Local Disease Control in Patients with Erdheim-Chester Disease treated with External Beam Radiation Therapy

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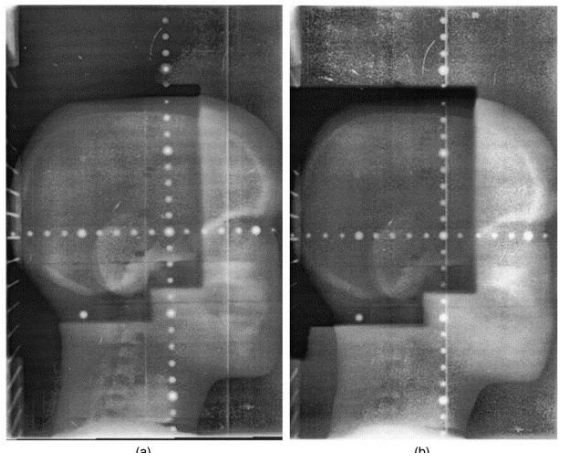
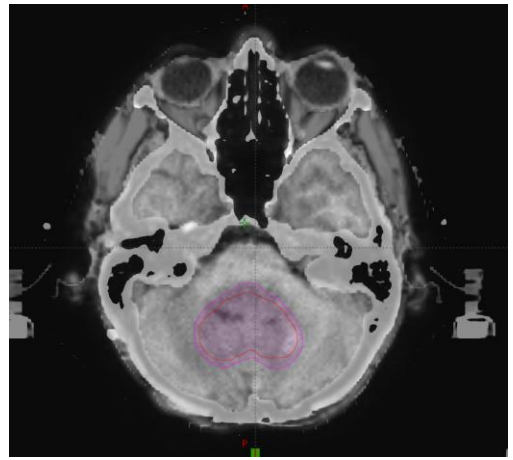
Background

- Radiation is effective for local control at low doses of **10-15Gy** for Langerhans Cell histiocytosis, but data in ECD is limited.
- Some retrospective studies have demonstrated up to **80% long-term control rate** with no significant lasting high-grade toxicity
- **Advances in radiation technology**
  - image guidance, more concise volumes, IMRT vs 3D vs SRS





Kriz et al. *BMC: Radiation Oncology*

Methods

- This is a single institution retrospective case series of patients at Mayo Clinic in Rochester, MN with a diagnosis of ECD
- We identified **8 patients** with history of ECD who have previously been treated with external beam radiation therapy
  - median follow-up of **7.1 years** (range, 0.5-16.6)
  - The median age at time of first radiation treatment **was 47 years (range, 29-67)**
  - Most patients were **female (6 out of 8)**
  - **Bone (57%)** and **CNS (24%)** were the most commonly treated sites

Radiation Site	Dose	Date	Indication	Response	Toxicity
Patient 1:					
Left Tibia	10 Gy in 5 fractions	9/2004	Palliative	• Radiographic progression	• None reported
Right Tibia	10 Gy in 5 fractions	9/2004	Palliative	• Poor pain response • Radiographic progression	• None reported
Right Tibia			Palliative, Re-radiation		• None reported
Patient 2:					
Right Forearm			Palliative		• None reported
Left Forearm			Palliative		• None reported
Bilateral clavicles			Palliative		• None reported
Left Leg			Palliative		• None reported
Right Leg			Palliative		• None reported
Patient 3:					
Right Ilium	10 Gy in 5 fractions	1996	Palliative	• No pain response • Adequate radiographic response	• None reported

NM Bone Scan 1/2013

NM Bone Scan 5/2017

Case #4-5: Two patients treated for cerebellar lesions

Radiation Site	Dose/fractionation	Radiation Date	Indication	Response	Toxicity
- Presented with dysarthria and ataxia - Biopsy and imaging consistent with ECD based cerebellar lesions -Started on prednisone	19.8 Gy in 11 fractions	9/1/2007 - MRI with radiographic progression - Worsening neurologic effects -Restarted on prednisone	Palliative	• Persistent neurologic symptoms • Poor radiographic response	• None reported Last followup: -Persistent neurologic symptoms
Whole brain	16 Gy in 8 fractions	2/14/1997	Palliative	• Initial improvement in neurologic symptoms • Repeat MRI in June 1997 with worsening disease Treated with Cladribine	• None

