

Intensity Modulated Radiotherapy [IMRT] in a Rare Case of Infratemporal Chondrosarcoma with Intracranial Extension in a Young Adolescent Patient

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Authors' contributions

This work was carried out in collaboration among all authors. Authors TL and MA has scripted the draft, conceptualization and data curation. Author SK has contributed with the microphotographs and the corresponding legends, review and editing. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Mesenchymal chondrosarcoma is a malignant neoplasm of cartilaginous origin. It is an uncommon subtype of chondrosarcoma which is described in the young adolescent population. We present a rare case of chondrosarcoma in a teenage group-a 16yr old pediatric patient. This 16 year old patient came to us with progressive, painless, right facial asymmetry of 3 months duration .MRI scan showed a large lobulated enhancing lesion in right infratemporal fossa and masticator space with destruction of right mandibular bone with intracranial extension. She underwent subtotal excision [R2 resection] followed with adjuvant radiotherapy by Intensity modulated radiotherapy technique (IMRT) and remains disease free for 1.5 years on follow up. Role of chemotherapy is not defined. Combined modality approach including surgery and radiotherapy maximizes the chances of cure despite the aggressive biology of mesenchymal chondrosarcomas.

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1. INTRODUCTION

Mesenchymal chondrosarcoma represents 1% of all chondrosarcomas with the majority of them occurring in the head and neck region. These rare tumors have peak incidence between 20-30 years of age. This variant is locally aggressive with a high potential to cause distant metastasis. Surgery remains the cornerstone for curative treatment, however in inoperable lesions, local adjuvant radiotherapy remains a potential curative tool. Role of chemotherapy, immunotherapy and other biological therapies are yet to be defined. In this case, we present a young girl who underwent a R2 resection [macroscopic residual tumor] and was offered photon based radiotherapy by intensity modulated radiotherapy [IMRT] which resulted in complete response on her first follow up post 8 weeks with tolerable toxicities

2. CASE REPORT

A sixteen year old adolescent patient, student by occupation presented with a history of right cheek swelling which was painless in nature and around the size of a coin, the patient neglected the swelling for 3 months and finally visited a dentist, who had examined her (Fig. 1). He did a biopsy and diagnosed it as chondrosarcoma, she underwent MRI imaging for characterization of the tumor. The patient was then referred to a cancer hospital. Slides/ block review was done at our centre. Section showed lesion with fragments of bone. Lesion showed a biphasic pattern, composed of lobules of hyaline cartilage and sheets of small round cells with hyper chromatic nuclei and scant cytoplasm. Mitosis was

discerned. Hemangiopericytomatous vascular pattern was seen. Areas of necrosis were seen suggestive of Malignant small round cell tumor with cartilaginous component. The morphology favored Mesenchymal chondrosarcoma. The ki67 labeling index was 25%.

2.1 Treatment Course

She underwent right temporal craniotomy and subtotal excision of the tumor of the infratemporal fossa on 14/11/18. Postoperative histopathological Examination revealed a mesenchymal chondrosarcoma around 8.5 cm size with infiltrating cellular neoplastic lesion composed of sheets of round cells against a myxoid background (Fig. 7A, B, C, D).

After 3 weeks post operatively, she underwent a MRI scan of head and neck region for assessment of residual lesion. It revealed a large lobulated irregular mass measuring 4.4 x 6.5 x 4.7 cm of heterogeneous signal morphology in right infratemporal fossa + masticator space, peripherally hypointense + centrally hyperintense on T1W, intermediate to hyperintense on T2W imaging with internal hypointense septae. There was destruction of the right mandibular bone, infiltration of masticator muscles, indenting over the posterolateral wall of maxillary antrum and widening of skull base foramen with small mid intracranial and extra-axial extension. No evidence of parenchymal invasion. Residual Chondrosarcoma with central haemorrhagic collection. Fluid intensity within mastoid air cells - Mastoiditis. Mildly enlarged right I b nodes, 8 mm in short axis – Reactive (Fig. 5A, B).



