

A CASE REPORT OF CONVENTIONAL CHONDROSARCOMA OF MANDIBLE**VAIDEHI R PATEL^{a1}, SWAPAN GOSWAMI^b, RAKESH TANDON^c, VISHAL BHUVA^d,
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A 19 year old female patient presented with chondrosarcoma in left mandibular region near angle of mouth. The commonest sub type of chondrosarcoma is Conventional Chondrosarcoma which may be low, intermediate or high grade. They are frequently seen in the 4th and 5th decades with a slight male predominance 1.5-2.0:1. They can also be classified on their location within bone into Central, Peripheral, and Juxtacortical Chondrosarcomas. Rare subtypes of chondrosarcoma like Dedifferentiated, Mesenchymal and Clear Cell Chondrosarcoma are also seen. The extent of the tumour within the bone and extension into soft tissue can be delineated by MRI preoperatively. Being radioresistant, it is usually surgically treated by wide resection. Prognosis of jaw lesions is poor as compared to the lesions affecting the long bones of the body. Death occurs usually by direct extension to the base of the skull or due to distant metastasis to lungs and other bones.

KEYWORDS : Conventional Chondrosarcoma, Mandible, Prognosis

Chondrosarcoma is a malignant tumour wherein the tumour cells form cartilage (Brad et al.,2004). It usually involves the pelvic girdle, chest wall and scapula (Fred et al. ,1991). It does not involve the jaws commonly and if it does, it affects the maxilla rather than the mandible (Cohen and Smith,1963 ; Kenichi et al., 1995). In a series at Mayo clinic chondrosarcoma involved the jaw in only 3% of cases (Dahlin and Unni, 1986). The molar region of mandible is affected more compared to ramus, condyle, coronoid process or symphysis. (Fred et al., 1991 and Takayuki et al., 2006). Chondrosarcoma are slow growing tumour with a tendency for local recurrence after surgery. However with recurrence they exhibit rapid and aggressive growth. Chondrosarcomas of head and neck present without pain in contrast to excruciating painful presentation of chondrosarcoma of long bones. Some authors believe there is no sex predilections, however others have found a male to female ratio of 2:1.(Brad et al.,2004) These tumours occur in a range of 10 to 80 years, with most seen between 30 to 50 years of life(Fred et al. 1991).

CASE REPORT

A female patient aged 19 years presented with chief complaints of a painful swelling in left mandibular region which was of one and a half month duration. In last

20 days, swelling increased in size. The patient had swelling on left mandibular region measuring 4x3x1.5cms with ill defined margins.

The swelling was firm to hard in consistency and skin over the swelling was intact with normal colour. Intra oral examination revealed a swelling on lingual side of mandible which was 4x3x1.5cms. The swelling was tender, nonmobile, attached to bone and the mucosa over the swelling was intact and non hyperemic.

Radiograph showed a hypodense lesion in left mandible measuring 4.7x1.2cms in axial plane, 4.9x1.5cms in coronal plane, 4.9x1.2cms in sagittal plane. Lesion was on left side, close to the angle of mouth. There was extension of lesion which was more intraorally reaching up to midline and also involving left temporo-mandibular joint. Radiographic features were suggestive of bone tumor. Radiographic differential diagnosis included- Ewing sarcoma, osteosarcoma, fibrous dysplasia, central fibroma with calcification and osteoblastic metastatic carcinoma.

Surgically excised tissue consisted of a single tumor mass and in surgical diagnosis ossifying fibroma and chondroma were considered. The tumor mass was composed of cartilaginous tissue which measured 2.5x2.5x2cms having pearly white colour with irregular surface. On cutting, it had a consistency of cartilage tissue.

