Ependymoma

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Contents

11.1 General Principles of Simulation and Target Delineation (Table 11.1)

- Staging with spine MRI and cerebrospinal fluid sampling is essential to determine if a patient has tumor dissemination.
- CT simulation with a themoplastic mask for immobilization with 1–2.5 mm slice thickness.
- Obtain MRI with T1 pre- and post-gadolinium, T2, and FLAIR for target delineation. Ependymomas often have a mixed pattern of enhancement and may be best visualized on FLAIR sequences.
- Fuse preoperative and postoperative T2/FLAIR and post-gadolinium MRIs to help delineate target volumes.
- If biopsy only, can use preoperative MRI only.

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Table 11.1 Suggested target volumes and doses

Target volumes	Definition and description
GTV1	Residual tumor extent and resection cavity on postoperative T2/FLAIR and T1 post-gadolinium images. Registered preoperative MRIs are helpful for determining residual disease and resection cavity
CTV1	GTV1+ 1 cm. This can be edited around natural boundaries where invasion is unlikely, such as the skull or tentorium (CTV1 is defined as GTV1 + 0.5 mm on COG ACNS0831)
PTV1	CTV1 + 0.3–0.5 cm depending on comfort of patient positioning, mask fit, and image guidance technique (AP/lateral imaging or cone beam CT)
GTV2	The intent of GTV2 is to administer a boost dose to GTV1 but allow volume reduction to limit dose to the spinal cord, brain stem, and optic chiasm after 54 Gy. If dose constraints can be met, GTV2 may be the same as GTV1
PTV2	GTV2 + 0.3–0.5 cm depending on comfort of patient positioning, mask fit, and image guidance technique (AP/lateral imaging or cone beam CT). There is no PTV2 for children under the age of 18 months of age at the start of radiotherapy if gross total resection of tumor is achieved

Suggested dose 54.0–59.4 Gy in 1.8 Gy fractions

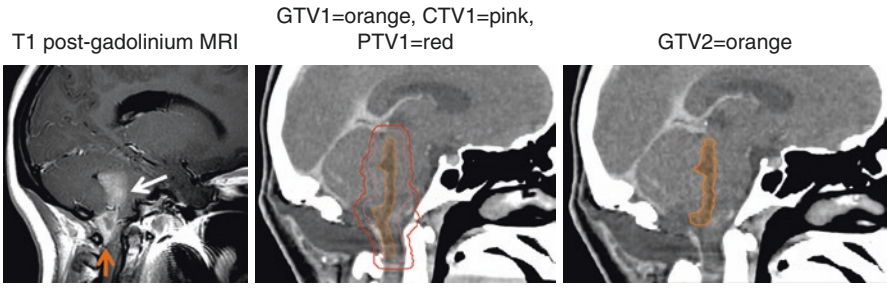

Fig. 11.1 Sagittal images for a patient with WHO grade III anaplastic ependymoma of the fourth ventricle. MRI before gross total resection shows tumor centered in the floor of the fourth ventricle (white arrow) with extension through the foramen magnum to approximately the C2 level (orange arrow). CT simulation with contrast demonstrates initial and boost contours using ACNS0831 guidelines

- If patient has contraindication to MRI, can obtain CT using 1–2.5 mm slice thickness with and without contrast.
- The GTV includes the postoperative residual disease and the edge of the postoperative tumor bed. The edge of any structure in contact with the preoperative tumor should be included, but the surgical tract does not need to be included. The CTV expansion into the brain stem should be limited where invasion or infiltration is not considered likely (Fig. 11.1).
- Contemporary 3D conformal or advanced techniques such as tomotherapy, IMRT, or proton therapy can be considered.

11.2 Dose Prescriptions

- Treatment of the brain after maximal safe resection
  - The current standard doses to the target for intracranial ependymoma are 54–59.4 Gy, and higher doses may be recommended for areas with residual macroscopic disease.
  - The extent of margin for focal radiation therapy continues to be studied with shrinking clinical target volume margins employed on completed and ongoing COG clinical trials.
- Treatment of patients with leptomeningeal dissemination
  - Patients with leptomeningeal dissemination with spinal deposits of intracranial ependymoma generally have a poor prognosis, and treatments should be individualized.
  - Given challenges in the interpretation of CSF cytology in ependymoma, cytology should be repeated in 10–14 days postoperatively to confirm results.

