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PINEAL PARENCHYMAL TUMOR OF INTERMEDIATE DIFFERENTIATION-A CASE REPORT

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Abstract

Pineal parenchymal tumours are uncommon, accounting for less than 0.3% of all primary CNS tumours. Pineal parenchymal tumours are diverse entities with a wide range of morphology. These tumours are histologically subdivided into pineocytoma (grade I), pineoblastoma (grade IV), papillary tumour of the pineal region (grade II or III), and pineal parenchymal tumour of intermediate differentiation (PPTID) (grade II or III) according to the WHO classification for tumours of the central nervous system (2021 revision). PPTID is an illness that has only recently been identified. Here, we report a case of pineal parenchyma tumor of intermediate differentiation**.** An adult female diagnosed with PPTID after surgery and treated with adjuvant EBRT alone to 54Gy in 30 fractions using shrinking field technique with CSI in initial phase. No other adjuvant treatment received.

Key Words: Pineal parenchymal tumors, WHO classification, CT brain, pineal parenchymal tumor of intermediate differentiation.

INTRODUCTION

Pineal parenchymal tumours are uncommon, accounting for less than 0.3% of all primary CNS tumours.1 Pineal parenchymal tumours are diverse entities with a wide range of morphology. These tumours are histologically subdivided into pineocytoma (grade I), pineoblastoma (grade IV), papillary tumour of the pineal region (grade II or III), and pineal parenchymal tumour of intermediate differentiation (PPTID) (grade II or III) according to the WHO classification for tumours of the central nervous system (2021 revision). PPTID is an illness that has only recently been identified.2 The WHO categorised it as a pineal parenchymal tumour with an intermediate prognosis between pineocytoma and pineoblastoma in 2000. PPTIDs are pathologically classified as grade II or III using the mitotic index and immunohistochemistry.3

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The treatment for pineal parenchymal tumours is determined by histology. Pineocytomas are treated surgically by resection.[4] Even without adjuvant treatment, full or partial resection results in a favourable outcome. In contrast, pineoblastomas should be treated with surgery as well as adjuvant treatments such as chemotherapy and craniospinal irradiation. In some cases, more severe treatment, such as myeloablative chemotherapy with stem cell rescue, may be required to treat pineoblastomas.[5]

CASE REPORT

A 42 year old female patient presented to our hospital with complaints of headache for 1 month, associated with neck pain. All the biochemical parameters are within acceptable limits. Known case of hypertension on medication with no significant family and personal history. Imaging done with CT brain shows hydrocephalus. Contrast Enhanced Magnetic Resonance (CEMRI) suggestive of posterior third ventricular mass-pineal neoplastic lesion. She Underwent endoscopic biopsy and ventriculostomy of third ventricle and tumour biopsy under general anaesthesia. Intra and postoperative periods were uneventful.

On clinical examination, no significant findings were found. Grossly tumour was composed of multiple small brown pieces of tissues. Microscopically the section shows closely packed sheets of round cell with moderately pleomorphic nucleoli with moderate amount of cytoplasm. Pineocytomatous possets are noted with fragments of necrotic tissue identified. Immunohistochemistry showed diffusely positive for synaptophysin and neurofilament protein. Based on the immunohistochemical reactivity and histology, pineal parenchymal tumor of intermediate differentiation (PPTID), low grade (Grade II) was made. Post op CEMRI brain done which showed 2.9x2.3x1.9 cm mass seen in the posterior third ventricle region extending posteriorly into quadrigeminal cistern. The above described lesion shows solid enhancing component with multiple peripheral cystic components. MR spectroscopy of the lesion shows significantly raised choline creatine. Choline NAA ratios and myoinositol peak at low TE. Mild obstructive hydrocephalus.

Differntial diagnosis includes: Astrocytoma and ependymoma.

Summary of Treatment: Patient received adjuvant EBRT using 6MV photons to a total dose of 54 Gy in 30 fractions to planning target volume (GTV) and a dose of 36Gy/20# to craniospinal irradiation by achieving organs at risk constraints as per QUANTEC guidelines in Tomotherapy.

