Case Report

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Second primary chondrosarcoma of the maxilla: a case report

Shreeya Pabi^{1*}, Yanpothung Yanthan¹, Sweety Gupta¹, Biswajyoti Das¹, Ajas Ibrahim¹, Naseef Kassim¹, Saurabh Varshney², Divya Singh³, Manoj Gupta¹

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*Correspondence: Dr. Shreeya Pabi,

E-mail: shreeya.pabi@gmail.com

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ABSTRACT

We are reporting an uncommon case of second primary chondrosarcoma of the maxilla in a 52-year-old male patient. Patient had first been diagnosed in 2004 with chondrosarcoma of the right maxilla. He had undergone right partial maxillectomy for the same. Since 2016 he had started noticing a swelling over the left side of the maxilla. Repeated biopsies from the growth showed no evidence of any malignancy. Hence, he was kept on routine follow up. However, in 2018, during follow up the growth had increased in size - hard swelling present in remnant post partial maxillectomy cavity, non-tender -and a punch biopsy was taken, revealing a well differentiated chondrosarcoma. A diagnosis of second primary chondrosarcoma of the maxilla was made based on Warren and Gates criteria. A brief discussion on the radiologic and histologic presentation of the tumor and the treatment modalities of this unusual tumor is discussed.

Keywords: Chondrosarcoma, Maxilla, Second primary, Adjuvant radiotherapy

INTRODUCTION

Chondrosarcoma is characterized by a neoplastic process with associated cartilage matrix production that is devoid of osteoid, a characteristic of osteosarcoma. It may arise at any age but typically occurs in middle-aged and older adults, in contrast to osteosarcoma. Chondrosarcomas of the head and neck region are rare malignancies, accounting for 1% of chondrosarcomas affecting the whole body. In the head and neck region, chondrosarcomas occur more commonly in the maxilla, nasal cavity, nasal septum, and mandible. Chondrosarcomas of the jaws are more common in men, with a male-to-female ratio of 2:1.

The second primary malignancy is defined as a second de novo malignant neoplasm with known cancer. Warren and Gates have given the criteria used for the diagnosis of second primary malignancies, they are as each of the tumors must be malignant, each confirmed by histology; each must be geographically separate and distinct. The lesions should be separated by normal mucosa or has to occur after 5 years of the first diagnosis; Probability of one being the metastasis of the other must be excluded.⁵

Clinical features

1% to 4% of chondrosarcomas affect the head and neck region, mainly the jawbones. The most common site affected is the anterior part maxilla and the posterior region mandible with a proportion of 1.75:1.⁶ It has been postulated that chondrosarcoma originates from remnants of embryonic cartilage precursors from nasal and septal development in the anterior part of the maxilla and from Meckel's cartilage precursors in the posterior aspect of the mandible.⁷

The most common clinical finding of is painless swelling, expansion of buccal and lingual plates, premature eruption

¹Department of Radiation Oncology, AIIMS Rishikesh, Uttarakhand, India

²Department of ENT, AIIMS Rishikesh, Uttarakhand, India

³Department of Pathology, AIIMS Rishikesh, Uttarakhand, India

or exfoliation of teeth.⁸ The mass is usually rapidly growing and covered with mucosa which can ulcerate and there can be pain at later stages. Rarely, there can be lymph node involvement. The symptoms vary from being painless to painful to headache and hearing loss with neurological problems depending on the tumor location. Jaw lesions may be associated with separation or loosening of teeth, expansion of cortical plates, and premature exfoliation of teeth.^{3,8}

Low-grade lesions have a <10% risk of metastases, intermediate-grade lesions have a 10% to 50% risk, and high-grade lesion have a 50% to 70% risk. 1 The lungs are the main site of metastases. Staging evaluation thus should include a chest computed tomography (CT) for intermediate- and high-grade lesions.

Histopathology

Histologically, chondrosarcoma continues to be defined as a malignant tumor composed of fully developed cartilage without tumor osteoid, being directly formed from a sarcomatous stroma. Myxoid changes, calcification and ossification may be seen. Evan's and co-workers have attempted to associate the histologic grade (grade I to III) of chondrosarcoma with the ultimate biologic behaviour of the tumour, depending on the cellularity, nuclei size, presence of mitotic figures, multinucleation, spindle cell formation and mineralization in the form of osseous development at the edge of the cartilaginous lobules.⁹

Radiological features

The radiological pattern of chondrosarcoma is variable. It includes single or multiple radiolucent areas. These lytic changes are prominent in more advanced cases. Other findings are opacification of air spaces, a densely calcified bone mass and root resorption. Also, it may reveal ground glass appearance or a sunburst appearance. Cortical destruction occurs late in the course of disease and periosteal bone formation is often limited. 10,11