1995-1996 — January 1997 — June 1997 — 1998 — August 1999

16 Gy in 8 fractions of whole brain radiation

(**19.8 Gy in 11 fractions**), left leg (**18 Gy in 10 fractions**), right mastoid (**18 Gy in 10 fractions**), and right mandible (**15Gy in 10 fractions**)

Case #6: 56 year old female presenting with inflammatory dyspnea, found to have an intra-atrial mass

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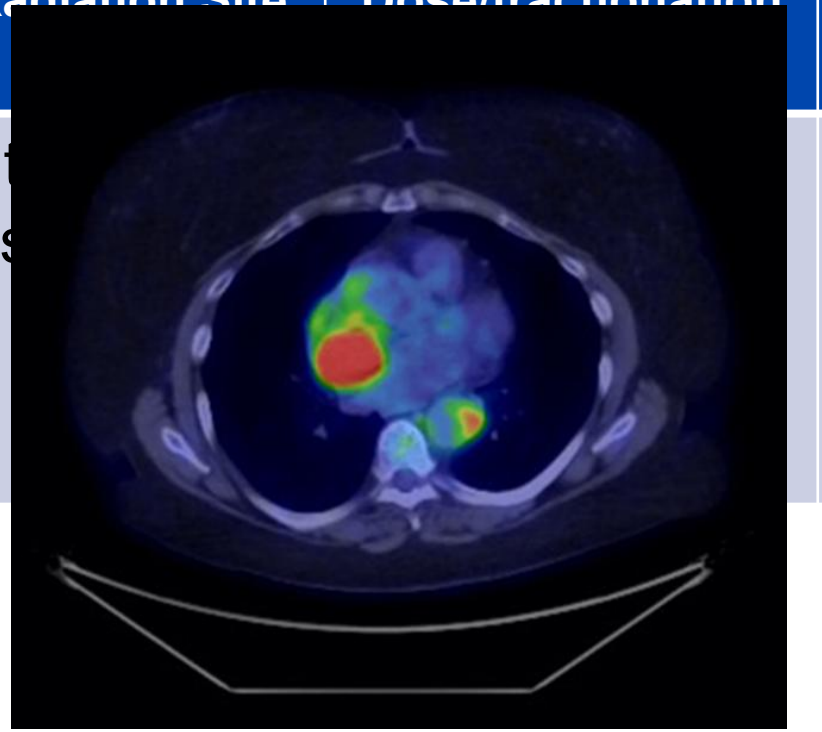
FDG PET: 10/2023



-Started on cladribine

FDG PET: 2/2024

Radiation Site	Dose/fractionation	Indication	Response	Toxicity
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- **18 Gy in 5 fra**
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- stable imaging

Case #7: 38 year old women presenting with lower back pain and sudden left foot to knee numbness, found to have cerebellar and conus medullaris masses

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Radiation Site	Dose/fractionation	Radiation Date	Indication	Response	Toxicity
Cerebellum	45 Gy in 25 fractions	5/4/2016	Palliative	• Stable MRI • Stable weakness/numbness	• Grade 1 fatigue • Grade 1 dermatitis and alopecia • Grade 1 headache
T10-L3 spinal canal lesion	30 Gy in 10 fractions	2/14/2016	Palliative	• Stable/mild improvement on MRI • Stable weakness/numbness	• Grade 1 fatigue • Grade 1 dermatitis and alopecia
C7-T2 spinal canal lesion	30 Gy in 10 fractions	2/23/2024	Palliative	• Pending followup	• Pending follow-up

