Sarcomatoid carcinoma of the mandible: report of a case

Gui-Young Kwon, Young-Jun Choi, Min-Seok Song, Kyoung-In Yun
Departments of Pathology, and Oral and Maxillofacial Surgery, College of Medicine, Chung-Ang University, Seoul, Korea

Abstract

Sarcomatoid carcinoma is a rare and occurs mainly in the upper aerodigestive tract such as the oral cavity, esophagus and vocal cords. It is a unique variant of squamous cell carcinoma. We report the case of a patient with spindle cell squamous cell carcinoma involving the mandible. At initial examination, overlying mucosa of that lesion was normal appearance. One week later, that lesion showed ulcerative and bloody change and rapid growth in size. This case showed unpredictable rapid growth although rapid growth in size was suspected of undergoing malignancy.

Key words: Sarcomatoid carcinoma, Mandible, Mandibular neoplasms

Ⅰ. Introduction

Sarcomatoid carcinoma is a rare and occurs mainly in the upper aerodigestive tract such as the oral cavity, esophagus and vocal cords. It is a unique variant of squamous cell carcinoma. We report the case of a patient with spindle cell squamous cell carcinoma involving the mandible. At initial examination, overlying mucosa of that lesion was normal appearance. One week later, that lesion showed ulcerative and bloody change and rapid growth in size. This case showed unpredictable rapid growth although rapid growth in size was suspected of undergoing malignancy.

Ⅱ. Report of the case

An 80-year-old male visited to our hospital because of painless submandibular swelling. The swelling appeared 2 weeks ago and decreased after lower teeth extraction. His medical history included surgical treatment for cerebral abscess after admission one month ago. The patient didn’t take any medicine. He was not a smoker.

On intraoral examination, tooth extracted area was completely healed state. Oral mucosa and gingiva appeared normal color and texture but a hard mass was palpable on right lower vestibular area. The tongue and floor of mouth was not elevated. On extraoral examination, a hard submandibular mass was palpable but overlying skin was normal appearance. On panoramic view, any pathologic findings were not showed. We recommended further radiographic examinations to him and his family but that was refused by him.

One week later, he was referred from the department of neurosurgery to evaluate the right lower gingival mass after admission. He was admitted to the department of neurosurgery for seizure and abnormal behavior two days before. Clinical examination revealed the swelling of right lower edentulous area with hematoma and elevation of the tongue and mouth floor. (Fig. 1. A) The patient complained of continuous bleeding and foul odor. He could not seal the lips because of a protrusive mass. (Fig. 1. B)

Computerized tomography showed that 5.3 × 3.2 cm sized, relatively well marginated, heterogeneous mass was noted in anterosuperior aspect of the right mandibular body and focal cortical destruction was seen in right parasympysis of the mandible. (Fig. 2) A biopsy of the hematoma-like lesion was performed and showed a sarcomatoid carcinoma. (Fig. 3. A) The microscopic examination of biopsy showed an infiltrative mass consisting of diffuse tumor cells. The tumor cells had round to spindle cytoplasms with hyperchromatic, bizarre nuclei. (Fig. 3. B) The immunohistochemical stainings were performed and the tumor cells were immunoreactive for pan-cytokeratin and vimentin, supporting the diagnosis of sarcomatoid carcinoma. (Figs 3. C, D)
Unfortunately, he expired due to multiple metastatic carcinomas from the lung or stomach.

### Discussion

Sarcomatoid carcinoma is a subgroup of malignant mixed tumors and is extremely rare in maxilla. The initial description of this type of malignancy was reported in 1864 by Virchow, who labeled it as carcinosarcoma. Later on, Saphir and Vass analyzed 153 cases of carcinosarcomas of various sites. They concluded that the sarcomatous component represented a variation in the squamous portion of the carcinoma or an inflammatory reaction of the underlying stroma.

Numerous hypotheses regarding the histogenesis of this type of tumor have been proposed. Three dominant pathogenetic theories have been proposed: the tumor (1) represents a “collision tumor” (carcinosarcoma), (2) is a squamous cell carcinoma with an atypical reactive stroma (pseudosarcoma), or (3) is of epithelial origin, with “de-differentiation” or transformation to a spindle cell morphology (sarcomatoid carcinoma). Recently, the third hypothesis has been supported by following evidences: their occurrence in the exact sites that normally have squamous epithelium and a preponderance of carcinomas rather than sarcomas; a superficial location; a polypoid appearance; the direct continuity and smooth transition of the spindle cells with areas of squamous epithelium; immunoreactivity with epithelial antigens; a dual expression of epithelial and mesenchymal differentiation with double labeling techniques in some neoplastic spindle cells; and the presence of epithelial only, sarcomatous only, or a duality of expression in metastatic deposits from laryngeal sarcomatoid carcinoma. Furthermore, recent molecular studies have shown evidence of a monoclonal origin from a stem cell capable of divergent differentiation.

When the malignant surface epithelium is histologically evident, the diagnosis of a sarcomatoid carcinoma is made with confidence. However, when the surface epithelium is ulcerated or denuded, the correct diagnosis is more difficult. Furthermore, the biopsies from the squamous cell component tend to be misdiagnosed as squamous cell carcinoma and biopsies from spindle cell component tend to be diagnosed as sarcoma. In our case, the lesion looked like an ecchymosis on initial clinical examination.
Sarcomatoid carcinoma of the oral cavity presents a male predominance at a mean age of 57 years and site predilection for the lower lip, tongue and alveolar ridge or gingiva although most tumors in head and neck region occur in the larynx. Growth configuration is often exophytic polypoid, but sessile, nodular or endophytic configuration has also been described. The lesion usually has an extensive surface ulceration with friable, fibrinoid necrosis of variable thickness or shaggy exudates. Radiation, trauma, tobacco use or alcohol consumption seemed to play a role in etiological factors.

The differential diagnosis includes a number of benign and malignant tumors, such as squamous cell carcinoma, fibrosarcoma, malignant fibrous histiocytoma, leiomyosarcoma, rhabdomyosarcoma, malignant peripheral nerve sheath tumor, osteosarcoma, mesenchymal chondrosarcoma, Kaposi’s sarcoma, angiosarcoma, synovial sarcoma, malignant melanoma, fibromatosis, leiomyoma, nodular fasciitis and reactive epithelial proliferations.

Wide surgical excision is the treatment of choice. Most authors agree that irradiation is ineffective. Radiation therapy is considered an acceptable alternative for inoperable patients. Furthermore, adjuvant irradiation might be of benefit in cases in which the surgical margins are positive or in patients with nodal metastasis at the time of diagnosis.

Prognosis is related to location, tumor size, depth of invasion, stage of disease and with the presence of any keratin staining in the spindle cells. A tumor of the oral cavity and oropharynx is potentially aggressive and seems recur easily and to metastasize easily. The incidence of metastases was 36% and the 2-year survival rate was 55% in tumors involving the oral cavity.

References