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Figure 1 : Gross Photograph Of Tumor Mass Showing Pearly White Cartilaginous Tissue With Irregular Surface

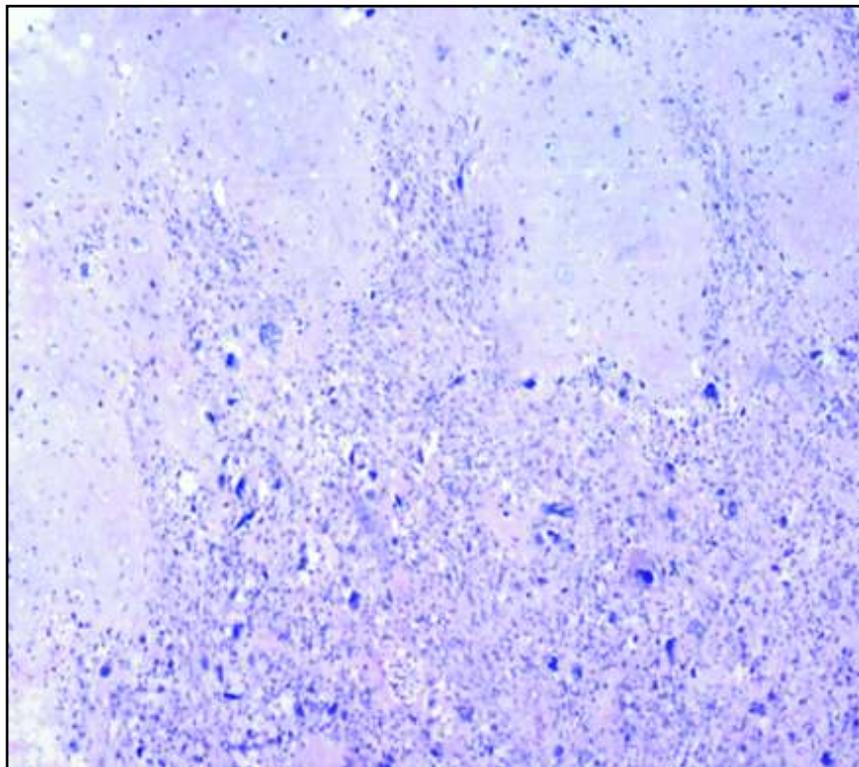


Figure 2 :A Low-Power Field Illustrating The Division of The Tumour By Fibrous Septa (haematoxylin And Eosin 40 X)

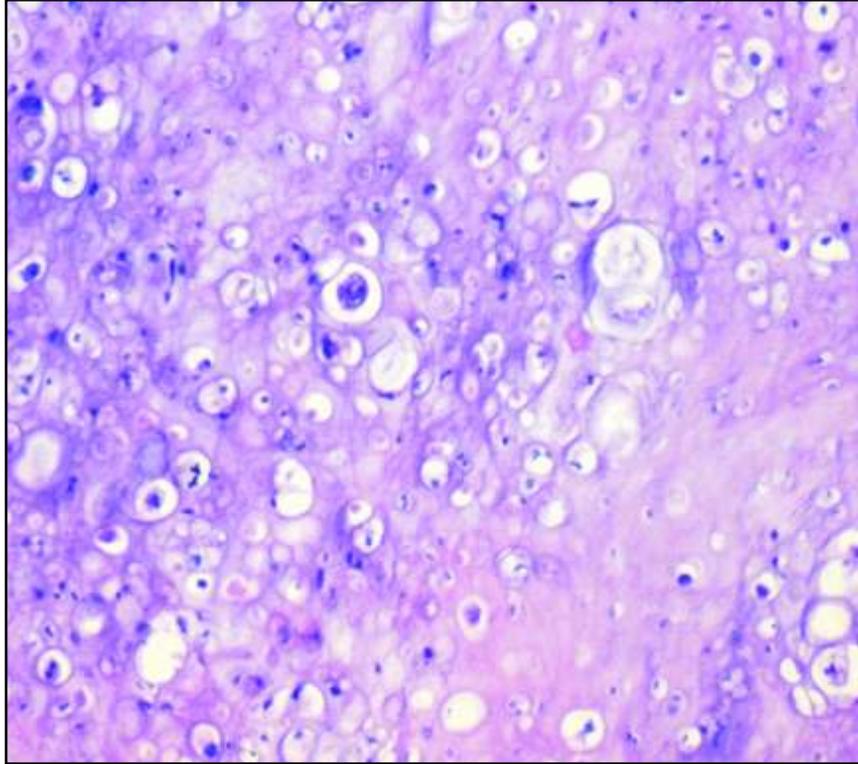


Figure 3 : Higher Magnification Showing Lacunae Containing Two Or More Cells And Binucleated Chondrocytes Seen Forming Malignant Cartilage. (Haematoxylin And Eosin 100 X)