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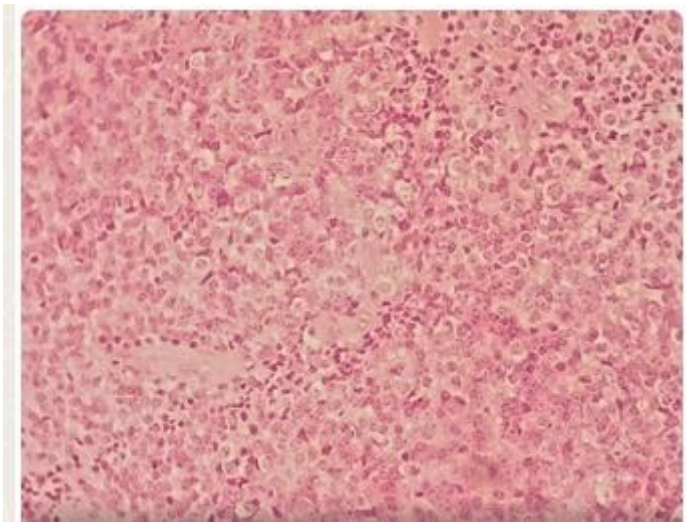

Figure 1: HandE stained picture showing pineacytomatous rosettes are noted, fragments of necrosis noted

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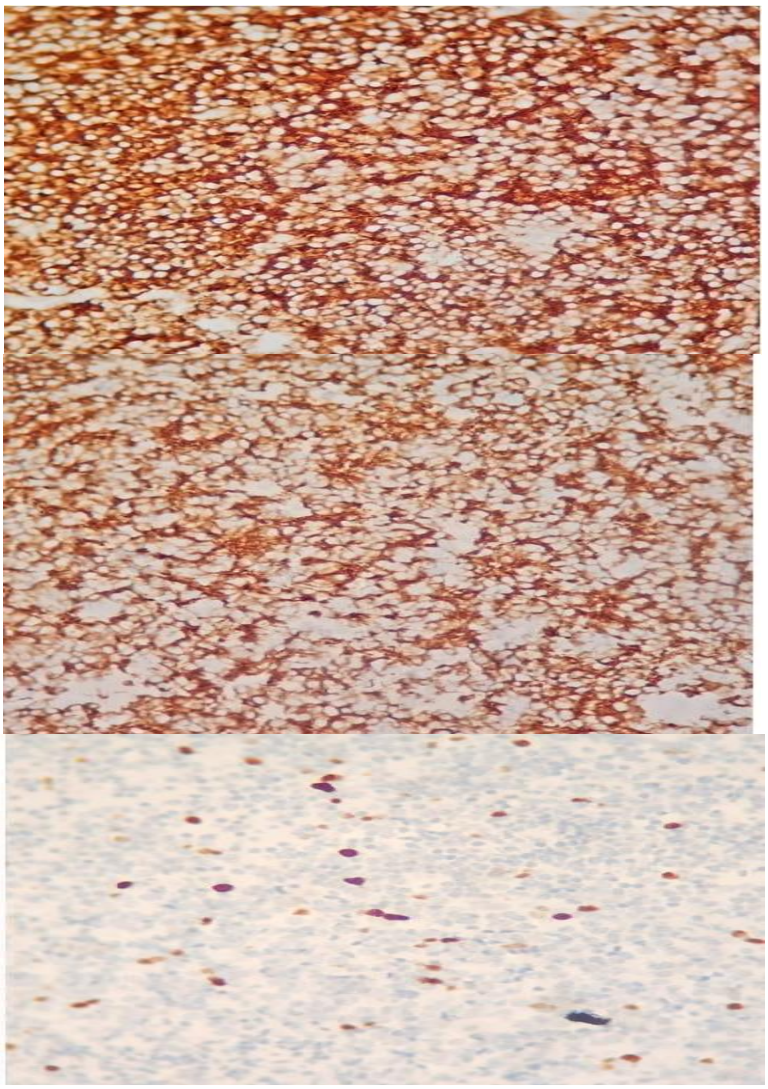

Figure 2: Synaptophysin- diffusely positive, NFP- positive in numerous tumor cells MIB- 5-6%.