Treatment and prognosis

The histologic grade and tumor location are important determinates of treatment approach. Surgical excision is the primary treatment modality for chondrosarcoma. For low-grade tumors, surgical resection alone is enough to achieve a high rate of disease control. This results in good local control rates. For the less common intermediate- and high-grade tumors, wide en bloc excision is the optimal surgical approach. Radiation therapy is indicated for incompletely resected high-grade or locally recurrent tumors and tumors that are unresectable. Doses of 50 Gy preoperatively and 60 to 66 Gy postoperatively for close or positive margins are typically used. Doses of ≥70 Gy are needed for definitive treatment (unresectable tumors).¹

We, hereby, present a rare case of second primary chondrosarcoma of the maxilla.

CASE REPORT

A 52 years old male patient presented to All India Institute of Medical Sciences (AIIMS), Rishikesh, with the complaint of swelling over the left side of the palate for the last two years. Patient had a history of swelling of the right maxilla 15 years back, in 2004, for which he had undergone Right Maxillectomy. His post-operative histopathology report showed: well differentiated chondrosarcoma grade 1. No adjuvant treatment was advised or taken. Following this, the patient was on regular follow ups every 3- 6 monthly. Patient was stable during follow up. In 2016 he had noticed a swelling over the left side of his hard palate. He then came to our institute where he was investigated for the same. Clinically, the mucosa over the hard palate was intact. Contrast enhanced computed tomography (CECT) of Face and Neck and Chest x-ray was taken as well. The CECT face and neck revealed a well-defined 2×2 cm sized lesion showing internal chondroid matrix seen arising from left maxillary alveolus placed between posterior wall of left maxillary sinus and ipsilateral pterygoid plates. Recurrent/residual lesion alternate benign bony lesion/ fibrous dysplasia, pansinusitis. Chest x-ray showed no evidence of any metastasis. An incisional biopsy was taken from the growth which was suggestive of inflammation. A repeat biopsy was done again which showed no evidence of any dysplasia or metaplasia. The patient was kept on routine follow up.

In May of 2018 he presented with complaints of increase in the size of the swelling over the left hard palate. On clinical examination of the oral cavity and oropharynx he was found to have a 1.5×1.5 cm hard, non-tender growth present in posterior part of the hard palate with induration extending anteriorly up to second molar, posteriorly up to retromolar trigine, medially 0.5 cm lateral to midline and laterally up to alveolus. Adequate mouth opening was present, dental hygiene was good and teeth in upper right alveolus were absent. Neck showed no swelling or palpable lymph nodes. Anterior rhinoscopy was performed and a palpable hard non tender swelling in left nasal cavity posteriorly was found.

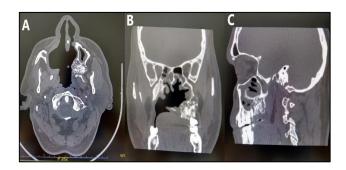


Figure 1: CECT PNS: axial section (A), coronal section (B), and sagittal section (C), showing a lytic expansile lesion in the left maxilla and mucosal thickening in bilateral maxillary and ethmoidal sinuses.

CECT of paranasal sinuses (PNS) was done which revealed a lytic expansile lesion in the left maxilla and mucosal thickening in bilateral maxillary and ethmoidal sinuses (figure 1). A punch biopsy was taken from the growth in the nasal cavity which was suggestive of recurrence of a well differentiated chondrosarcoma.

Patient was diagnosed as a case of second primary chondrosarcoma of the maxilla according to Warren and Gate's criteria. He was planned and taken up for surgery. He underwent left partial maxillectomy in July 2018. The post-operative histopathology of the resected specimen (figure 2) revealed well differentiated chondrosarcoma of the left hard palate, grade II; resected tumor was of the size: $2.3 \times 2 \times 2$ cm; superior and posterior margins were involved by the tumor, rest of the margins were free from any tumor. Pathological stage was pT1.

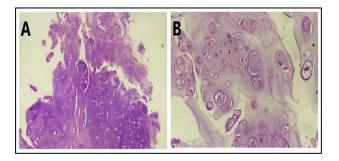


Figure 2: Microscopic images, (A) (100×) and (B) (400×), showing a lobulated cartilaginous tumor with increased cellularity having enlarged nuclei with irregular nuclear contours present in groups within hyaline cartilaginous stroma.

Patient was then planned and taken up for adjuvant radiotherapy to the post-operative tumor bed by Intensity modulated radiation therapy-volumetric modulated arc therapy (IMRT-VMAT) technique, 66 Gy in 33 fractions using linear accelerator (LINAC) photons. Patient had tolerated the treatment well without any significant toxicity.