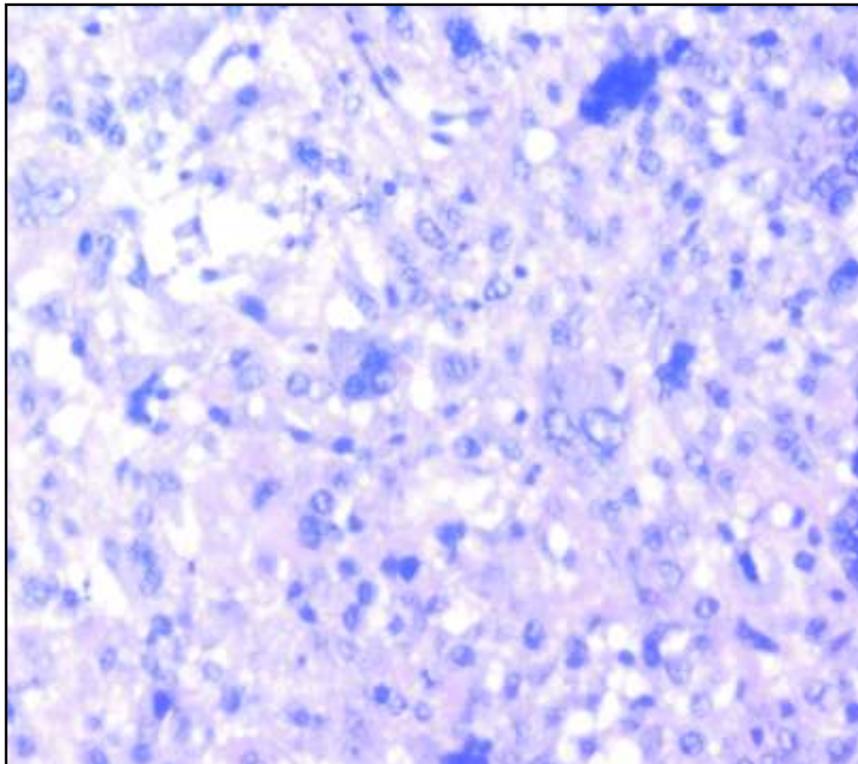


Figure 4 : Tumour Cells Showing Hyperchromatism And Mitotic Activity.(haematoxylin And Eosin 200x)

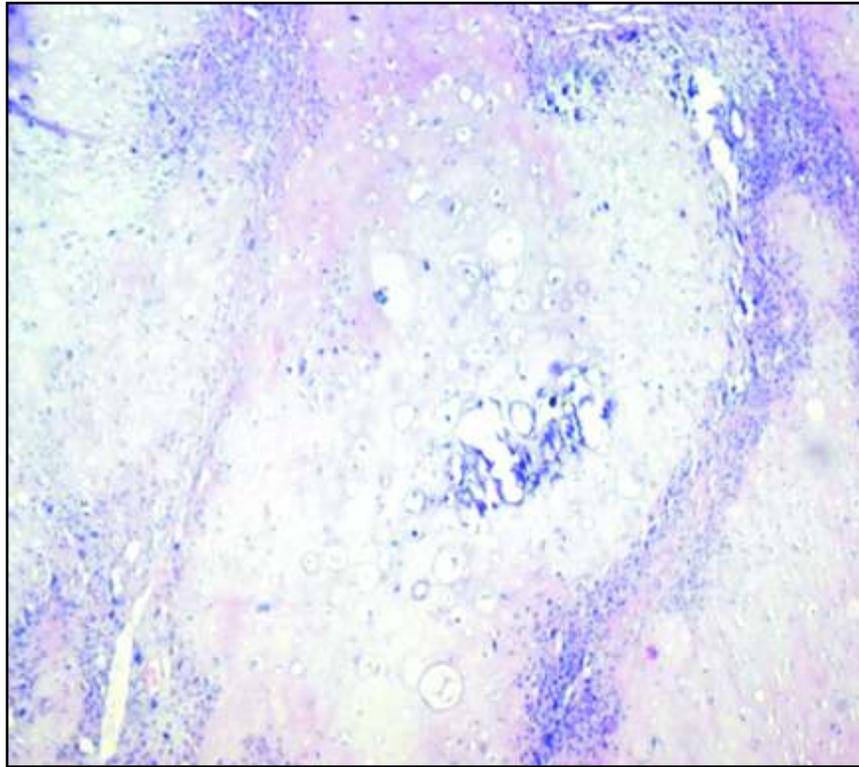


Figure 5 : A photomicrograph of one of the calcified spicules. These are fragments of calcified chondroid (Haematoxylin and Eosin 100 x)

Multiple sections were processed and submitted for histopathologic examination.