DISCUSSION

PPTID first featured in the WHO classification in 2000. PPTID is a type of pineocytoma that is midway between pineocytoma and pineoblastoma. PPTID was split into WHO grades II and III in the 2021 WHO classification, albeit the histological grading standards were not specifically described. PPTID can occur at any age and is similar to pineoblastoma. Neuronal cell markers, such as synaptophysin and NFP, are immunohistopathologically positive, whereas glial cell markers are negative. According to reports, the MIB-1 index ranges between 3% and 10%. Using the mitotic index and NFP expression, Jouvet et al. 4 categorised PPTID into grades II and III;

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grade II contains mitoses less than 6/10 high-power fields and NFP positive expression, whereas grade III has mitoses greater than 6/10 high-power fields and NFP negative expression.

PPTID (pineal parenchymal tumour of intermediate differentiation) is a rare neoplasm with little known about its clinical history and patient fate. It is a more aggressive tumour with more heterogeneity than pineocytoma, with local infiltration and distal CSF dissemination.6 tumours with histological traits intermediate between pineocytoma and pineiblastoma, or tumours with areas of both aforementioned entities, fall under this category.7

A PPTID's clinical appearance is comparable to that of other pineal area tumours. The most prevalent symptoms are dizziness and headache. PPTID, if severe enough, can produce hydrocephalus, resulting in signs of high intracranial pressure such as ataxia. PPTID has a greater patient age range.[8]

In large published series, it has a wide range of reported mitotic counts ranging from 0 to rarely >6/10 hpf. A. Jouvet et al suggested a four-grade prognostic grading system: In low grade PPTID, the 5-year survival rate is 74%, with recurrence occurring in 26% of cases. High grade PPTID has a 5 year survival rate of 39%, with recurrence occurring in 56% of cases.[9]

PPTIDs are classified as grade II or III pathologically based on the presence of necrosis, mitotic rate, and immunohistochemistry expression of neurofilament protein. Grade 2 tumours have 6 mitoses and are strongly immunopositive for neurofilaments, while grade 3 tumours have >6 mitoses or 6 mitoses but no significant immunostaining for neurofilaments. PPTID has been shown to transform into Pineoblastoma.[10]

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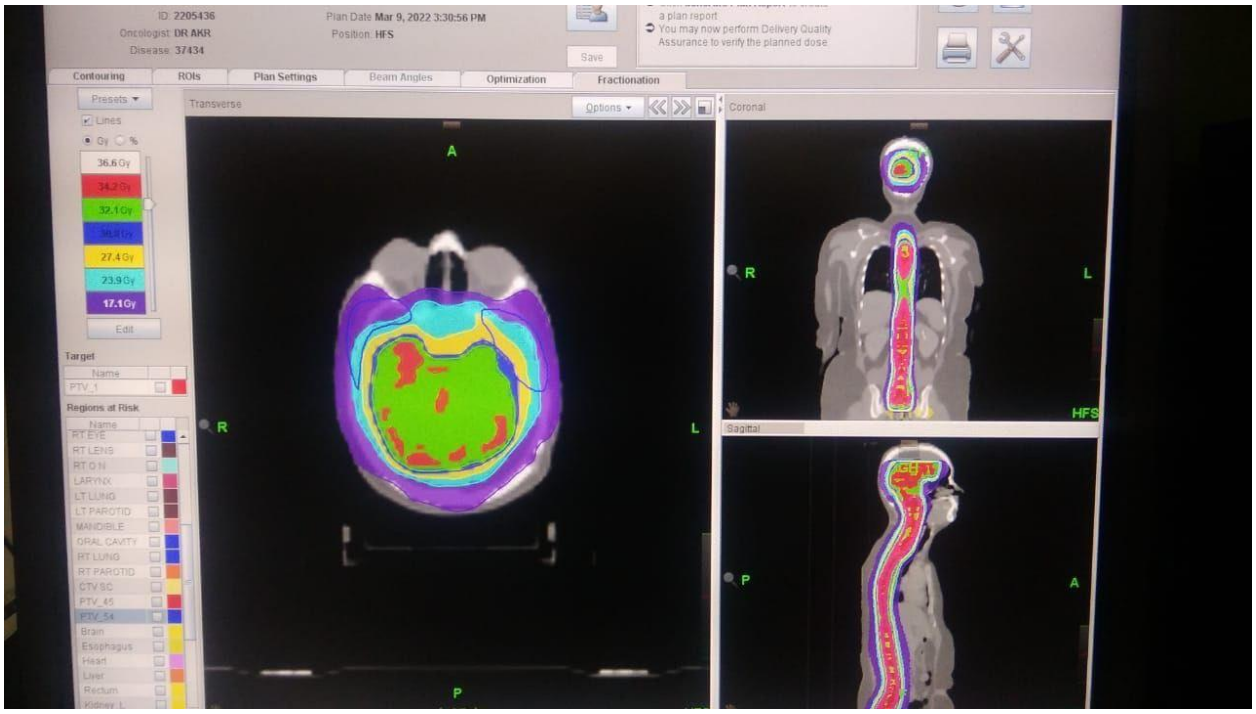

