



RADIOTHERAPY IN ADULT PINEOBLASTOMA – A CASE REPORT

Radiotherapy

Dr. Diplu Choudhury	Assistant Professor, Dept. of Radiation Oncology, State Cancer Institute, Gauhati Medical College, Guwahati, Assam.
Dr. Kaustav Kumar Das*	Senior Resident, Dept. of Radiation Oncology, State Cancer Institute, Gauhati Medical College, Guwahati, Assam. *Corresponding Author
Dr. Tarjina Begum	Assistant Professor, Dept. of Medical Oncology, State Cancer Institute, Gauhati Medical College, Guwahati, Assam.
Dr. Nisarga A	Junior Resident, Dept. of Radiation Oncology, State Cancer Institute, Gauhati Medical College, Guwahati, Assam.
Mr. Kalyanjit Dutta Baruah	Junior Physicist, Dept. of Radiation Oncology, State Cancer Institute, Gauhati Medical College, Guwahati, Assam.

ABSTRACT

Pineoblastoma, a rare and aggressive embryonal tumour of the pineal region, predominantly affects children and young adults. It accounts for less than 1% of all central nervous system (CNS) tumours. These tumours, classified by the World Health Organization (WHO) as Grade IV pineal parenchymal tumours, are known for their rapid growth and tendency to spread via cerebrospinal fluid (CSF). Although histologically similar to medulloblastoma, Pineoblastoma has a more aggressive clinical behaviour. This report describes the successful management of an adult Pineoblastoma case. A 25-year-old male with symptoms of diplopia and headache was diagnosed with a pineal mass causing obstructive hydrocephalus. The patient had undergone endoscopic third ventriculostomy to relieve pressure symptoms followed by craniospinal irradiation (CSI) subsequently followed by chemotherapy. Radiotherapy was delivered using Helical Tomotherapy. Chemotherapy with Carboplatin and Etoposide was scheduled after radiotherapy, resulting in no residual tumour on follow-up imaging. The treatment strategy, including CSI and chemotherapy, aligns with current guidelines and offers insight into the management of Pineoblastoma in adults, where treatment protocols remain less standardized. The patient achieved significant recovery, with no signs of disease recurrence after two months. This case emphasizes the importance of personalized, treatment approaches and ongoing monitoring for this rare tumour.

KEYWORDS

INTRODUCTION:

Pineoblastoma is a highly malignant variety of embryonal tumours in the pineal region affecting mostly the children and young adults.[1] Pineal tumour constitutes less than 1 % of primary tumours of the central nervous system, The incidence is generally similar between males and females, though some studies suggest a slight male predominance.

In the United States between 2000 and 2017, the age-adjusted incidence rate (AAIR) of pineoblastoma was highest in children aged 0–4 years (0.049 per 100,000).[2] The World Health Organisation classifies broadly the tumours of the pineal gland into ; germ cell tumours(50-75%) and pineal parenchymal tumours (15-30%).

The pineal parenchymal tumours are further classified on the basis of the histological grade : Pineocytoma (WHO Grade I) , Pineal parenchymal tumour of intermediate differentiation (WHO grade II/III), Pineoblastoma (WHO Grade IV).[3]

Traditionally, pineal tumours fell into the broader category of CNS supratentorial primitive neuroectodermal tumours (SPNETs).

Although histologically they closely resemble medulloblastoma but the clinical behaviour of Pineoblastoma is more aggressive than that of medulloblastoma. [4-5]

Pineoblastoma are aggressive tumours with a propensity for cerebrospinal fluid (CSF) dissemination, Often warranting craniospinal irradiation (CSI). However due to their rarity in adults , the optimal radiotherapy approach remains undefined and standardized treatment strategies are lacking. This report presents a case of adult pineoblastoma managed successfully at our Institute.

CASE REPORT:

A 25 male old patient presented with diplopia and headache for one year without no history of loss of consciousness and seizure evaluated at SMCH and was referred to the Dept of Neurosurgery, CN centre, GMCH for further management. The CEMRI Brain done prior at

SMCH, Silchar revealed T1 isointense and T2 hetero-intense ill defined mass lesion of size 1.5cm x 1.7cm x 1.9 cm.(AP x CC x TR) showing diffuse restriction on DWI and heterogenous enhancement on post contrast noted arising from the pineal gland.

