CASE REPORT

Osteogenic Sarcoma of Lower Jaw

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Abstract

A rare case of osteogenic sarcoma affecting the lower jaw is presented. Osteogenic sarcoma of the jaw is a very rare tumor of the head and neck comprising less than 0.5% of all malignant tumors of head and neck. Pertinent literature is reviewed to emphasize the various treatment modalities and histopathological features for diagnosis of this rare tumour.

Key Words

Osteogenic sarcoma, lower jaw.

Introduction

Osteogenic sarcoma (OS) is an uncommon tumour accounting for 7,400 new cases and 4,200 new deaths in the United States annually. Only 10% occur in head & neck (1). Osteogenic sarcoma represent about 2.4% of all childhood cancers and 56% of all malignant bone tumors in children (2). As per literature, the osteogenic sarcoma of jaw comprises of less than 0.5% of all malignant tumors of head & neck (3). The involvement of facial bones is rare, with maxilla and mandible being the commonest bones affected along with occasional involvement of frontal sinus and zygomatic bones (4). The involvement of facial bones in descending order of frequency, is maxilla, mandible, skull and cervical spine (5). The incidence of OS of jaw is higher in males than females with ratio of 3:2 and is more common in the younger age group i.e. 20-30 years (4). Several epidemiological risk factors, related to development of OS include– a history of ionising radiation exposure; fibrous dysplasia; hereditary retinoblastoma; or prior exposure to thorium oxide (1). The incidence of OS substantially increases when the risk factors are combined.

We report a case of osteogenic sarcoma of the lower jaw in a 20 year old male patient who failed to avail proper medical/surgical treatment well in time leading to huge size of the tumor giving gross disfigurement.

Case report

A 20 year old male patient with painless swelling of the lower jaw including floor of mouth, more so on the left side reported in the ENT OPD. Five years back, he started with a swelling in the floor of the mouth on the left side, which gradually increased in size. The swelling was not associated with pain or any other symptomatology of any system. There was no history of trauma or exposure to radiations. Local examination revealed an irregular swelling about 20cm×15cm in size, more towards the left side. The swelling involved the both rami of mandible and floor of mouth. At this time it was not possible to pinpoint the site of origin. The swelling was non-tender, with well defined margins,

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having visible pulsations. It was immobile, fixed, non-compressive with raised local temperature and was firm to hard in consistency. The skin over the swelling was stretched and showing dilated visible veins over it. There was a small ulcerated area on the floor of the mouth towards left side. Loosening of teeth was also seen (Fig. 1). The patient was having moderate difficulty in swallowing and his voice was altered. The ear, nose and throat examination revealed no abnormality. Systemic examination also failed to reveal any abnormality. Routine investigations were all within normal limits. A biopsy was taken and the histopathological examination revealed an osteogenic sarcoma (Fig. 2).

Radicle resection of the tumor under general anaesthesia followed by chemotherapy was planned for this patient, but the patient refused surgery. For his personal beliefs, he agreed only for radiotherapy. He was given full course of palliative radiotherapy — 40 Gy/20 fractions with 12×12 field size directed to anterior lower mandible. But the patient did not show any response. Patient was motivated to undergo surgery, but he again refused and did not turn up for follow-up.

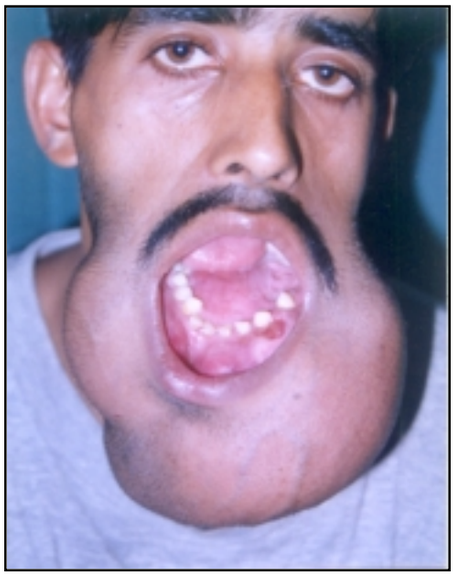

Fig. 1. Swelling arising from floor of mouth with loosening of teeth & ulcer on floor of mouth.