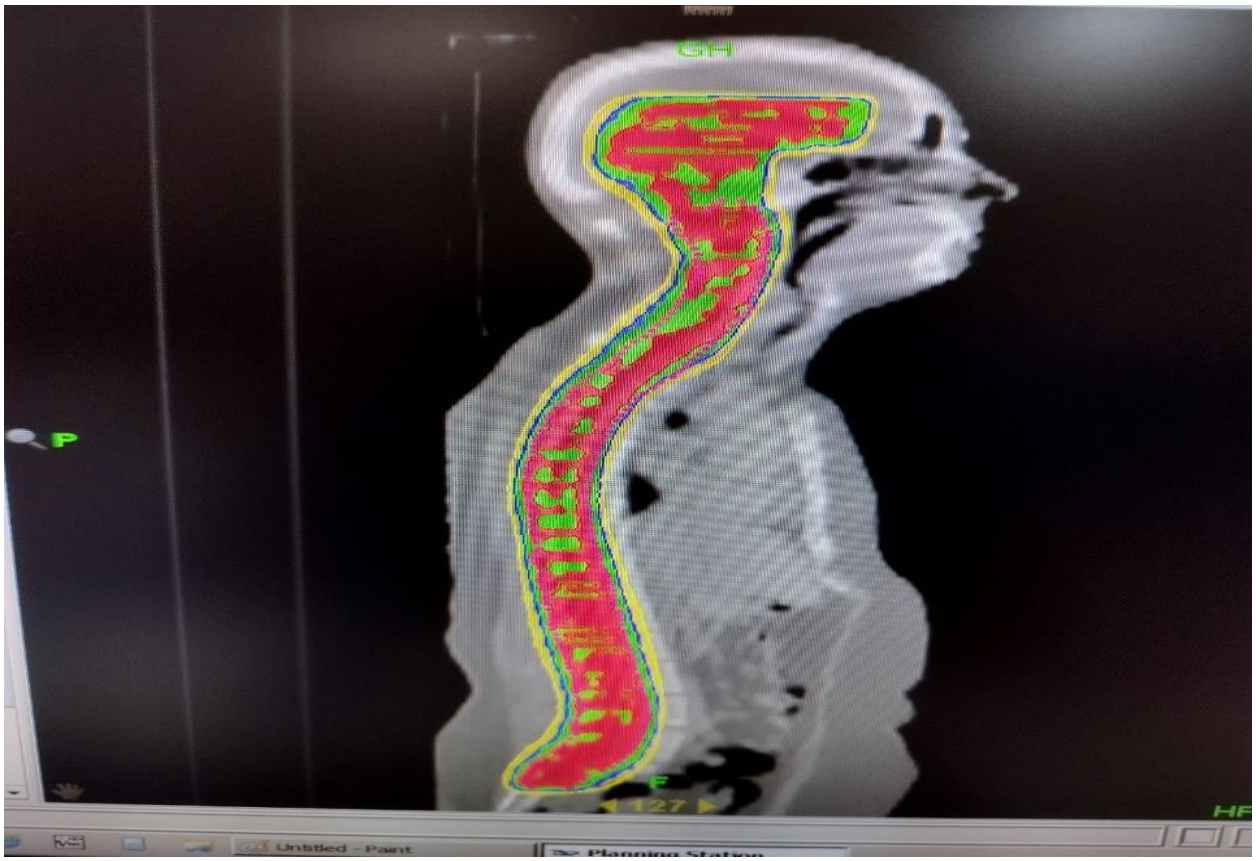