    Craniospinal irradiation is typically indicated after surgery. Target volumes and doses are similar to high-risk medulloblastoma.

11.3 Treatment Planning Techniques

- Contemporary 3D CRT or advanced techniques such as IMRT, VMAT, or proton therapy may be used with the goal of sparing portions of the brain stem, supratentorial brain, hypothalamus, pituitary, optic apparatus, and cochleae (Table 11.2).
- Treatment planning aims to cover 95% of the PTV volume by 95% of the prescribed dose for photon plans and 100% of the CTV volume by 100% of the prescribed dose for proton plans (Fig. 11.2).

Table 11.2 Recommended normal tissue constraints for 1.8 Gy per fraction schemes

Organs at risk	Suggested dose constraints
Optic nerves and chiasm	D50% ≤ 54 Gy and D10% ≤ 56 Gy (goal)[a] D50% ≤ 56 Gy and D10% ≤ 58 Gy (maximum)[a]
Optic globes	D50% ≤ 10 Gy and D10% ≤ 35 Gy (goal)[a] D50% ≤ 20 Gy and D10% ≤ 54 Gy (maximum)[a]
Cochlea	D50% ≤ 35 Gy (goal)[a] D50% ≤ 20 Gy (preferred)[a]
Brain stem (photon)	D50% ≤ 61 Gy and D10% ≤ 63 Gy (goal)[a] D50% ≤ 62 Gy and D10% ≤ 64 Gy (maximum)[a]
Brain stem (proton)	D50% ≤ 52.4 CGE and D0.1cc ≤ 56.6 CGE (goal)[a] D50% ≤ 54 CGE and D0.1cc ≤ 58 CGE (maximum)[a]
Cervical spinal cord (superior-most 6 cm)	D50% ≤ 26 Gy and D10% ≤ 57 Gy (goal)[a] D50% ≤ 50 Gy and D10% ≤ 59 Gy (maximum)[a]

[a]COG ACNS0831 trial [1]

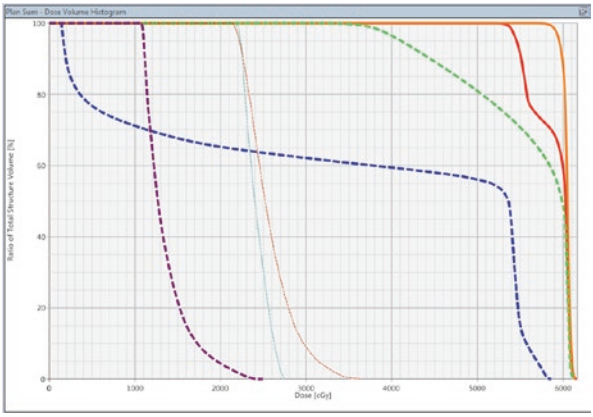

Fig. 11.2 Sample dose-volume histogram for the above patient with WHO grade III anaplastic ependymoma of the fourth ventricle treated with an IMRT plan. PTV2, orange; PTV1, red; brain stem, dashed green; spinal cord, dashed blue; left cochlea, dotted brown; right cochlea, dotted teal; and optic chiasm, dashed indigo

Table 11.3 Side effects

Acute	Hair loss, fatigue, headaches, nausea, diarrhea, fatigue, alopecia, hearing changes, myelosuppression, and cerebral edema causing neurological symptoms
Long-term	Neurocognitive decline, decreased growth, hypopituitarism, hypothyroidism, hearing loss
Uncommon or rare risks	Lhermitte's syndrome, gonadal dysfunction, brain or brain stem injury, secondary malignancies

11.4 Side Effects

Please see Table 11.3.

11.5 Treatment of Recurrence

- While the long-term prognosis for patients with recurrent disease is poor, there is growing evidence that reirradiation is beneficial and has successfully provided local control in carefully selected cases. Patients treated with focal reirradiation remain at risk for development of disseminated metastases or primary site recurrence
- Data in the literature showed that stereotactic radiosurgery can be used for treatment of recurrent intracranial ependymoma; the recurrent tumor, gadolinium enhanced in most cases, alone is targeted, and the typical dose used ranged from 12 to 24 Gy (median 18 Gy) in 1 fraction; the local control rate is 70–80%, but distant failure occurs in at least one quarter of the patients.

Reference

1. https://clinicaltrials.gov/ct2/show/NCT01096368

Further Reading

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