CASE REPORT

Atypical case of primary intraosseous adenoid cystic carcinoma of mandible

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ABSTRACT

The primary central salivary gland neoplasms of the mandible are infrequent. Their clinical and radiographic features may be similar to odontogenic tumors, which are otherwise common. Their accurate diagnosis becomes troublesome. Hence, diagnosis should depend on stringent diagnostic criteria. Adenoid cystic carcinoma is well known for its prolonged clinical course and its tendency for delayed onset of distant metastases. The long-term survival of these patients is therefore poor. Treatment modalities include surgery, radiotherapy and chemotherapy. The purpose of this paper is to report a case of primary central adenoid cystic carcinoma of mandible with an atypical presentation.

Key words: Adenoid cystic carcinoma, central salivary gland carcinomas, mandible

INTRODUCTION

Salivary gland carcinomas located centrally within the mandible are rare, comprising less than 0.4% of all salivary gland carcinomas. Mucoepidermoid carcinomas are the most frequently reported types of primary central salivary gland carcinomas of the mandible followed by adenoid cystic carcinomas (ACC), adenocarcinomas and acinic-cell carcinomas. ACC is so rarely seen in the jaw bones usually occurs in the posterior mandible of adults in 4th to 6th decade, causing pain due to peri-neural invasion. It is well known for its prolonged clinical course and its tendency for delayed onset of the distant metastases. When such neoplastic transformation occurs centrally within the mandible, the radiological and clinical presentations may mimic odontogenic tumors making their diagnosis troublesome. Hence, regardless of the histology the diagnosis should rely on the application of strict diagnostic criteria.

We hereby report a case of primary intraosseous ACC atypically involving whole of the mandible with no distant metastasis in spite of its unusual size. Literature search on pubmed did not reveal any ACC case with such an extensive involvement of mandible.

CASE REPORT

A 64-year-old male patient reported to the outpatient department with the chief complaint of pain in the lower jaw. There was history of pain since five months, which was resistant to analgesics. Clinical examination revealed a slight, diffuse swelling throughout the buccal aspect of mandible. Gingiva appeared inflamed due to chronic generalized periodontitis with no other evident change in the mucosa. Submandibular lymph nodes were palpable but non-tender. Panoramic radiograph and intraoral periapical radiographs showed an irregular lytic lesion involving almost whole of the mandible. All teeth were vital.

The possibility of a malignant tumor was considered based on the clinical findings. On performing incisional biopsy, histopathological findings showed predominantly solid, tubular and cribriform pattern in very few areas. The tumor cells were basaloïd in nature with scanty cytoplasm and oval to round nucleus. Peri-neural invasion was seen; and surrounding stroma was dense. Majority of the areas (>30%) showed solid pattern, hence the diagnosis of solid adenoid cystic carcinoma was made. To rule out the possibility of local and distant metastasis, subsequent examination including ultrasound scanning (USG), computed tomography (CT), chest radiograph and bone scan was advised. Distant metastasis was ruled out with no other lesions of the major or minor salivary glands.

Considering the size and bilateral extension of tumor, subtotal mandibulectomy (sparring the condylar stumps)
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along with bilateral suprathyroid neck dissection was planned as tumors >4 cm have been associated with an unfavorable outcome.[4] Due to its peculiar clinical presentation, adequate soft tissue margin was removed prophylactically on both buccal and lingual aspect of the lesion. Sublingual and submandibular salivary glands were also removed bilaterally. Stabilization was done using 2.7 mm titanium reconstruction plate.

Histopathology of the excised specimen showed intraosseous involvement of solid variant of adenoid cystic carcinoma [Figures 2 and 3], lymph nodes and excised salivary glands were negative. A final diagnosis of solid intraosseous ACC was given. Postoperative radiotherapy was given. Follow-up after 15 months since the initial diagnosis showed no sign of recurrence.

DISCUSSION

Central salivary gland neoplasms are extremely rare, ACC (12 cases) is the second most common histological subtype in the mandible preceded by mucoepidermoid carcinoma (60 cases). The most common site is posterior body or angle of the mandible.[5] In 1979, Batsakis proposed a diagnostic criteria for central salivary gland neoplasm: (1) radiographic evidence of osteolysis, (2) presence of intact cortical plates, (3) presence of intact mucous membrane overlying the lesion, (4) absence of any primary tumor within major or minor salivary gland, (5) histological confirmation of the typical architecture and morphological features of a salivary gland tumor.[6] In our case, it was thought that the tumor first occurred in the mandible, and later expanded widely throughout the medullary spaces of the mandible.