DISCUSSION

Chondrosarcomas are uncommon malignant tumors. They are rarely found in the head and neck region accounting for 5.76% of all the cases of chondrosarcomas. Our case occurred in a 52-year-old male patient with complaints of a painless swelling over the hard palate, with intact mucosa. Initially, in 2004, his disease was on the right side of maxilla. In 2016, he had presented to us with a painless, slow growing swelling over the left side of the maxilla. Gross appearance of the swelling looked benign as the mucosal surface appeared smooth. Upon biopsy the report showed only inflammation, another repeat biopsy was done which showed hyperplasia. Going by these reports, we assumed that there was no malignancy. On routine follow up in 2018, patient had come back to us with the same complaint of a swelling and this time it was gradually

increasing in size. No swelling or palpable lymph nodes were seen over the neck.

A punch biopsy was taken which reported a well differentiated chondrosarcoma. Our case fulfilled all of Warren and Gates' criteria for a second malignancy, viz a viz a confirmed histology of the swelling (both from 2004 and 2018); both the swellings were geographically separate and distinct (right side in 2004 versus left side in 2018) and has been diagnosed after 5 years of the first diagnosis (in our case, second diagnosis happened after 14 years). Hence, we have termed out case as a second primary chondrosarcoma of the maxilla. Most of the limited data and literature available is for recurrent cases of chondrosarcoma of the maxilla.

In this case report we are going to discuss our experience with the clinical manifestations, imaging findings and our treatment strategy. To our knowledge, it is a first if its kind case report.

Upon work-up, he was found to have recurrent chondrosarcoma of the maxilla, non-metastatic. Clinical findings of the present case are consistent with the observations reported by authors of different studies.⁷⁻⁹ A case report by TC Huang was reviewed and the patient was of a much younger age and treated with upfront surgery. Post-operative biopsy showed well differentiated chondrosarcoma grade 1, hence adjuvant treatment was deferred. But patient returned with recurrence at the end of 9 months.7 However, our patient did not have any local recurrence, but was a second primary as per the Warren and Gates criteria. There is sufficient evidence regarding the role of field cancerization in squamous cell carcinoma of the oral cavity. In squamous cell carcinoma of the oral cavity the possible explanation could be that of lateral field cancerization which suggests the lateral spread of tumors, which occurs due to a progressive transformation of the tissue adjacent to the tumor rather than the expansion of pre-existing cancer cells into the adjacent tissue.⁵

Clinically, the chondrosarcomas of jaws are relatively invasive and destructive, similar to our case.

Our treatment strategy was to treat in lines of a recurrent chondrosarcoma of the maxilla. As most of the case reports have reported, recurrence had occurred within the first one year of radical surgery. 9 And if margins were free of tumor post-surgery, then no adjuvant radiation had been administered. Our patient had undergone left partial maxillectomy. Following this the post-operative histopathology of the resected specimen had revealed well differentiated chondrosarcoma of the left hard palate, grade II; superior and posterior margins were involved by the tumor. In view of margin positivity, patient was then planned and taken up for adjuvant radiotherapy to the postoperative tumor bed since the tumor involvement at the resected margins is the only other poor prognostic sign. We had prescribed a dose of 66 Gy in 33 fractions since the recommended doses for close or positive margins typically used was 60 to 66 Gy postoperatively. IMRT-VMAT technique was used and treatment was delivered using LINAC photons. Patient had tolerated the treatment well without any significant toxicity. Patient is currently on routine follow up every 3 monthly and is doing well clinically.

There is sufficient evidence regarding the role of field cancerization in squamous cell carcinoma of the oral cavity. In squamous cell carcinoma of the oral cavity the possible explanation could be that of lateral field cancerization which suggests the lateral spread of tumors, which occurs due to a progressive transformation of the tissue adjacent to the tumor rather than the expansion of pre-existing cancer cells into the adjacent tissue.¹²

CONCLUSION

Chondrosarcoma of the maxilla is a rare type of sarcoma. Unlike the expanding high-grade chondrosarcoma of the bones presenting with excruciating pain chondrosarcomas of head and neck tend to be painless on presentation. Examination often discloses a firm mass that has an intact mucosal covering, and our case was not different in this respect. Hence a thorough work-up including clinical history, correct imaging techniques and a thorough interpretation of the histopathology is important in order to reach a diagnosis. The most effective therapeutic modality is wide surgical excision. These tumors have a high chance of recurrence, even after attaining tumor-free margins post operatively. As reported in our case, there even stands a chance of a second primary appearing after several years of living disease free. The second primary was located on the opposite side of the same subsite. There is no literature regarding the role of field cancerization being the possible explanation for recurrences in chondrosarcomas of oral cavity tumors. Further investigations are needed to gain a better understanding.

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