Fig. 1. Extra oral and intra oral view of the swelling over the right cheek

CECT chest was done and nothing abnormal was detected. A multi-disciplinary consult was taken and she was advised adjuvant radiotherapy. On local examination, she had a scar over the right temporal region of the scalp adjacent to pre auricular region and neck. No neck nodes were palpable. Right cranial nerve 5th and 7th palsy was demonstrated [postoperative complication]. She underwent 66Gy of 6MV photon therapy in 33 fractions, 2Gy per fraction on a linear accelerator using intensity modulated radiotherapy [IMRT] with sequential field reduction in 7 weeks from January to march 2019. Acute toxicities included RTOG grade 2 dermatitis with hyper pigmentation and patchy epilation of scalp hair in the irradiated zone (Fig. 2). On follow up after 6 weeks, MRI was done for response assessment. MRI revealed ill defined mixed intensity area on T1W + T2W imaging in right temporal, infra temporal + masticator space with central fat intensity with no evidence of significant restriction on Diffusion Weighted Images (Fig. 6A, B). Right mastoid air cells are replaced with fluid intensity - Mastoiditis. No evidence of intracranial lesion. It was suggestive of a complete response. The patient is on regular follow up and her clinical examination is normal

except for partial recovery of right facial nerve palsy. Right trigeminal nerve has attained full recovery. Regrowth of scalp hair along with disappearance of skin changes over face (Fig. 4).

3. DISCUSSION

Chondrosarcomal tumors have many variants including conventional, mesenchymal, clear cell and dedifferentiated subtypes. In 1954, Mesenchymal Chondrosarcoma (MCS) which is an uncommon histological variant of chondrosarcoma was first described by Lichtenstein and Bernstein as an occurrence in the bone [1]. In the head and neck region, there may be soft tissue or skeletal involvement. Among the bony involvement, mandible and maxillary region are more frequent and infratemporal fossa region may be rarely affected [2,3]. It accounts for 1% of all chondrosarcoma with around 800 cases of Mesenchymal chondrosarcoma reported [4]. This tumor affects children and young adults, usually between the ages of 15-35 years, like in our case our patient is 16 years old. Females are affected slightly more often than males.



Fig. 2. Postoperative images-before the commencement of adjuvant radiotherapy



Fig. 3. Radiation induced acute toxicities-RTOG grade 2 dermatitis with hyper pigmentation and epilation of scalp hair over involved field



Fig. 4. On follow-up -post 9 months of radiotherapy-no major cosmetic deficits

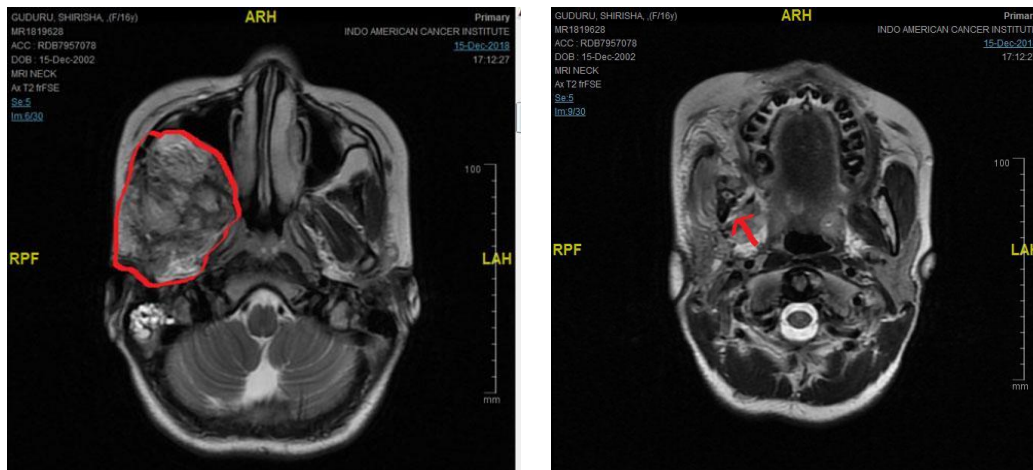


Fig. 5(A, B). Post operative MRI neck with contrast showing residual tumor with destruction of mandibular bone [shown in arrow]

