



RADIOTHERAPY IN ANEURYSMAL BONE CYST OF THE OCCIPITAL BONE : A CASE REPORT

Radiotherapy

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ABSTRACT

Introduction: Aneurysmal bone cysts (ABCs) is a rare benign tumour of bone with an annual incidence of 0.14 per 100000 population mostly effecting the appendicular skeleton: femur tibia, fibula, humerus followed by skull bones and posterior elements of the spine. The pathogenesis of ABC still remains a controversial issue **Methods And Materials** Here we are going to describe a case of Aneurysmal Bone Cyst involving the occipital bone in a 8 year old boy. The boy had initially presented with headache and vomiting for which he was evaluated at Neurosurgery Dept after which he had undergone subtotal resection. The HPE showed it to be aneurysmal bone cyst. After discussion in the tumour board he was taken up for adjuvant radiation. He received a total dose of 30.6 Gy in 17 fractions through IMRT technique using daily image guidance. **Discussion** The optimal prescription of radiation dose is essential for avoiding the development of adverse reactions to radiotherapy, the recommended dose is between 26 and 30 Gy using standard fraction size for ABCs. Dose constraints are to be kept in mind for critical OARs while planning an IMRT plan for such location of the disease. **Conclusion** Aneurysmal Bone Cyst is an aggressive benign lesion with high rates of recurrence. Mainstay of treatment is surgery with radiation reserved for adjuvant and unresectable tumours. However the exact treatment of choice depends on the treating physician

KEYWORDS

BACKGROUND

Aneurysmal bone cyst (ABC) is a rare vascular and benign tumor-like lesion of bone. They account for 1-2% of all primary bone tumors and cranial location is described in 3-6% of all cases, the occipital location is very rare.

The annual incidence is 0.14 per 100000 population, mostly effecting the appendicular skeleton: Femur, Tibia, Fibula, Humerus followed by skull bones and posterior elements of the Spine. Lichtenstein has proposed that ABC is a reactive lesion rather than a true neoplasm and that vascular disturbances in bone leading increased intraosseous pressure, causing local destruction and distension of the bone.[2]

Usually the skull bones are rarely affected. Here we describe a case of a Aneurysmal bone cyst involving the Occipital bone in a 8 year old boy which is rare and the first case to receive adjuvant radiation at our institute.

Case Report

An 8year old boy presented with vomiting, headache for 4 months with no history of seizure and loss of consciousness. He was initially evaluated at Deptt of Neurosurgery, CEMRI was done which revealed a large well-defined lesion in occipital bone in midline with erosion of the same. It measured 6.20 cm x 4.30 cm x 5.00 cm in CS and CC dimensions. The lesion was anteriorly abutting and compressing the cerebellum with mild ascending trans-tentorial herniation crowding at foramen magnum and resultant tonsillar herniation 1.2 cm below McRae's line with upstream dilatation of third ventricle and bilateral lateral ventricles.

After thorough work up the patient had undergone, Suboccipital Craniotomy with Subtotal resection of the cyst at Neurosurgery Dept, CN Center, GMCH.

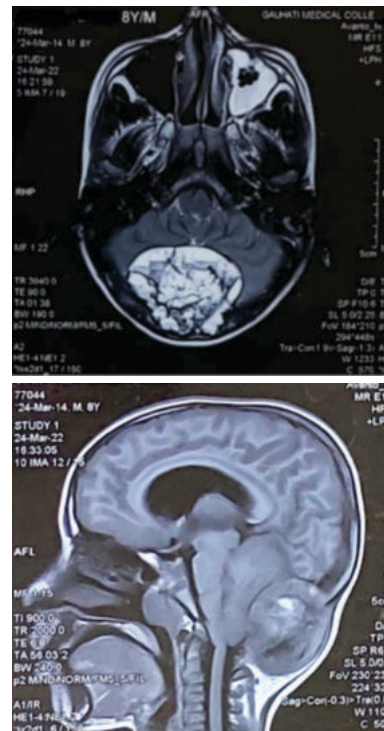
He was then assessed by Radiation Oncology team for adjuvant treatment. Post op his ECOG

Performance status was 1, his higher mental function as well as his cranial nerve function were intact. On motor system examination,

power in all four limbs were 4/5. The HPE slide was reviewed at our institute which showed Aneurysmal bone cyst.

The case was taken up for discussion in Multi-Disciplinary Tumour Board and the decided for adjuvant radiation.

Fig 1: Pre op CEMRI Brain: showing the lesion.(Axial and sagittal views)



Radiotherapy technique

CT simulation was done in SIEMENS® SOMATOM DefinitionAS-20 CT scanner and images were acquired with a slice thickness of 3 mm. The images were then transferred to the treatment planning system in DICOM format and with proper image registration and fusion, contouring was done for GTVr, PTV and OARs respectively. GTVr was contoured as the residual tumour evident on the planning CT images as well as observed on post op MRI scan and PTV was given with appropriate margin according to the institutional protocol. Important OARs like Brainstem, spinal cord, pituitary glands, optic chiasm etc were contoured. Planning was done in ECLIPSE® software using IMRT-IGRT technique and the treatment was delivered using 9 co-planar beams to a total dose of 30.6 Gy in 17 fractions with fraction size of 1.8 Gy per fraction in the Varian® Truebeam Linac .He successfully completed the prescribed radiation treatment.

