

# A Rare Case of Humerus Metastasis From Acinic Cell Carcinoma of Parotid Gland

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Received: 02 May 2017; Accepted: 18 Dec. 2017

**Abstract-** Seventy-five percent of salivary gland tumors occur in parotid tumors. These tumors are mainly (75% to 85%) benign and around 25% turn out to be malignant. The most common cases of malignancy are mucoepidermoid carcinoma and adenoid cystic carcinoma (they totally represent half of the malignant tumors). Acinic cell carcinoma (ACC) is not of high incidence and is often seen in major salivary glands, particularly parotid gland. ACC makes up for 1-3% of salivary gland tumors and 3% of total parotid tumors. It is a low-grade malignant tumor with metastatic and invasive ability. Advanced stage, painful and fixed tumor, desmoplasia, anaplasia and differential character of the tumor, high mitosis, and necrosis, and nerve invasion, incomplete removal of the tumor, large size, and invasion to the deeper lobe of parotid worsen the prognosis of this tumor. We present a case report of a 52-year-old man with ACC metastasis to superior left humerus 15 years after surgery.

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*Acta Med Iran* 2018;56(8):547-550.

**Keywords:** Humerus; Metastasis; Acinic cell carcinoma; Parotid gland

## Introduction

Seventy-five percent of salivary gland tumors occur in parotid tumors (1). These tumors are mainly (75% to 85%) benign, and around 25% turn out to be malignant (1). The most common cases of malignancy are mucoepidermoid carcinoma and adenoid cystic carcinoma (they totally represent half of the malignant tumors). Acinic cell carcinoma (ACC) is not of high incidence and is often seen in major salivary glands, particularly parotid gland (2).

ACC makes up for 1-3% of salivary gland tumors and 3% of total parotid tumors (2,3). It is a low-grade malignant tumor with metastatic and invasive ability (although the incidence of remote metastasis is lower in head and neck than in other parts). It was classified as a benign tumor until in 1953 Buxton showed for the first time its metastasis and relapse. Spiro introduced two cases of ACC relapse, 30 years after primary surgery (3). Metastases mainly involved bones and axial skeleton. Its acinic name is due to the glandular shape of tumoral cells (4). The ACC treatment includes full excision of the parotid gland and sometimes radiotherapy (5,6).

Advanced stage, painful and fixed tumor,

desmoplasia, anaplasia and differential character of the tumor, high mitosis, necrosis, nerve invasion, incomplete removal of the tumor, large size, and invasion to the deeper lobe of parotid worsen the prognosis of the tumor (7).

In our research, no ACC metastasis to humerus was observed, and therefore we present a report of a 52-year-old man with ACC metastasis to superior left humerus 15 years after surgery.

## Case Report

A 52-year-old man with pain in his superior left arm referred to hospital. His pain started three months before and gradually intensified. Common painkillers failed to calm his pain. The patient had no fever or other clinical symptoms. He had a history of left parotid gland surgery 15 years ago, but he did not have his pathology report to present. The examination of his superior left arm showed tenderness. Paraclinical tests were natural, and only his CRP was reported at 103 mg/dl (Normal range <10). Report of MRI (his left shoulder and arm) were a T1 low T2 high signal mass lesion involving the humeral neck and proximal shaft with a length of approximately 70 mm

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## Humerus metastasis from acinic cell carcinoma

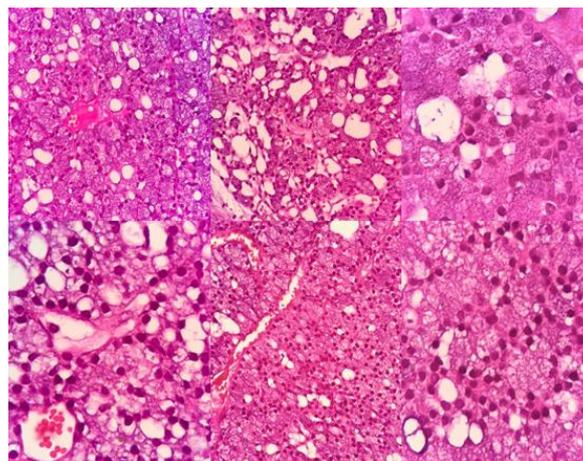
associated with a break in the anterior cortex of the neck with a soft tissue component extending outside displacing the biceps tendon (Figure 1). Report of CT Scan (his left shoulder and arm) was intramedullary soft tissue mass lesion centrally located in metaphysis and proximal shaft of humerus at a segment of about 70 mm length extending mildly to the head and was accompanying cortical thinning with some areas of cortical resorption and disruption as well as mild extra-cortical extension in medial aspect (Figure 2). Whole body bone scan after IV administration of 740 MBq 99 m Tc-MDP showed zones of increased flow and local hyperemia in the angiographic and blood pool phase in the upper arm. In delayed (3<sup>rd</sup>) phase of the study ,whole body bone scanning was carried out and demonstrated abnormally intense increased radiotracer uptakes in the proximal end of the left humerus.