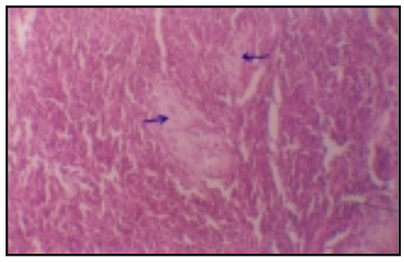

Fig. 2. Photomicrograph showing diffuse sheets of malignant cells directly laying down the osteoid-arrows (H&E 100X)

Discussion

The recent literature from head & neck surgery contains only infrequent case reports and small series of osteosarcomas. The prognostic factors and optimal management of OS of head and neck are, therefore, poorly defined. Etiology of osteogenic sarcoma is unknown. Radiation therapy (RT), paget's disease, fibrous dysplasia and a history of trauma have all been implicated in the pathogenesis of this tumor (4). OS of jaw bones differ from OS of other regions and it occurs a decade later. OS grows rapidly and invades locally but have a limited tendency to metastise. The most common presenting sign and symptoms of OS include– an enlarging mass with or without pain or paraesthesia; loosening of teeth; and firm, fixed swelling (6). The present case also has the similar features.

Radiology of OS shows areas of medullary destruction with osteosclerosis or osteolysis. "Sunray" spiculations radiating from the cortex into adjacent soft tissues may be a feature. Over 80% of the patients have micro-metastatic disease at the time of diagnosis, most commonly in lungs (1). Distant metastasis occur within 1 year in 80% of patients of OS of long bones, but the rate of metastasis is high in case of jaws bones (3). Histologically, OS consist of a malignant undifferentiated stroma and neoplastic osteoid formation along with increased mitotic rate of proliferating stromal cells. The stromal component is characterised by dense cellularity and pleomorphism

(1). Similar findings have been observed in our presentation. P^{53} mutations, increase in serum alkaline phosphatase and immunohistochemical overexpression of P^{185} have also been reported, with overexpression of P^{185} correlated with poor outcome (4,7,8). Overexpression of P^{185} (the product of HER-2/neu oncogene, a– 185^{ICD} protein) is seen in approximate 40% of primary OSs and even higher proportion (77%) in metastatic tumors (2). The prognostic variables in addition to stage of presentation of OS, which have been seen to risk stratify OS patients include clinicopathological parameters (e.g., p-glyco-protein, expression and abrogation of P^{53}, cyclin-D P^{16}) (8).

The optimal management of patients with OS of head and neck is unclear. Different authors have given different treatment modalities e.g., radical surgery, combined therapy employing surgery, R.T. and/or chemotherapy. Improved local control and survival have been seen with adjuvant RT with or without chemotherapy. Five year survival rate have been reported in 70-75% of patients receiving pre-operative radiation followed by wide surgical excision and post-operative chemotherapy (1). The OS of head & neck have less risk of distant metastases but a higher rate of local recurrence which may be due to difficulty in achieving wide surgical margins in head & neck surgeries due to anatomic and cosmetic reasons. Radicle surgery alone results in local failure (1, 3). Chemotherapy definitely reduces the risk of metastatic recurrences (9). The modern management approach involves- initial biopsy, then combination chemotherapy followed by surgical excision aiming to achieve clear surgical margins. Hemimandibulectomy or radicle maxillectomy is undertaken in preference to radiotherapy because OS is a relatively radioresistent tumor (10). OS of jaws is more curable than those that occur in long bones. The five-year survival rate for OS of all site is 65-80% whereas that for OS of jaws is 30% (3, 9). The death is usually due to direct extension of the tumour into the brain tissues (1). Preoperative radiotherapy prolongs the survival as has been documented in OS of long bones (3). In our patient the pre-operative radiotherapy was given with the aim that pre-operative radiotherapy enhances the local control with surgery and chemotherapy by preventing or suppressing distant metastases. After undergoing combined treatment with surgery and chemotherapy, 50-60% of patients who present with a localized OS are cured. But the prognosis in patients with metastases at the time of diagnosis is significantly worse, with few long-term survivors (7). Furthermore, the skull lesions have worse prognosis compared to others (1).

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