The pathogenesis of central salivary gland neoplasms is unknown. Some theories propose the following: (1) enclaved retromolar mucous glands during embryological development of the jawbones, (2) submandibular and sublingual glands closely apposed in bony defects or cavities in the lingual cortex of the mandible, or (3) fragments of submandibular and sublingual glands that have undergone embryologic evagination.[5,7,8]

Histologically, the cribriform or tubular growth patterns is associated with a better prognosis, in contrast presence of (>30-50%) solid areas indicates an aggressive clinical course.[4] The present tumor belonged to ACC category with predominance of solid pattern. Reports suggest that clinical staging is a more reliable prognostic tool than histologic grading.[2]

According to Brookstone and Huvos, lesions that are located within undisturbed, intact cortical bone and overlying periosteum and that show no signs of cortical expansion offer the best prognosis and therefore indicate stage I disease. Stage II disease is characterized by lesions surrounded by intact cortical bone that has undergone some degree of expansion. An instance of cortical perforation, breakdown of the overlying periosteum or nodal metastatic spread is categorized as stage III disease.[7]
Radiographically, the lesions, in general, are poorly defined and have infiltrative margins. The center of the lesion, as was the case with our patient [Figure 4], usually show low density on the computed tomography (CT) scan and radiograph. ACC infrequently occupies almost the whole area of a sparse component without any influence on surrounding structures, as seen in our case. The changes on magnetic resonance (MR) images are characterized by an increase in intensity with maintenance of the shape. CT and MRI play a pivotal role in predicting the peri-neural spread and submucosal extensions. The differential diagnosis for ACC should include polymorphous low-grade adenocarcinoma (PLGA), basaloid squamous carcinoma (BSC), mucoepidermoid carcinomas, acinic cell carcinoma and malignant mixed tumors, adenocarcinoma and squamous cell carcinoma.

PLGA shows similar histological patterns (solid, cribriform and tubular), growth pattern and peri-neural spread. Polymorphous architecture, single file/single cell infiltration and foci of papillary growth are some of its main characteristic features. Histological features of high grade i.e. increased mitotic figures, coarse chromatin, apoptosis and necrosis are commonly associated with ACC (solid) than PLGA. Solid ACC also needs to be differentiated from BSC. Both tumors produce basement membrane like material, but the latter tends to dissect between tumor cell rather than forming cribriform spaces. Necrosis and basaloid cells with prominent nucleoli and coarse chromatin are common features, although single cell necrosis, rapid mitotic rate and greater degree of nuclear atypia are more frequent in BSC.

Surgery is the predominant treatment modality for central salivary gland tumors of the mandible. It ranges from enucleation or curettage to en bloc or radical excision, but no method of treatment reduces the potential for recurrence. Postoperative radiation therapy enhances local and regional control in ACC. Our patient is being regularly followed up for any evidence of recurrence or distant metastases since 15 months from the original diagnosis. However, long-term follow-up is indispensable regardless of the site because of the tumor’s susceptibility for late recurrence and metastasis.

In the present case, ACC had spread to involve almost whole of the mandible. In spite of the size, no metastasis was discovered, which otherwise, is a common finding in such cases. The extensive involvement of the mandible could possibly be due to the insidious clinical course that characterizes ACC as it infiltrates nerves, peri-neural spaces and tissue planes.

**CONCLUSION**

Although salivary gland tumors arising within the mandible are uncommon, their significance should not be minimized. Because of their unique morphology and clinical behavior, they should be considered in the differential diagnosis of aggressive lesions of the mandible. If intraosseous ACC is suspected, a multidisciplinary diagnostic approach should always be adopted. Although in our case, there is no recurrence after 15 months, long term follow-up is essential for ACC in order to avoid metastasis in addition to recurrence of this lesion. This case underlines the importance of early detection, total surgical resection and long-term follow-up as it is essential in clinical management due to their higher recurrence rate.

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