



Full length article

Osteosarcoma of the jaw: Challenges in the diagnosis and treatment

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ABSTRACT

Purpose: Osteosarcomas rarely affect jaw bones. Patients are usually older than those who suffer long bone sarcomas, with a rare incidence of metastasis. This is suggestive of a different pattern of behavior compared with long bone sarcomas. This study aimed to present NCI, Cairo University experience in treating patients diagnosed with osteosarcomas of the jaw, including the diagnostic challenges and treatment outcome.

Patients and methods: This is a retrospective case series study of all cases of osteosarcomas of mandible and maxilla that were treated at the NCI, in the period between 2006 and 2013. Patients' data, including demographic data, various clinical presentations, results of investigations, treatment modalities performed and outcomes, were collected from hospital records kept in the Biostatistics Department at NCI. **Results:** Records showed 21 cases of osteosarcoma of the jaw. The mandible was affected in 15 cases, the maxilla in six. Two cases had sun-ray periosteal reactions. Erroneous biopsy results were found in 4 cases compared with final pathology reports of surgical resections. All cases underwent surgical resections, with 8 cases having positive margins. The median follow-up period was 19.3 months (range 0.3–98.0 months). The cumulative disease-free survival (DFS) was 27.5% and the median DFS was 72 months. The cumulative overall survival at end of the study was 77.4%.

Conclusions: Osteosarcoma of the jaw is challenging both to diagnose and manage. This is due to the high incidence of mistakes in biopsy results, rare specific radiological features and difficulties in proper resection due to proximity to vital structures.

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Introduction

Bone tumors are rare malignancies. According to the Surveillance, Epidemiology, and End Results Program's (SEER) registry, they represent only 0.2% of new cancer cases in the USA with an estimated 3260 new cases to be diagnosed in the year 2017 [1]. Although they are rare tumors, they are the seventh most common cancer in childhood [2] and they have high mortality rates. In the USA, it is estimated that 1550 deaths will occur in the year 2017 due to bone tumors [1]. Osteosarcoma is the most common malignant bone tumor (excluding bone marrow tumors), with mandibular and maxillary osteosarcomas accounting for only 6% of all osteosarcomas [3]. Mandibular and maxillary osteosarcomas usually afflict patients 10–20 years older than those afflicted by long

bone osteosarcomas and have lesser incidences of distant metastasis [4]. This suggests that mandibular and maxillary osteosarcomas behave differently than osteosarcomas of long bones. The sites of these tumors also impose a different set of problem for resection and reconstruction than long bone tumors.

The aim of this study to present the National Cancer Institute (NCI), Cairo University experience in treating patients diagnosed with osteosarcoma of the jaw. A summary of anatomical sites, diagnostic challenges, surgical procedures and treatment outcome will be discussed.

Patients and methods

This is a retrospective case series study involving all cases who presented to the NCI during the period between 2006 and 2013 with primary jaw osteosarcomas. Demographic data, as well as data regarding clinical presentations, treatment modalities and their outcomes were all collected from hospital records of the NCI, Cairo University.

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Statistical method

The IBM® SPSS® Statistics version 22 was used for statistical analysis. Mean, standard deviation and range were used to describe numerical data while qualitative data were described using frequency and percentage. Survival analysis was done using Kaplan-Meier method. Overall survival calculated from date of diagnosis till date of death or last follow up while disease free survival was calculated from date of surgery till date of recurrence whether local or distant or death or last follow up date.

Results

Twenty-one patients with osteosarcoma of jaw bones were diagnosed and treated at the NCI during the period studied. Demographic features and clinical presentations of the cases are illustrated in (Table 1).

Fifteen cases (71.4%) had lesions in the mandible and 6 cases (28.6%) in the maxilla.

Radiology

A local CT scan of the jaw was conducted on 21 cases, and an MRI on one case. Periosteal reactions in the form of “sun ray” appearances were detected in only in two cases (9.5%) (Figs. 1 and 2). Only two cases (9.5%) had suspicious lymph nodes

(detected by CT scanning). Lung deposits were reported in one case (4.7%). They were discovered by X-ray conducted at presentation.

Pathology

Different methods were used to obtain needed biopsies. Open biopsy was used in 14 cases (66.7%), and core needle biopsy in 7 cases (33.3%). Inaccurate biopsy results were initially delivered in 4 cases (19.04%) compared with final pathology reports of surgical resections. Three cases were diagnosed by biopsy as chondrosarcomas and one as case of osteoblastoma. Final pathology of these cases revealed them all to be osteosarcomas.

Different pathological variants of osteosarcoma were found (Table 2). Thirteen cases (61.9%) were resected with a negative margin whilst 8 cases (38.1%) were resected with positive margins. Cases that had positive margins were 4 maxillary cases (66.7% of maxillary cases) and 4 mandibular cases (26.6% of mandibular cases).

Surgical treatment

Surgical resection was done for all cases. Type of operation illustrated in (Table 3)

The relation between types of mandibular operations (Hemi-mandibulectomy vs. Segmental mandibulectomy) and marginal status (positive or negative) was statistically insignificant (p-value = 0.6).

Chemotherapy

Only twelve cases received chemotherapy; 2 cases (9.5%) received neo-adjuvant chemotherapy; 5 cases (23.8%) received adjuvant chemotherapy; 5 cases (23.8%) received both neo-adjuvant and adjuvant chemotherapies. Chemotherapy regimens were in the form of 4–7 cycles of cisplatin or carboplatin plus doxorubicin except for one case which received 6 cycles of methotrexate-based regimen.

Recurrence and outcome

The survival analysis included 17 cases since there were 4 cases lost follow up after doing surgery so they were excluded from

Table 1
Patients' demographic features and clinical presentation.

Age	Mean ± SD	Range
Gender	29.7 ± 11.9 years	15–54 years
	Males	Females
	10	11
Clinical presentation	Number of cases	
Mass	21	
Size of tumor	Mean ± SD	Range
	4.9 ± 2.7 cm	2–13 cm
Pain	6	
Ulceration in oral cavity	6	
Trismus	2	



Fig. 1. CT of Osteosarcoma of the right side of the mandible for 16 years old female show sun rays periosteal reaction.

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