Histological examination showed (figure1) presence of lobules of cartilage surrounded by highly cellular, pleomorphic fibrocartilagenous tissue. The stromal cells of the fibrocartilagenous tissue were not small and round but showed various degree of pleomorphism, giant anaplastic cells and increased atypical mitosis. The Nuclei were enlarged, having open chromatin, irregular contour and prominent nucleoli. Lacunae containing two or more cells and binucleated chondrocytes were seen.

Focal areas of calcification were seen. Areas of haemorrhage and necrosis were seen. No evidence of tumor osteoid was seen.

Few thin walled blood vessels were seen, however haemangiopericytoma like pattern was not apparent, as is seen in Mesenchymal Chondrosarcoma. Overall features were that of Conventional Chondrosarcoma grade III.

DISCUSSION

It was in 1930 that Phemister stated that sarcomas, involving bone, which contained cartilage were chondrosarcomas, since then chondrosarcoma was recognized as a distinct entity (Fred et al., 1991). This concept was supported by Ewing who stated that chondrosarcomas and osteogenic sarcomas were two separate lesions (Fred et al. 1991). In 1942, Lichtenstein and Jaffe defined chondrosarcoma as a lesion which developed directly from a sarcomatous stroma, developing from a full-fledged cartilage and not showing neoplastic osteoid tissue and bone.

Ewans et al.,1977 classified Chondrosarcomas into three grades I, II and III based on cellularity, nuclear size and mitotic rate. Various types were identified viz. Conventional Chondrosarcomas, Clear Cell Chondrosarcomas, Myxoid Chondrosarcomas, Dedifferentiated Chondrosarcomas and Mesenchymal Chondrosarcomas.

Amongst cartilaginous tumours, the malignant tumours are quite common compared to benign ones (Rajendran and Sivapathasundaram, 2006). Diagnosing a well differentiated chondrosarcoma from a chondroma is fraught with difficulties (Fred et al., 1991). Due to indistinct boundaries between benign and malignant chondromatous tumours, there is difficulty in diagnosis of chondrosarcoma of the jaws and it might ultimately rest on the behavior and clinical course of the tumour.

Tumors like chondroma and chondroblastic osteosarcoma are difficult to distinguish from chondrosarcoma. Chondroma is very rare in facial bones and jaws, affecting usually small bones. Thus any cartilagenous tumours in an adult with increase in size or causing pain should be viewed with suspicion of malignancy (Takayuki et al., 2006). Also microscopic examination of the biopsy will reveal subtle differences like hyperchromatism and abnormally enlarged nuclei favouring malignancy. (Lichtenstein and Jaffe, 1942). pointed out that appearance of plump nuclei, more than an occasional cell with two or more nuclei and giant cartilage cells with large solitary or multiple nuclei or showing chromatin clumps are features supporting malignancy in cartilaginous tumors.

Cartilage forming neoplasm of the jaws are twice likely to be malignant than benign, so some recommend all cartilaginous tumours of craniofacial region should be excised widely even if histologically benign. If tissue is insufficient for study an improper diagnosis is made, hence multiple biopsies from different sites are required to confirm malignancy (Fred et al., 1991).

Fu and Perzin in 1974, described three prognostic factors: location and extent of the lesion, adequacy of surgical therapy, and degree of differentiation of the tumor. It was found that 5 year survival rate was 40 -60 %, and few cases showed recurrence even 10 to 20 years later. Hence a life long follow up after surgery is recommended (Fred et al., 1991).

Our case too, later on examination of excision biopsy confirmed the suspicion of malignancy and was diagnosed as conventional chondrosarcoma (Well differentiated type).

The patient was referred to a higher oncology centre and was advised wide excision of the affected area. Preoperative chemotherapy was initiated. However patient developed secondary complications and expired before the surgical procedures could be carried out.

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