**Figure 1.** A T1 low T2 high signal mass lesion involving the humeral neck and proximal shaft with a length of approximately 70 mm associated with a break in the anterior cortex of the neck with a soft tissue component extending outside and displacing the biceps tendon



**Figure 2.** Intramedullary soft tissue mass lesion centrally located in the metaphysis and proximal shaft of the humerus at a segment of about 70 mm length is seen extending mildly to the head which is accompanying cortical thinning with some areas of cortical resorption and disruption as well as mild extra-cortical extension in the medial aspect



**Figure 3.** Microscopic examination shows a neoplasm composed of neoplastic polygonal acinar cells with relatively abundant basophilic granular cytoplasm and uniform round basally located nuclei which are arranged in sheets with foci of microcystic formation and delicate vascular spaces in between tumor cells. In some foci, the neoplastic cells with cytoplasmic foamy appearance are seen. The tumor cells cytoplasmic granules are PAS positive and diastase resistant

Afterward, the patient underwent surgery, and the lesion in his superior left arm was removed. In the macroscopic exams, several cream-brown irregular pieces, measuring 4x3x2, underwent processing. The microscopic exam showed that the lesion was a neoplasm composed of neoplastic polygonal acinar cells with relatively abundant basophilic granular cytoplasm and uniform round basally located nuclei which are arranged in sheets with foci of microcystic formation and delicate vascular spaces in between tumor islands. In some foci, the neoplastic cells with cytoplasmic foamy appearance are seen (Figure 3). The tumor cells' cytoplasmic granules were PAS positive and diastase resistant and based on these findings; the ACC diagnosis was finalized.

## Discussion

ACC is an uncommon tumor in the salivary glands. It constitutes only one percent of total salivary neoplasms and 10-17 percent of malignant neoplasms in the salivary glands. Parotid glands are the most common site for these tumors (81-98%) after submandibular glands (11%) and minor salivary glands (3-12%). ACC was considered benign until 1953 when Buxton showed that this tumor was metastatic and recurrent (8,9).

Previous studies showed that this tumor was recurrent between 8.3% and 45% (9,10). Between 4% and 16% of these tumors had struck lymph nodes and between 2.6% and 14% metastasized to other areas (10). The standard treatments for these tumors were total parotidectomy

while preserving facial nerve (8,10). Radiotherapy has been proposed as an adjuvant treatment when the tumor has not been fully removed. ACC has so far been reported to have metastasized to the liver, lungs, orbit, and neck lymph nodes (11). ACC remains a malignant tumor with a low rate of incidence. In previous studies, there has been report of recurrence 30 years after the first diagnosis of malignancy (11). Metastasis to distant areas has been mainly seen in axial skeletons particularly in thoracic vertebrae. Previous studies had reported two rare cases of metastasis to the vertebral column.

The previous study shows a case of metastasis to cervical vertebrae in a post-surgery recurrent ACC (11). Vidyadhara *et al.*, described an ACC recurrence and its metastasis to T4 that had caused myelopathy (10). Recently a sort of salivary gland tumor, known as MASC (Mammary analog secretory carcinoma), with ETV6-NTRK3 translocation is reported (12,13).

MASCs are classified as adenocarcinomas, not otherwise specified (ANOS) or acinic cell carcinomas by the current World Health Organization classification (13,14). MASCs can be differentiated from acinic cell carcinomas by a lack of periodic acid-Schiff diastase-positive zymogen granules and S-100 protein positivity (13).

ACC happens in adults aged over 60 at 35% while 33% of cases happen in people aged between 40 and 59 (3). The ratio of ACC incidence in women to men is 59.5% to 40.5% (3). In patients with local disease, the survival rate of 5, 10, and 20 years stand respectively at 100%, 99.15%, and 94.37%. Furthermore, in the patients with metastasis, the survival rates were respectively 59.24%, 31.52%, and 21.99%.

A 20-year survival in patients with good prognosis was 97.79%, in those with quite good prognosis was 83.33% and in those with weak prognosis was 38.06% (8).

ACC has four morphological patterns (10), identified as below: solid, microcystic, papillary cystic, and follicular. These patterns may emerge alone or combined. In terms of morphology, in ACC tumor cells are polygonal and have basophilic and granular cytoplasm. In addition to serous cells, clear cells, intercalated duct cells, vacuolated cells, and nonspecific glandular cells may be seen. Zymogen-style cytoplasm granules are key to ACC diagnosis. Proteinaceous deposits and calcifications were infrequently reported by immunohistochemistry. The tumor cells are positive for cytokeratin and  $\alpha$ -1 antichymotrypsin and 1-antitrypsin markers (10).

Clinically, patients with major salivary gland tumor often refer to a tumor or a painless inflammation. The

presence of a parotid mass along with facial nerve paralysis generally indicates a malignant rather than a benign tumor (11). Minor salivary tumors inside the mouth may appear in the form of a submucosal mass or a mucus injury. The lymphatic drainage of these tumors varies depending on the location of salivary glands. In parotid malignancies, intraparotid is the first place to see metastasis, followed by the levels I and II of the lymphatic nodes of the neck (12).

Submandibular salivary gland tumors first metastasize to adjacent nodes before extending to the neck lymphatic glands. Sublingual gland extends into submental chin and submandibular lymph glands and minor salivary glands into post-nasal lymph nodes. ACC is uncommon, but it has had delayed metastasis to farther areas. The most common areas to have suffered metastasis are lungs, bones, and liver. In the ACC patients, there is a high risk of contracting other malignancies like non-Hodgkin and renal cell carcinoma (12).

This report was a rare case of ACC metastasis to humerus bone. Based on this report, axial and even non-axial bone pains must be taken seriously in these people. Metastasis to far areas may happen even after years.

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