Primary Intraosseous Adenoid Cystic Carcinoma of the Jaw: Clinical and Histopathologic Analysis

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Purpose: Adenoid cystic carcinoma (ACC), formerly known as cylindroma, is a malignant epithelial neoplasm typically derived from the salivary glands. Of all salivary gland tumors, the incidence of malignant salivary gland tumor has been 15 to 32% in the parotid glands, 70 to 90% in the sublingual glands, and about 50% in the minor salivary glands. Intraosseous ACC of the jaw has rarely been reported and is poorly understood. The aim of the present study was to analyze this tumor clinically and histopathologically to improve the diagnosis, management, and treatment.

Materials and Methods: We collected the records of 16 patients with intraosseous ACC from 1998 to 2013, who had been treated at our hospital, including clinical data and follow-up information. We then analyzed the patients' clinical features, diagnosis, treatment, and prognosis.

Results: The average age of the 16 patients was 56.8 years, and the male/female ratio was 0.8. The primary manifestations of the tumor were obviously different. Tumor excision was performed and followed by radiotherapy or chemotherapy, or both. The average follow-up period was 57.2 months, and the average follow-up period for patients who were alive and tumor free was 52.3 months. The survival rate was 68.8% after treatment. All these results were generally in agreement with those from previous reports.

Conclusions: The differential diagnosis of intraosseous ACC from other common tumors of jaws should be determined by the clinical, radiographic, and histopathologic subtypes. For treatment, surgery is the first choice for patients, and radiotherapy or chemotherapy might improve the prognosis in the postoperative period. In addition, the histopathologic subtypes and biologic processes of ACC are related to patient prognosis.

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Table 1. PATIENT CHARACTERISTICS

Characteristic	Value
Age (yr)	
Mean ± standard deviation	56.8 ± 18.9
Range	24-82
Gender (n)	
Male	7
Female	9
Site (n)	
Maxilla	8
Mandible	
Anterior body	2
Posterior body	5
Ramus	1

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the sublingual glands, and about 50% in the minor salivary glands,[1-4] but this lesion has rarely affected the jaw.[5] The typical feature is a painless smooth neoplasm; however, it can invade adjacent nerves and vessels. Unlike most carcinomas of the head and neck, regional lymph node metastases of ACC have been rare, although distant metastases have been common. We might not conclude that the short-term prognosis of ACC was much better than that in the long term.[6,7] ACC has been characterized by indolent growth, metastasis, and a poor prognosis.[8]

Central malignant salivary gland tumors of the jaw are rare, constituting less than 0.4% of all salivary gland carcinomas. Martinez-Madrigal et al[9] described 16 patients with primary salivary gland carcinomas arising centrally within the mandible who were treated at the University of Texas MD Anderson Cancer Center and the Institut Gustave Roussy from 1950 to 1990. Of the 16 patients, 5 had histologic types of carcinoma diagnosed, 7 mucoepidermoid carcinoma, and 4 adenoid cystic carcinoma.[9] Intraosseous adenoid cystic carcinomas of the jaws are extremely rare,[10] with only approximately 26 previously reported cases of ACC arising centrally within the mandible as of 2009.[11] The first case of central ACC within the mandible was reported in 1955 by Bumsted.[12] Mano et al[8] reviewed 17 cases from published studies in 2010 and simultaneously reported an unusual case of intraosseous ACC with multiple bone metastases.

Table 2. PRIMARY SYMPTOMS AND DIAGNOSES

Variable	Patients (n)
Primary symptom	
Swelling	8
Toothache	4
Paresthesia	2
Epistaxis	1
Pain	1
Preoperative diagnosis	
Cyst	1
Ameloblastoma	3
Malignant tumor	6
Squamous cell carcinoma	2
Adenoid cystic carcinoma	4
Histologic type	
Cribriform	8
Tubular	2
Solid	6

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The origin of the tumor is still controversial, and two hypotheses have been proposed regarding its histiogenesis: *1)* it arises from ectopic mucous glands and is entrapped within the bone of glandular remnants during development; or *2)* it arises from the neoplastic transformation of mucus cells derived from the odontogenic epithelial lining of cysts.[4,5,9,13,14]

Intraosseous ACC can mimic odontogenic cysts and malignant tumors in terms of the clinical and radiographic subtypes. Therefore, a final definitive diagnosis can only be obtained by histopathologic examination.[9] Such a histopathologic examination should be considered to differentiate ACC from cystic lytic lesions and other malignant tumors.

The aim of the present study was to improve the diagnosis, management, and treatment of ACC by analyzing the data from 16 patients who were treated at the Shanghai Ninth People's Hospital.

Materials and Methods

The present study was a retrospective analysis, and the independent ethics committee of Shanghai Ninth People's Hospital, affiliated with Shanghai Jiao Tong University School of Medicine, approved the study. We also followed the Declaration of Helsinki guidelines in the present investigation. We included all hospitalized cases of intraosseous ACC at Shanghai Ninth People's Hospital from 1998 to 2013. The clinical data, histopathologic materials, surgical records, and follow-up information were analyzed.

Strict diagnostic criteria were established to confirm the central origin of salivary gland tumors involving the jaw. All of the following diagnostic criteria were satisfied in all patients: *1)* radiographic evidence of osteolysis, *2)* clinical and radiographic presence of intact cortical plates, *3)* an absence of any primary lesions within the major or minor salivary glands, and *4)* histologic confirmation of the typical architecture and morphologic features of ACC.[4,5,9,13-15]