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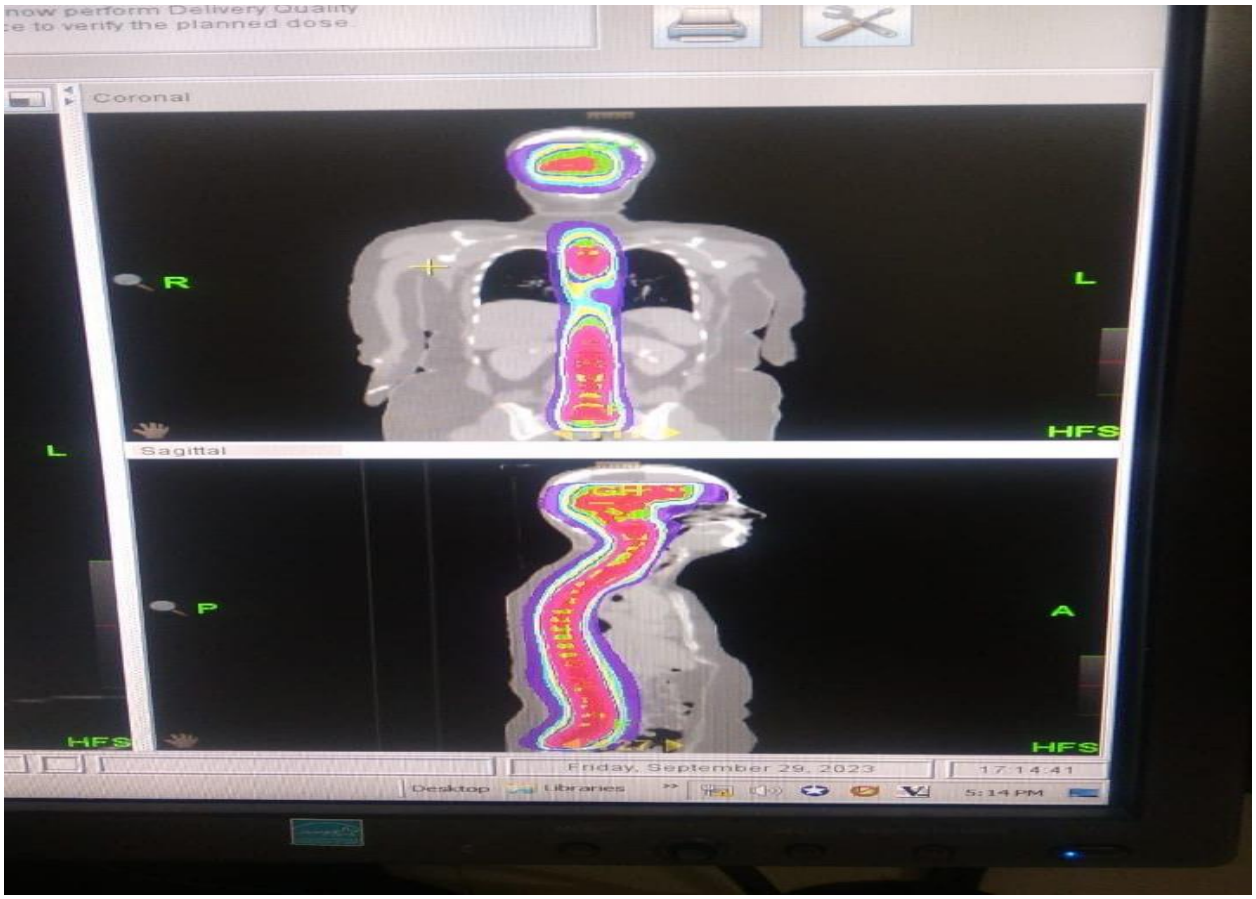