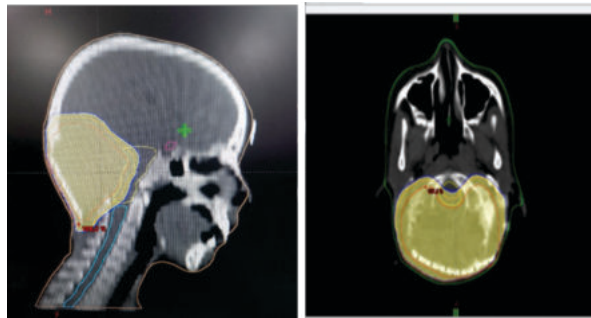


Fig 2 A
Fig 2 A: 98%(yellow) Isodose coverage of the Target volume (Sagittal)
2 B:98% (yellow) Isodose coverage of the Target volume (axial)

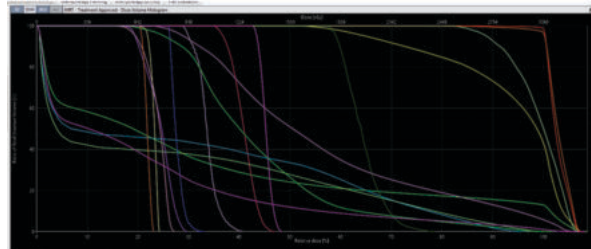


Fig 3 :Dose volume Histogram showing the RT doses to the OARs and GTVr. GTVr (Red),PTV (Orange),Brainstem (Yellow),Spinal cord (Sky Blue), Optic chiasm(Dark pink), left temporal lobe(light green), right temporal lobe (light pink) Pituitary (pink)

Table 1 : Dose constraints achieved for the PTV and critical OARs.

	PTV	BRAIN STEM	SPINAL CORD	OPTIC CHIASM	PITUITARY GLAND
Max dose(Gy)	33.26	32.8	29.31	14.44	14.81
Mean dose (Gy)	31.48	28.61	9.05	12.48	13.83
V107%[%]	0.6	0.0	0.0	0.0	0.0
V100%[%]	98.5	42.9	0.0	0.0	0.0

Follow up

After two months of treatment he came for follow up with an overall improvement in his performance status and general condition, after another 2 months on follow up with no specific complaints , MRI was done which revealed a heterogeneously enhancing cystic lesion in occipital bone with no mass effect or haemorrhage with no evidence of restricted diffusion. The patient was kept on follow up with consultation with Neurosurgical team.

DISCUSSION:

Aneurysmal bone cyst(ABC) is considered as one of the differential diagnosis of primary benign cranial bone tumours. The patient with ABC most commonly presents in the first two decades of life. Recently, genetics have been implicated in the etiology of ABC, leading some to believe that ABC is a true neoplasm rather than a reactive process. In a histopathologic study, Oliveira et al reported that gene rearrangements localized to t(16;17) in 36 of 52 primary ABCs (69%) in which the ubiquitin-specific protease 6 oncogene was placed under the regulatory influence of the highly active cadherin-11 promoter.[1] The patient with primary ABC typically reports pain and

may present with soft-tissue swelling or a palpable expansile mass. Leithner et al reported the expression of Insulin like growth factor [IGF 1] in most of the specimens.[4] Enneking divided the lesions into three stages: inactive, active, and aggressive. The inactive tumour is the most benign because the lesion is contained. An active lesion typically produces mild symptoms such as pain, with expansion and cortical thinning as well as a layer of reactive bone separating the lesion from normal bone . Aggressive tumours are rapidly expansile and destroy surrounding tissues[3].Imaging modalities used for diagnosis are X-Ray, CT scan , MRI.[5] The treatment of choice is surgical excision. There is a role of Adjuvant Radiotherapy but may be reserved for high risk patients based on grade, stage and for recurrence. Feigenberg et al , in their study of nine patients who received adjuvant radiation showed that no patient experienced a local recurrence (median follow up 17 years) [6].

When megavoltage radiation is used skin dose is reduced and distribution is more homogenous due to less side scatter.[7] At higher doses of radiation there is more likelihood of radiation induced second malignancy, therefore it is advised to use megavoltage radiation with less dose to minimise the occurrence of second malignancies.[8,9]. Some researchers advocate that all ABCs are component of a secondary process , either benign or malignant.[10, 11, 12]

Zhu et al reported no recurrences and no complication at final follow up following radiotherapy[13]. Although the adverse effects following radiotherapy must be considered upon, but with the advent of modern radiotherapy delivery techniques, the treatment have been made much safer than earlier times. The optimal prescription of radiation dose is essential for avoiding the development of adverse reactions to RT. Feigenberg et al recommended a dose between 26 and 30 Gy using standard fraction size for ABCs . While planning IMRT for these cases with a lots of surrounding critical structures ,such as the brainstem, pituitary, spinal cord, optic chiasm etc. the constraints of the various OARs must be kept in mind for a optimal plan.

CONCLUSION

Thus, Aneurysmal Bone Cyst is an aggressive benign lesion with high rates of recurrence. Mainstay of treatment is surgery with radiation reserved for adjuvant and unresectable tumours. However due to lack comparative trials in the use of adjuvant treatment the exact treatment of choice depends on the treating physician's experience and institutional preferences.

ABBREVIATIONS

- GTVr – Residual Gross tumour volume
- PTV – Planning target volume
- OAR- Organs at risk
- ECOG- Eastern Cooperative oncology group
- CEMRI – Contrast enhanced Magnetic resonance Imaging
- RT- Radiotherapy
- CC- Craniocaudal
- CS -Cross sectional
- IMRT- Intensity modulated Radiotherapy
- HPE- Histopathological examination

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