CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 5 (2014) 330–334

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Central papillary cystadenocarcinoma of the mandible: A case report and review of the literature

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ARTICLE INFO

Article history:
Received 26 March 2014
Accepted 3 April 2014
Available online 13 April 2014

Keywords:
Papillary cystadenocarcinoma
Central
Intraosseous
Mandible
Salivary gland neoplasm

ABSTRACT

INTRODUCTION: Central papillary cystadenocarcinoma of the jaw is an extremely rare tumor with only three previously reported cases in the English literature. This tumor is a histologically low-grade cancer, affecting more commonly in the mandible than in the maxilla.
PRESENTATION OF CASE: A 65-year-old woman presented with a two months history of a rapidly growing, painless mass of the right ascending ramus of the mandible. The pathologic report from incisional biopsy was a papillary cystic tumor with a differential diagnosis of cystadenoma versus cystadenocarcinoma. Segmental mandibulectomy, parotidectomy and submandibular gland resection were performed. The final pathology was intraosseous papillary cystadenocarcinoma.
DISCUSSION: Clinical features of central papillary cystadenocarcinoma of the mandible mimic an odontogenic lesion and metastatic bone disease, careful review of radiograph and pathology should be done. Surgical excision with wide margins is the appropriate treatment. Postoperative radiation therapy should be considered in histologically aggressive or high-stage tumor.
CONCLUSION: This is the fourth case of central papillary cystadenocarcinoma of the mandible in the English literature. Although it is usually a low-grade cancer, en bloc resection with adjuvant postoperative radiotherapy in a high-stage disease, and long-term follow-up allow the patient to have a favorable prognosis.

1. Introduction

Malignant salivary gland neoplasms that occur primarily in the mandible are rare, with most of them being mucoepidermoid carcinoma.[1,2] Papillary cystadenocarcinoma of the salivary gland is uncommonly found[3] and when it originates intraosseously in the mandible, only three cases have been previously reported.[2,4,5] Histologically, the tumor is characterized by a predominantly cystic and invasive growth pattern with a papillary component and defined as a low-grade malignant glandular tumor.[3,6] The radiologic finding consists of a unilocular or multilocular radiolucency mandibular lesion which mimics an odontogenic lesion and metastatic bone disease.[2,4] The authors report the fourth case, and discuss clinical features and management of central papillary cystadenocarcinoma of the mandible.

2. Presentation of case

A 65-year-old woman presented with a two months history of a rapidly growing, painless mass of the right face without trismus, facial or trigeminal neuropathy, or other neck mass. Her history included controlled hypertension and a left thyroid lobectomy for toxic adenoma several years ago. Physical examination revealed a hard consistency mass about 7 cm × 7.5 cm over the right mandibular ramus (Fig. 1). Fine needle aspiration biopsy (FNAB) was performed but the result was unsatisfactory. Computed tomography scan showed a large multilocular low density area at the ascending ramus of the mandible attached to the parotid gland and an ill-defined border between submandibular gland and the mass (Fig. 2). An open incisional biopsy was performed and the pathological report was a papillary cystic tumor with a differential diagnosis of cystadenoma versus cystadenocarcinoma.

Segmental mandibulectomy, parotidectomy and submandibular gland resection were performed (Fig. 3). Neck dissection was not performed because no lymphadenopathy was detected in CT scan. The final pathology was intraosseous papillary cystadenocarcinoma of the mandible with both major salivary glands being normal tissue. Microscopically, the tumor contained multiple cystic

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http://dx.doi.org/10.1016/j.ijscr.2014.04.006

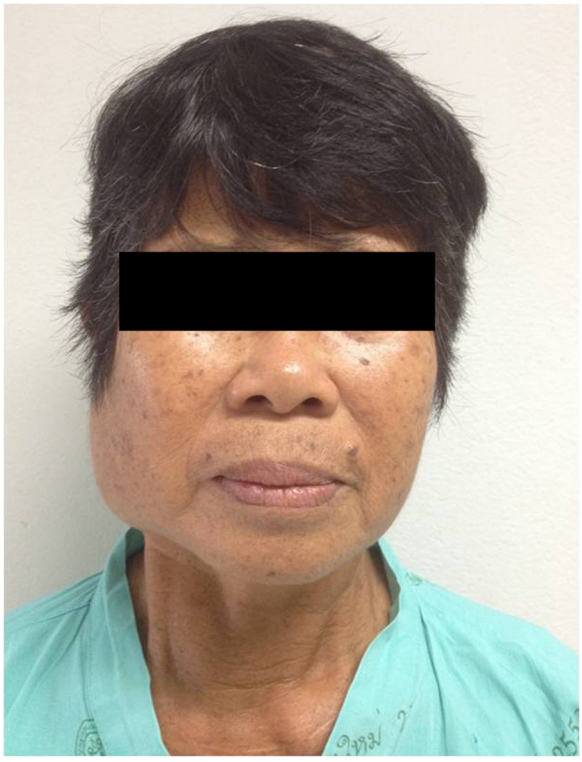

Fig. 1. A 65-year-old woman with a hard consistency mass at right mandible.

cavities with papillary projection. The neoplastic ducts infiltrate into the bone and surrounding skeletal muscle. The lining epithelia are mostly single layer of cuboidal cells with mild cellular and nuclear pleomorphism. The mitotic activity is low. The necrosis and perineural invasion is not present (Fig. 4).