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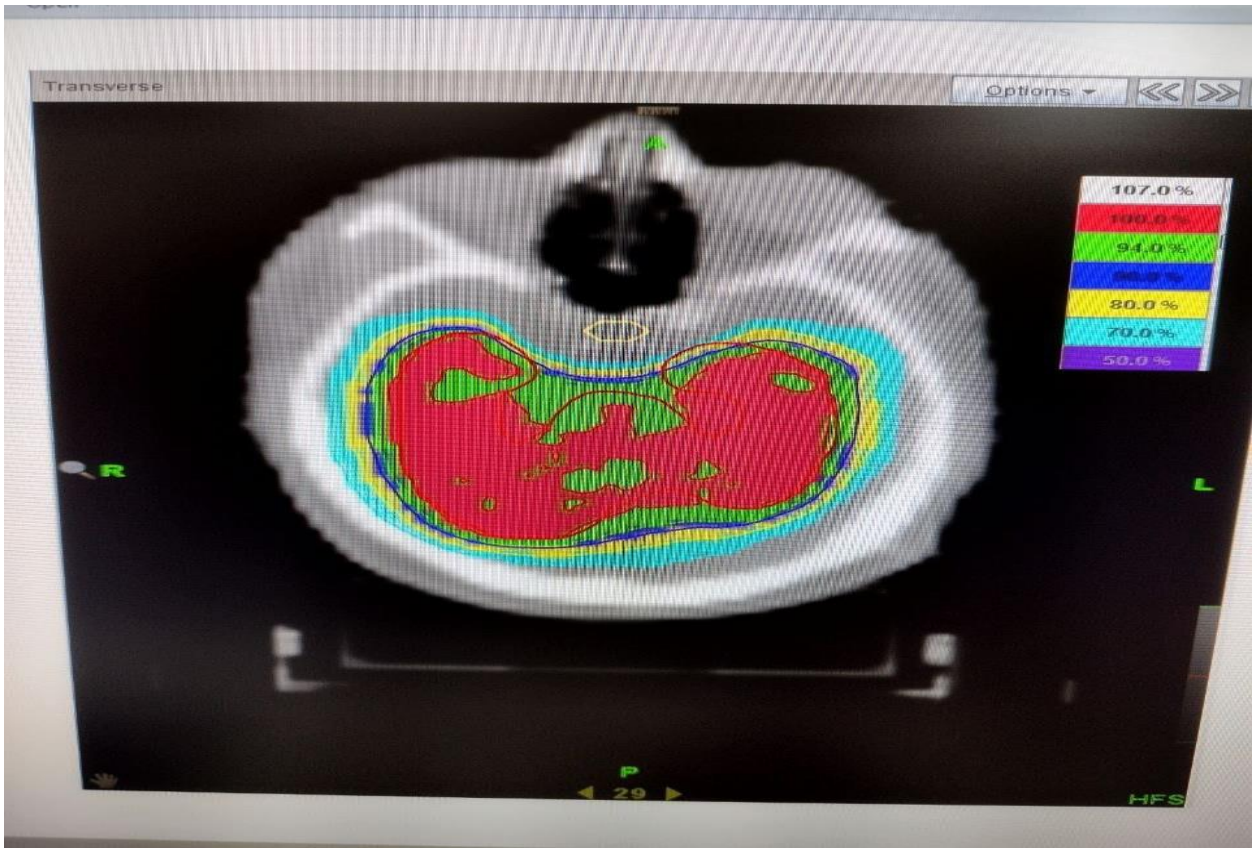