Lesion was extending into the tectum and causing mass effect leading to gross upstream dilatation of bilateral lateral ventricle and 3rd ventricle (obstructive hydrocephalus) On MRS choline peak was noted. All these feature were suggestive of pineoblastoma. Then the patient had undergone endoscopic 3rd ventriculostomy to get relief from the obstruction and pressure symptoms.

CSF cytology was normal with absence of any malignant cells. Biochemical markers like : LDH, *beta*-HCG , *alpha*-fetoprotein, which were in normal range.

Subsequently the patient was referred to the dept of Radiation Oncology for further course of treatment.

Post shunting clinical assessment was carried out by radiation oncologist and clinically the patient's ECOG Performance Score was 2 , with average built and nutrition. Higher mental functions were intact. Cranial nerve functions were intact, sensory and motor systems did not showed any abnormality .Power in all the four limbs were recorded to be 5/5.

A second CEMRI brain along with spine was performed at our institute which revealed, a 2.6x2 cm sized lobulated heterogenous enhancing FLAIR hyperintense Lesion seen in the region of pineal gland causing mass effect on the third ventricle with resultant mild supratentorial obstructive hydrocephalus – suggestive of Pineoblastoma. No significant abnormality identified in whole of spine.

Neurosurgery opinion was again taken for surgery or biopsy but the disease was not feasible for surgical resection.

The case was thoroughly discussed in MDTB of our institute, and was decided for CSI+ tumour boost followed by Chemotherapy.



Figure 1 : Pre Treatment CEMRI brain : T1 isointense lesion in the pineal region



Figure 2 : Pre Treatment CEMRI brain : T2 hetero intense ill-defined mass lesion in pineal region.

Radiotherapy Planning:

CT simulation was done in SIEMENS® SOMATOM Definition AS-20 CT scanner and images were acquired with a slice thickness of 3 mm. Immobilisation was achieved using Orfit® thermoplastic mask. The images were then transferred to the treatment planning system in DICOM format and with proper image registration and fusion, contouring was done for GTV, PTV and OARs respectively.

The contouring was based upon SIOPE – Brain tumour group consensus guideline [6].

For CSI :

CTV cranial, was delineated by outlining the **inner table of the skull** using the **bony window** including the **cribriform plate**, the **most inferior portions of the temporal lobes**, and the **entire pituitary fossa** then modified to encompass the **extension of cerebrospinal fluid (CSF) within the dural sheaths of the cranial nerves**.

CTV spinal included the entire subarachnoid space and encompassing the extensions along the nerve roots laterally. It is delineated caudally till the lower limit of the thecal sac (S2 vertebra)

Planning Target Volume (PTV)

The PTV margin was given according to the institutional protocol.

For Tumour Boost :

GTV was taken as the tumour evident on the planning CT scan fused with MRI scan **CTV** was given with a margin of 1.5 cm around the GTV and a **PTV** margin was added according to institutional protocol .

OARs;

The OARs delineated for the CTVcranial include the bilateral eye balls, lens, optic nerve , optic chiasm, brainstem, cochlea and parotid glands.

Adjacent to the CTVspinal, the OARs delineated include the larynx, oesophagus, thyroid, lungs, heart, liver, stomach, intestine, pancreas, kidneys and testis.

Planning was done in Accuray Precision® software using IMRT-IGRT technique and the treatment for CSI phase was delivered using 6MV photons to a dose of 36 Gy in 20 fractions with 1.8 Gy per fraction for five days a week in the Accuray Radixact Helical Tomotherapy and was followed by primary tumour boost of 18 Gy in 10 fractions to a total dose of 54Gy in 30 fractions to the tumour. Daily treatment was completed in 679.3 seconds. Weekly blood parameters like total counts and the other metabolic functions were monitored thoroughly throughout the treatment . He successfully completed the prescribed radiation treatment without any default or any break in the course.

Table 1 : Dose Constraints Achieved For The Important OARs. (both phases)

S.no	OAR	Dmax(Gy)	Dmean(Gy)
1	Brain	52.1	42.3
2	Spinal cord	36.12	35.35
3	Right optic nerve	40.37	37.88
4	Left optic nerve	40.30	37.48
5	Right eye	33.67	16.96
6	Left eye	33.91	15.35
7	Right lens	7.04	6.03
8	Left lens	6.72	5.76
9	Optic chiasm	44.32	40.11
10	Right lung	28.82	8.44
11	Left lung	29.94	7.70
12	Heart	12.68	3.70
13	Spleen	9.24	5.85
14	Right kidney	20.08	8.62
15	Left kidney	18..36	8.04
16	Liver	23.29	6.70