The tumor cells are immunoreactive for CK7, CK19 and CEA but not for CDX-1, CK20, CA-125 and TTF-1.

The diagnosis was central papillary cystadenocarcinoma of the mandible. Post-operative radiotherapy with a total dose of 66 Gy (2 Gy per fraction) at the primary site and upper neck (levels I–III) was administered because of the large tumor size with cortical bone erosion although the surgical margins were negative and perineural invasion and high grade features were absent.

One year after surgery and ten months post-radiation therapy, the patient has no tumor recurrence both at the primary and neck (Fig. 5).

3. Discussion

Papillary cystadenocarcinoma of the salivary gland described by the WHO in 1991 is extremely rare.[3,7] The tumor is also called malignant papillary cystadenoma, atypical type of adenocarcinoma, low-grade papillary adenocarcinoma, and mucous-producing adenopapillary carcinoma.[3,8] This tumor is more commonly found in the major salivary glands (65%) and most often occurs in the parotid gland (95%).[6] The minor salivary glands involved by the tumor are found in the lip, buccal mucosa, palate, tongue and retromolar trigone.[6] Tumor histology is characterized by a cyst and papillary endocystic projection and defined as a low-grade glandular tumor with an indolent behavior.[3,6] However, the presence of intense nuclear and cellular pleomorphism and numerous mitosis with a rapidly progressive clinical course indicate a high-grade malignant tumor.[3]

Central or primary malignant salivary gland neoplasm of the jaw is a rare neoplasm arising intraosseously.[1,4,5,9] The tumor is

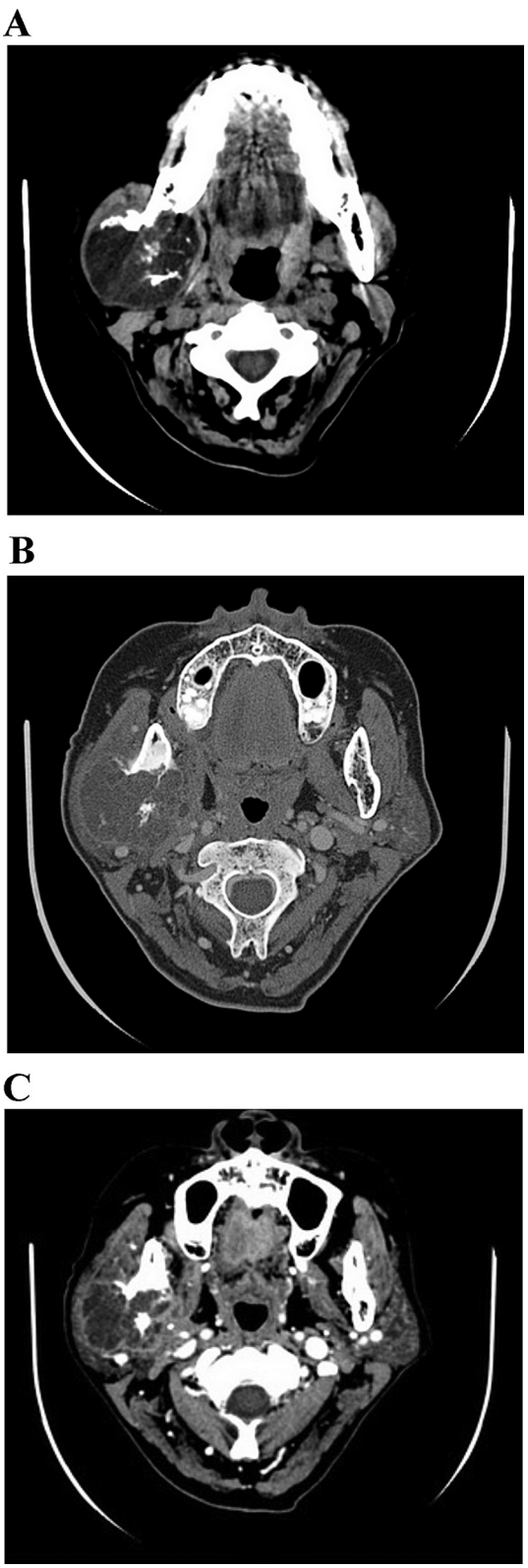

Fig. 2. Axial non-contrast enhanced CT scan of the mandibular ramus shows a circumscribed margin, multilocular cystic mass at the right side with internal coarse, irregular calcifications (A). Bone window image reveals a very thin bone around the expansile mass (B). The tumor has contact surface with the right parotid gland and caused lateral displacement of the gland in a contrast enhanced film (C).