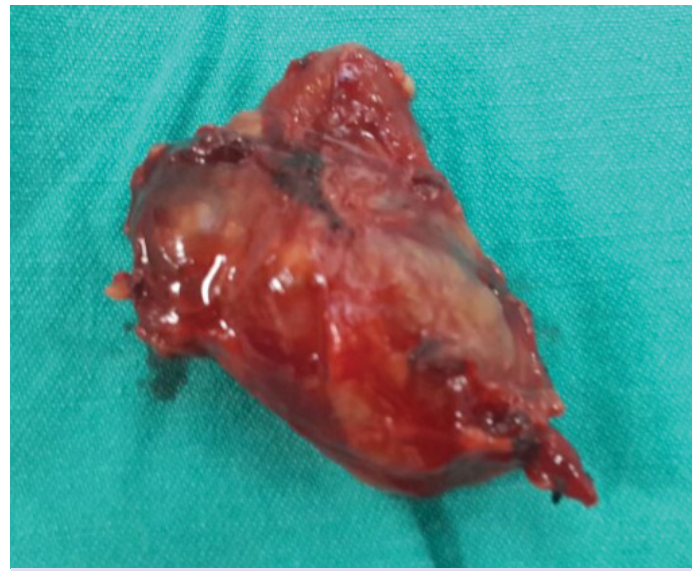


FIGURE 2. Image of surgical excision specimen, nearly 6x4x3 cm and multilobular.

EMC is a typically a low-grade malignant neoplasm, but in rare cases, the tumor may metastasize to distant sites. It has been highlighted that EMC develops more commonly in women and has a higher incidence in the fifth to eighth decades [2]. The symptoms of EMC vary depending on its anatomic position and size. When it encroaches at the cervical area, as found in our case, various symptoms may occur depending on the carcinoma's size, ranging from no symptoms to voice changes and respiratory obstruction. The major problems with our patient were dyspnea and swelling in the neck.

EMC is diagnosed using an optical microscope and using immunohistochemistry. On histology, EMC is characterized by the presence of tubules with two distinct types of cells. The main histological features of EMC are epithelial cells in the inner layer of the lumen and myoepithelial cells surrounding the outer layer [2, 6]. In immunohistochemical analysis, myoepithelial cells in the outer layer are positive for calponin, p63 protein, glial fibrillary acidic protein, S-100 protein, and SMA. Epithelial cells in the inner layer are positive for cytokeratin-7 and epithelial membrane antigen [2].

In our case, the outer myoepithelial cells tested positive for SMA and p63, while the inner epithelial cells were positive for cytokeratin; hence, EMC was diagnosed (Fig. 3, 4).

Other diseases that should be considered in the differential diagnosis of EMC are myoepithelial carcinoma, pleomorphic adenoma, and adenoid cystic carcinoma.