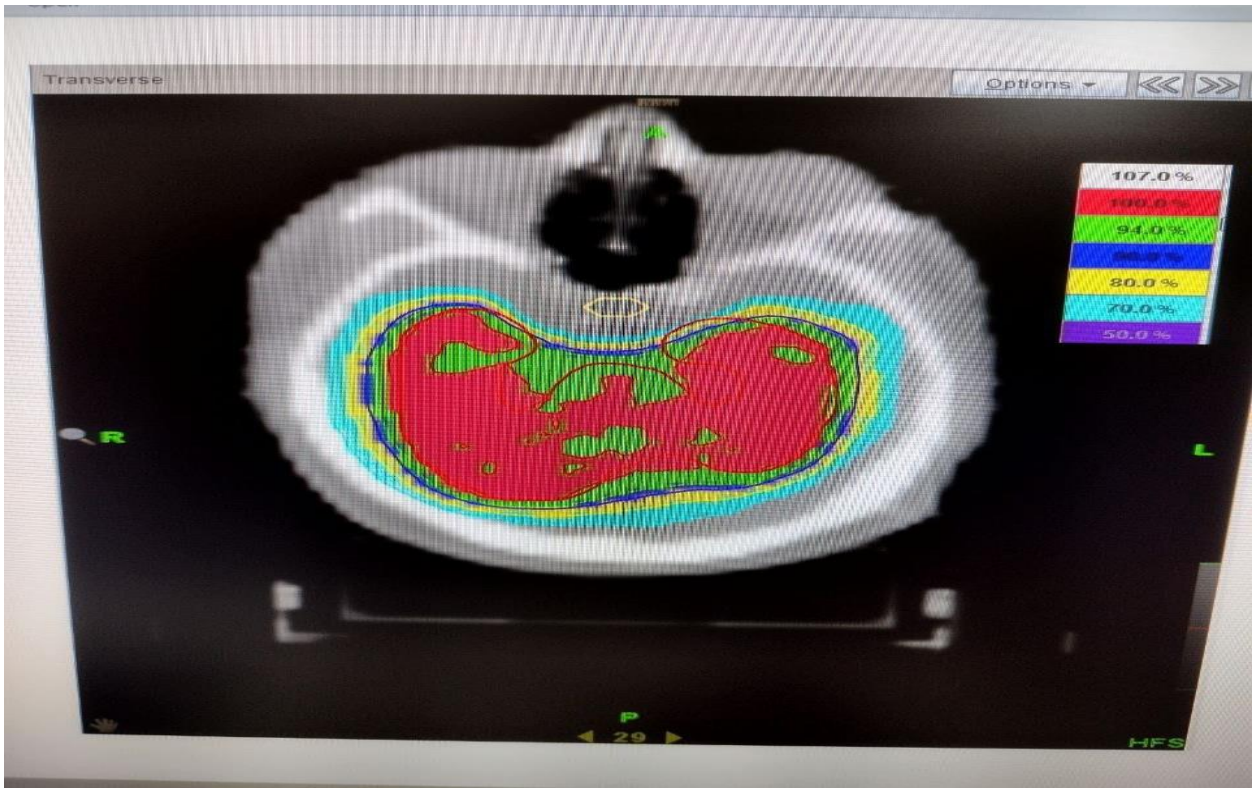

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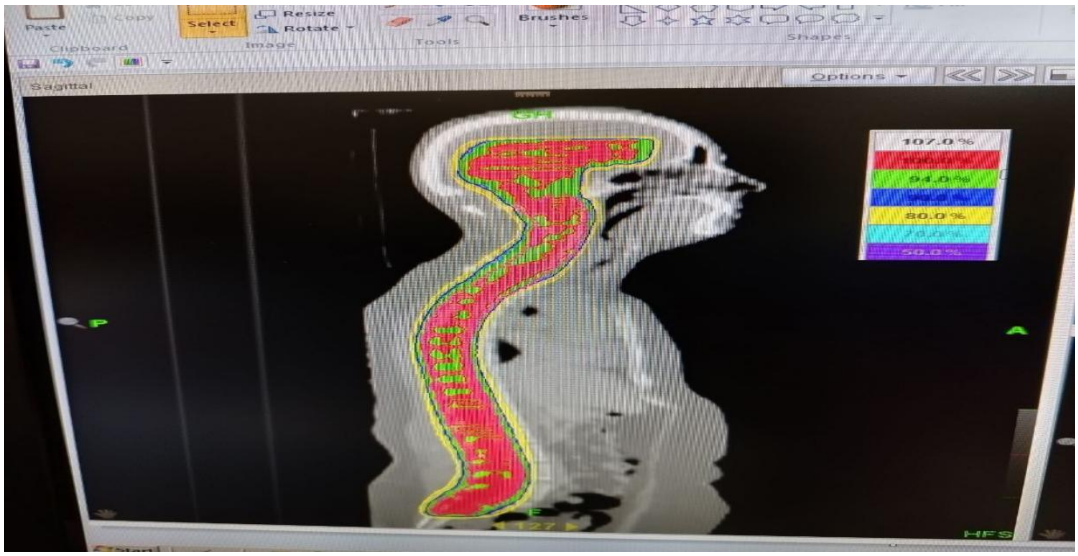

Radiation therapy plan for pineal parenchymal tumor of intermediate differentiation . Isodose distribution around the pineal tumor delivering 54Gy in 30 fractions to the planned target volume of which 36Gy in 20 fractions to craniospinal axis and 46Gy to the brain .

CONCLUSION

PPTID are exceedingly rare tumour entities, but their ideal treatment options are similarly difficult to define due to a lack of research and a small number. Because of the small number of cases and limited data available on the pathological features and biological behaviour of PPTID, the relevance of pineal parenchymal tumour (PPT) grading criteria such as mitotic count, NFP expression, and Ki-67 proliferation index requires further research. Criteria have yet to be developed, and the treatment technique and prognostic prognosis are still debatable.

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