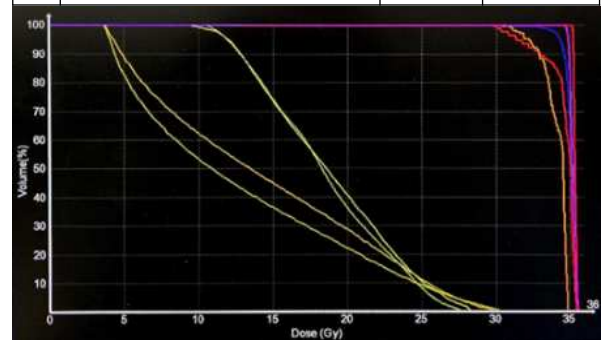


Fig 3A

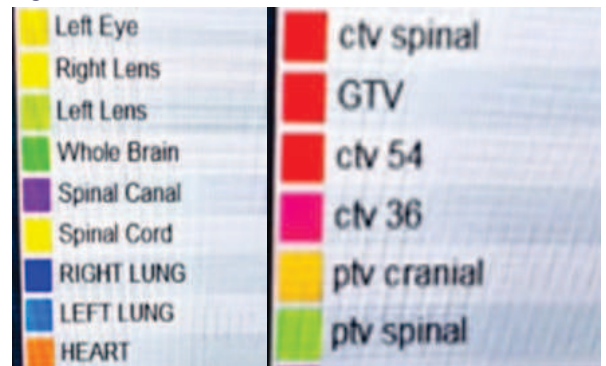


Fig 3B

Figure 3: A) Dose volume histogram for the target volumes and OARs B) Colour codes for different target volumes and OARs

Chemotherapy:

After completion of the Radiotherapy ,the patient received chemotherapy with three weekly cycles of Carboplatin and etoposide

to a total number of 6 cycles. He successfully completed the prescribed chemotherapy without any side effects.



Figure 4: Dose color wash of CSI in helical tomotherapy (36 Gy)

Follow up:

After two months of treatment the patient attended the OPD for follow up with an overall improvement in his performance status and general condition, after another 1 month on follow up with no specific complaints an CEMRI brain was done for the patient which revealed no obvious residual lesion in pineal region, but only treatment related sequelae in the form of micro-haemorrhages in the form of heterogeneous signal intensity.

He was kept on further follow up every three months for the initial first year.



Figure 5: CEMRI Brain: showing no residual disease after Radiotherapy

DISCUSSION:

The general consensus that adjuvant RT should be considered standard (category 2A in the NCCN guidelines) [7]. The prognostic significance of RT dose has been outlined with studies reporting better outcomes with higher doses. Schield and colleagues concluded that patients with PB have a high risk of spinal failure and should receive 30-45 Gy on the cranio-spinal axis and at least 50.4 -54 Gy to the primary tumour.[8]

A multicenter dosimetric study evaluated five different craniospinal irradiation (CSI) techniques—3DCRT, IMRT, VMAT, Tomotherapy, and Proton Pencil Beam Scanning—using a standardized benchmark patient [9]. The analysis showed that modern radiotherapy techniques provided superior dose distribution and organ-at-risk sparing compared to conventional 3DCRT. However, all IMRT-based approaches were associated with a higher integral dose, which raises

concerns about an increased risk of secondary malignancies. This potential long-term risk is a key reason why 3DCRT continues to be used in paediatric patients, despite its less favourable dosimetric profile.

The use of chemotherapy in adult patients of Pineoblastoma is not well-defined and is largely based on treatment approaches and clinical trials done for patients of medulloblastomas.

In the German trial HIT 2000, designed for children and young adults with non-metastatic SPNETs and Pineoblastomas, Gerber and his colleagues have used concurrent weekly vincristine and adjuvant vincristine-lomustine-cisplatin and have found out a five-year OS benefit of 64% in the latter (Pineoblastoma) and acceptable toxicity profile.[10] In a series of 17 PBs by Biswas et al, the most common chemotherapy regime was a combination of Carboplatin and Etoposide with acceptable toxicity profiles.[11]

Abbreviations:

GTV : Gross tumour volume

CTV : Clinical target volume

PTV ; Planning target volume

OAR : Organ at risk

CSI : Craniospinal irradiation

IGRT: Image guided radiotherapy

IMRT : Intensity modulated radiotherapy

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