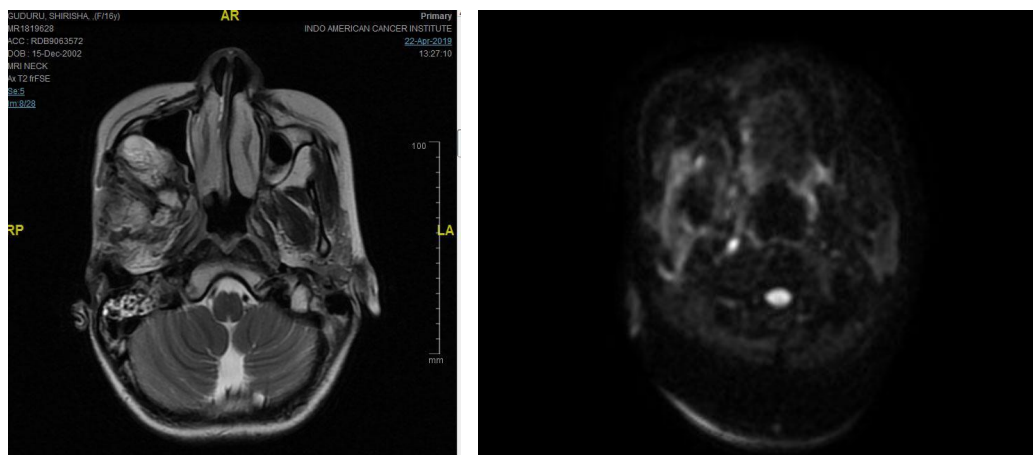


Fig. 6(A, B). T2 weighted and diffusion weighted images of MRI done 1 month after adjuvant radiotherapy

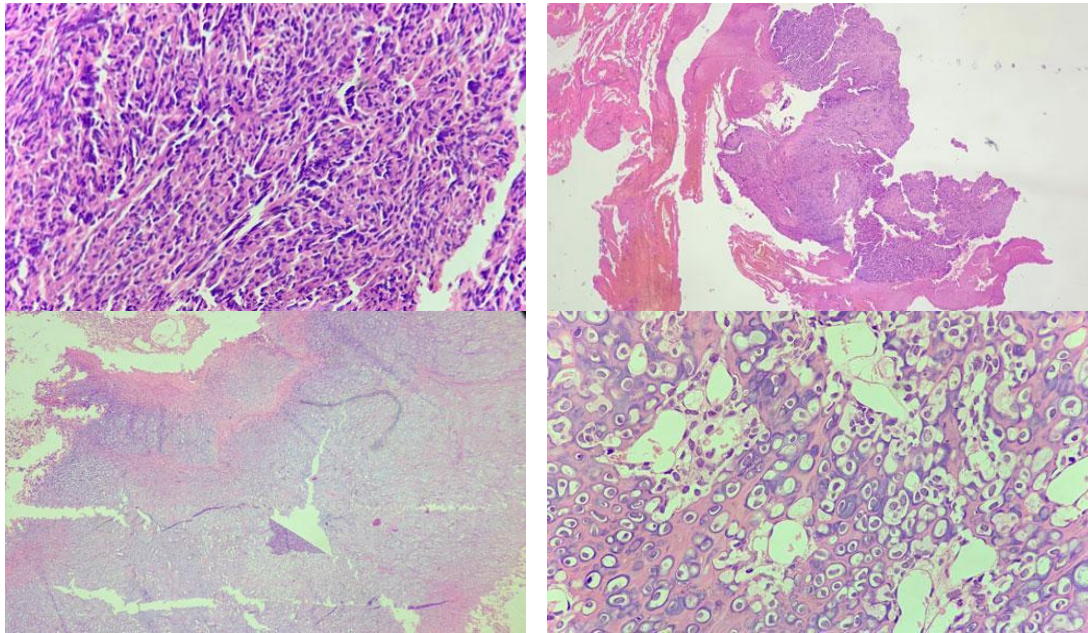


Fig. 7. (A). Bundles of plump spindle cells having hyperchromatic nuclei with intervening hyalinised stroma 100x (B). Infiltrating cellular neoplastic lesion composed of sheets of round cells against a myxoid background H&E 40x (C). Clusters of undifferentiated small blue cells mixed with islands of hyaline cartilage H&E 100x (D). Lobules of mature hyaline cartilage showing mild pleomorphism and large dark nucleus H&E 400x