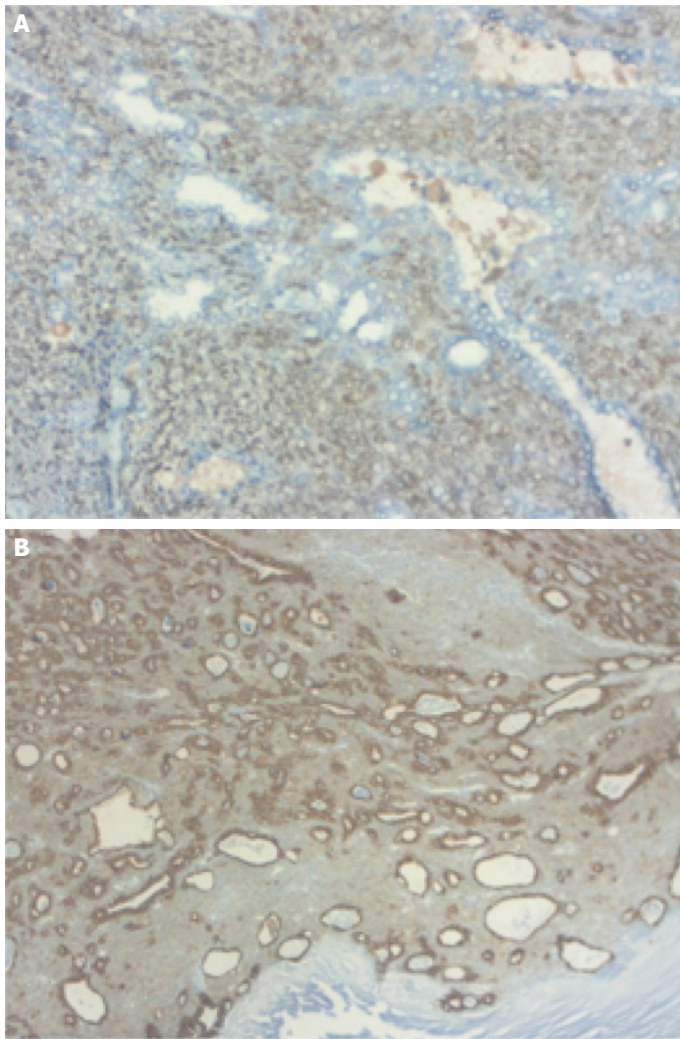


FIGURE 3. (A) Outer myoepithelial cells stain positive for smooth muscle actin (magnification 100), (B) inner luminal cells and outer myoepithelial cells positive for cytokeratin 5/6 (magnification 100).

Similar local morphological features of EMC can be observed in adenoid cystic carcinoma and acinic cell carcinoma [7]. These carcinomas should be widely excised.

EMC is regarded as a low-grade malignancy tumor, and the treatment protocol involves wide surgical excision with secure clear margins [2]. Although postoperative radiotherapy is generally performed to reduce local recurrence, the effects of chemotherapy are not clear yet [2, 8]. Tumors >4 cm are often associated with local recurrence [2]. EMC is a low-grade malignancy, recurrence rate and metastasis account for 23 to 80% and 14 to 25% respectively. Most surgeons prefer wide excision with clear margins, followed by radiotherapy in selected cases to avoid recurrence [9]. Pierard et al. [10] proposed use

in stabilization of pulmonary metastasis in these cases. In our case, we preferred chemoradiotherapy because of the tumor was >4 cm, and the surgical margin was positive.

As a conclusion, the presented case relates to an EMC originating from the supraclavicular region, where there has been no similar case reported previously. EMC is a low grade-malignancy, and the treatment protocol involves wide surgical excision. Postoperative chemoradiotherapy can use big size and positive margin.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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REFERENCES

1. Donath K, Seifert G, Schmitz R. Diagnosis and ultrastructure of the tubular carcinoma of salivary gland ducts. Epithelial-myoepithelial carcinoma of the intercalated ducts. [Article in German]. *Virchows Arch A Pathol Pathol Anat* 1972;356:16–31. [CrossRef]
2. Oh HJ, Do NY, Kee KH, Park JH. Epithelial-myoepithelial carcinoma arising from the subglottis: a case report and review of the literature. *J Med Case Rep* 2016;10:45. [CrossRef]
3. Patra SK, Panda NK, Saikia UN. Epithelial-myoepithelial carcinoma of the maxillary sinus: a rare case. *Laryngoscope* 2012;122:1579–81.
4. Konoglou M, Cheva A, Zarogoulidis P, Porpodis K, Pataka A, Mpaliaka A, et al. Epithelial-myoepithelial carcinoma of the trachea-a rare entity case report. *J Thorac Dis* 2014;6 Suppl 1:S194–9.
5. Hattori K, Murai N, Kusano J, Takahashi Y. Bilateral epithelial-myoepithelial carcinoma of the parotid glands. *Head Neck* 2016;38:E73–5.
6. Peters P, Repanos C, Earnshaw J, Stark P, Burmeister B, McGuire L, et al. Epithelial-myoepithelial carcinoma of the tongue base: a case for the case-report and review of the literature. *Head Neck Oncol* 2010;2:4.
7. Simpson RH, Clarke TJ, Sarsfield PT, Gluckman PG. Epithelial-myoepithelial carcinoma of salivary glands. *J Clin Pathol* 1991;44:419–23.
8. Corio RL, Sciubba JJ, Brannon RB, Batsakis JG. Epithelial-myoepithelial carcinoma of intercalated duct origin. A clinicopathologic and ultrastructural assessment of sixteen cases. *Oral Surg Oral Med Oral Pathol* 1982;53:280–7. [CrossRef]
9. Erkan AN, Bal N, Caylakli F, Kiroğlu F. Epithelial-myoepithelial carcinoma of the submandibular gland: a case report. *Kulak Burun Bogaz Ihtis Derg* 2007;17:167–70.
10. Pierard S, Gregoire V, Weynand B, Machiels JP. Epithelial-myoepithelial carcinoma of the submandibular gland with symptomatic lung metastases treated with chemotherapy. *Eur Arch Otorhinolaryngol* 2006;263:1158–60. [CrossRef]