CASE REPORT

Maxillary Carcinosarcoma: A Case Report and Review of the Literature

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ABSTRACT

We report on a patient who presented with pain and swelling to the left maxillary region of the face. A biopsy of the area was performed and subsequently diagnosed as maxillary carcinosarcoma (CCS). After reviewing the pathology results and the size of the tumor, it was decided that a radical dissection, followed by radiation therapy was the best option.

Keyword: Carcinosarcoma, Computed tomography, Sinonasal, Tumor.

How to cite this article: Sepúlveda I, Frelinghuysen M, García C, Spencer ML, Platín E, Alarcon J, Ulloa D. Maxillary Carcinosarcoma: A Case Report and Review of the Literature. *Int J Otorhinolaryngol Clin* 2014;6(3):114-117.

Source of support: Nil

Conflict of interest: None

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We report on a 66-year-old male patient who presented to the ENT service without a significant clinic history. The patient exhibited swelling in the left maxillary region of the face and palatine area that began 2 months earlier. Clinically, a necrotic tumor mass was observed in the nasogenian folds and periphery of the oral and orbital cavity.

A CT was acquired demonstrating a large expansive, invasive and heterodense process on the left half-face.

The mass seemed to originate from the maxillary region extending into the ipsilateral orbital, oral and nasal cavities (Figs 1 and 2). Postintravenous contrast injection, the mass was moderately enhanced revealing central necrotic areas (Figs 3 and 4).

The histopathology revealed fused cells with marked pleomorphism, atypical mitosis, myxoid degeneration and isolated multinucleated giant tumor cells. A sector showed an infiltrative superficial epithelial tumor with cords and nests; polygonal cell tumor involving the lateral margins (Fig. 5). The immunohistochemistry revealed a positive intense vimentin diffused in both components of the tumor, cytokeratin positive in isolated fused cells, positive diffuse in epithelial sector and negative for actin, desmin, CD 34 and S 100. In summary, the biopsy showed a malignant neoplasm with necrosis and characteristics of carcinosarcoma (CCS) (Figs 6 and 7).

Following review by the head and neck cancer committee that recommended radical excision of the tumor followed by radiation therapy. This recommendation was made based on the clinical findings, results of the biopsy and imaging studies. Also, the committee recommended enrollment of the patient in a pain management and palliative care program.

Subsequently a left maxillectomy was performed with orbital floor reconstruction. During surgery, infiltration of the orbital floor was noted, without apparent compromise of the orbital contents.

Conventional radiotherapy treatment consisted of a total dose of 66 Gy delivered to the affected area in 2 Gy increments during a 7-week period (Fig. 8). Following radiotherapy, an slight decrease in tumor size was noted. The patient reported pain, insomnia and a decrease in food intake consequently being referred to the pain and palliative care program.

DISCUSSION

Head and neck malignant neoplasms predominantly occur in the oral cavity, pharynx, ear, nose and larynx. Ninety to ninety-five percent of head and neck tumors correspond to squamous cell carcinomas.^{1,2} Sarcomas constitute less than 1% of all malignant neoplasms in the body including the head and neck region. Carcinosarcoma is rare and tends to be aggressive. It is a biphasic tumor

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Fig. 1: Axial CT bone window, showing a destructive, expansive process eroding the maxillary sinus walls

Fig. 2: Coronal CT soft tissues window demonstrating a hetero-dense maxillary mass compromising the subcutaneous muscle and fat

Fig. 3: Axial CT showing moderate enhancement after intravenous contrast, administration revealing areas of necrosis

Fig. 4: Coronal CT demonstrating the lesion expansion into the nasal cavity, buccal space and hard palate region

Fig. 5: HE stain

Fig. 6: Vimentin stain

comprising both a malignant epithelial (carcinomatous) and mesenchymal (sarcomatous) components.³⁻⁶ It is more prevalent in men than women with a ratio of 11:1 most often occurring between the ages of 60 and 70 years.

Carcinosarcoma is associated with snuff use, alcohol consumption, poor oral hygiene and exposure to radiation. It was first described by Virchow in the year 1864.^{1,2,7} Several theories have been postulated to explain

Fig. 7: Keratin stain

its origin but most believe that a multipotential cell, capable of both epithelial and mesenchymal differentiation are responsible for the origin of this rare tumor.^{1,9} This is a divergent differentiation of a single stem cell into epithelial and mesenchymal elements known as the monoclonal hypothesis.^{1,5,9-16,20} Convergent differentiation suggests this happens from two-cell lines known as the multiclonal hypothesis.^{5,14,16}

A well-accepted theory of the etiology is given by the transformation of carcinoma into sarcoma, as there is evidence that these lesions are originally epithelial and then acquire a potential to express a mesenchymal phenotype.¹ In the head and neck CCS involve mainly the salivary glands, parotid gland, larynx and the esophagus less frequently. Ultimately, it affects the oral cavity in the

following descending order, the lips, tongue and gingiva. Carcinosarcoma has also been reported in the thyroid, thymus, lung, breast, gastrointestinal tract, hepatobiliary system, genitourinary tract, uterus and skin.^{2,5,7,13,16-18}

The onset of signs and symptoms of CCS manifest quickly. In the maxillary region, patients experience localized pain, tooth mobility and burning sensation. As the tumor infiltrates the maxillary sinus, patients usually experience nasal congestion as the tumor obstructs the affected sinuses. In addition, the infraorbital nerve becomes affected causing sensory disturbances on the cheek. As the tumor progresses to the skull and reaches the floor of the orbit, it presses on the eyeball creating difficulty with mobility and its position in the orbital cavity. Also, it usually invades the ethmoidal area.^{2,14,19} The macroscopy usually shows a polypoid mass while the histology shows epithelioid (squamous) cells with pleomorphism and spindle cells arranged in nodular aggregates with central necrosis, cellular atypia and irregular hyperchromatic nuclei.^{1,2,14} Immunohistochemical examination results in the expression of vimentin and keratin in epithelial cells.^{1,2,5,8,13,14,16,17,20} Positive cytokeratin helps to establish the diagnosis of CCS.¹ Negative expression for smooth muscle actin has also been reported^{5,14} Regardless of treatment, CCS of the maxillary region tends to be aggressive with a high rate of recurrence.^{18,21}

The treatment of choice consists of surgical resection combined with adjuvant radiotherapy and chemotherapy. Reconstruction with the use of prosthesis to replace the resected tissue or tissue transfer is sometimes performed

Fig. 8: Tridimensional conformal radiation plan

depending on the extent of the surgery.^{2,5,14-17,19-21} Based on the stage of tumor, the 5 years survival rate ranges from 40 to 60%.^{16,19} Metastases to the lung occurs in 5% of cases and it has also been found in the brain, subcutaneous tissues, lymph nodes, pleura, diaphragm, bone and liver.^{1,5}

The prognosis is determined by the size, location, morphology, depth, presence of metastases, recurrence and prior treatment; however, it is very unfavorable regardless of treatment.^{1,2,16}

CONCLUSION

Carcinosarcoma is a rare and aggressive biphasic entity, with malignant epithelial (carcinomatous) and mesenchymal (sarcomatous) components. Imaginology is very important for diagnosis and planning treatment giving value information about growing pattern and extension to critical anatomic organs. The treatment of choice instead of scant literature, might consist of surgical resection combined with adjuvant radiotherapy and chemotherapy.

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