Mesenchymal chondrosarcoma is assumed to originate from remnants of the embryonic cartilage or metaplasia of meningeal fibroblasts. The characteristic DNA change in mesenchymal chondrosarcoma is the deletion of a part of chromosome 8 that ends up linking to DNA genes together in a new way. The genes are termed HEY1 and NCOA2. These abnormal genetic changes appear to occur spontaneously for unknown reasons. This tumor is unique because of its aggressive growth with a high tendency for late recurrence and delayed metastasis [2]. It is regarded as a high-grade sarcoma in the grading systems of the French Federation of Cancer Centers Sarcoma Group, the National Cancer Institute [5] Muller et al. [2], have found that it expresses type II collagen which differentiates it from other small cell sarcomas such as Ewing's, synovial sarcoma and haemangiopericytoma [6].

Mesenchymal chondrosarcoma (MCS) differs from typical chondrosarcoma in the following respects. First, MCS tends to be more aggressive, with 5- and 10-year survival rates of 54.6% and 27.3%, respectively [1]. Second, classical chondrosarcomas show a predilection for middle-aged to elderly males whereas MCS has a slight female preponderance and occurs in

the nervous system in patients at age 20 to 30 and in soft tissues in patients at age 40 or older. Although they arise in the bones mostly, around 22% - 50% of them occur in the soft tissues as well especially the brain and its meninges. MCS has a high propensity to metastasize to the lungs, lymph nodes, and other bones [1]. According to Schneiderman et al, cranial tumors behave clinically different suggesting better overall survival in young patients (compared with axial and appendicular locations) and a worse survival outcome in older patients [7].

Wide local excision to achieve R0 resection is commonly regarded as the "gold standard". But, treatment without surgery is associated with poorer overall survival and event-free survival. Negative surgical margins could significantly bring down the local-recurrence rate and are associated with a higher event-free survival rate. The addition of radiation therapy is not significantly associated with the overall or event-free survival. However, local radiation is recommended as the salvage therapy for patients with positive margin so as to achieve better local control. Postoperative radiation therapy is thus usually administered in a majority (60%–70%) of patients as it has been shown to

significantly decrease the local recurrence rate by a factor of almost 4 [8]. The toxicities associated with radiotherapy mainly depend on the volume, technique of radiotherapy, dose of radiotherapy and proximity to normal tissues. With the use of modern radiotherapy techniques like IMRT, acute and late toxicities may be minimized especially in the proximity of critical normal tissue like in our case allowing us to achieve tumoricidal dose with local radiotherapy boost. Although local control can often be achieved, distant disease (lung metastases) develops in 90% of patients and the 5-year survival rates drops to 0% - 18%.

The role of adjuvant chemotherapy is controversial. Various chemotherapeutic agents are used in MCS, including dactinomycin, carboplatin, cisplatin, cyclophosphamide, doxorubicin, etoposide, ifosfamide, high-dose methotrexate, and vincristine. Patients with MCS can be divided by the histologic categories into two groups to select a treatment protocol [7]. Those exhibiting a small cell pattern received a chemotherapy regime consulting Ewing family of tumors (focusing on adriamycin), while the hemangiopericytomatoid pattern received a reported regime in treatment of osteosarcoma. But in spite of this, adjuvant chemotherapy regime does not benefit the overall survival.

4. CONCLUSION

It is recommended that for tumors not amenable to ablative surgery due to its proximity with critical structures in the head and neck region, radiation therapy especially with conformal techniques like IMRT is an effective curative tool to bring down the recurrence rate. For patient with more malignant MCS in pathological behavior, the addition of adjuvant chemotherapy may play a more important role.

CONSENT AND ETHICAL APPROVAL

As per international standard or university standard guideline participant consent and ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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