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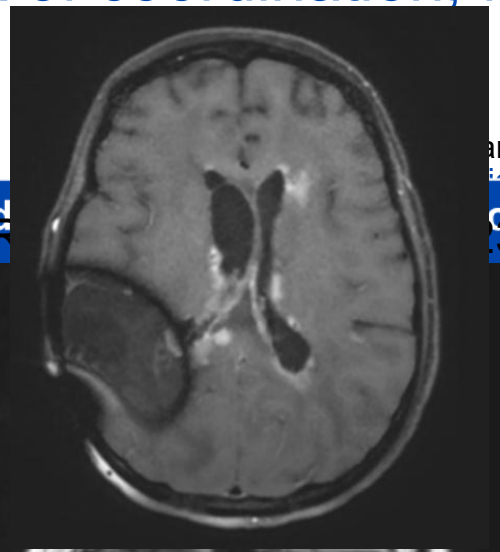
2024

- progression on MRI

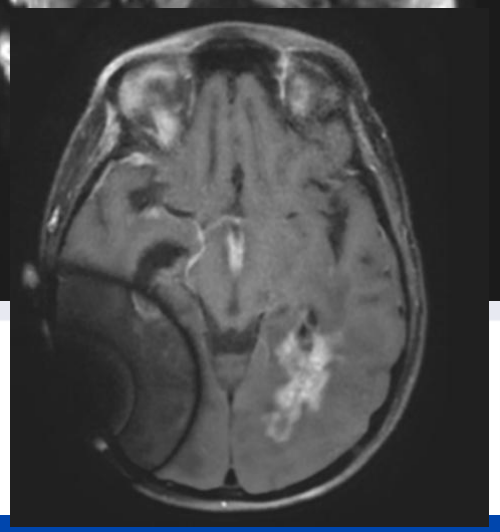
- progression on MRI + new left sided weakness

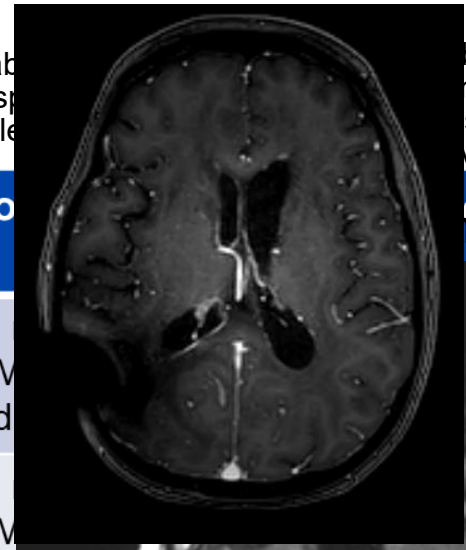
RT at 30 Gy in 10 fractions to T2

Case #8: 27 year old woman presenting with worsening headaches, vertigo, and lack of coordination, found to have leptomeningeal enhancement

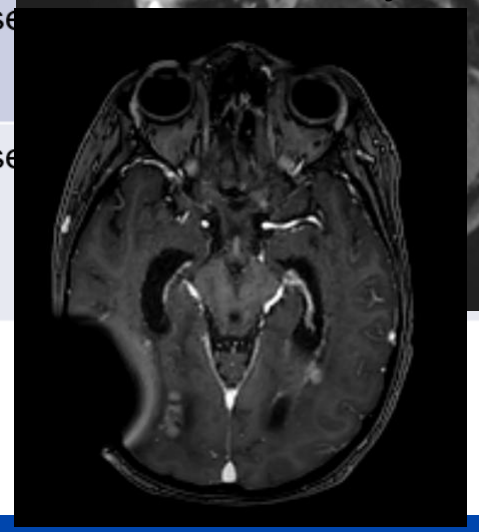


MRI Brain T1: November 2023





MRI Brain T1: February 2024



Results

- There were **17 distinct sites** of disease treated with radiotherapy
  - In disease sites treated with **less than 18 Gy**, 8 out of 9 sites developed in-field recurrence
  - In disease sites treated with **18 Gy or more**, recurrence was noted in 1 out of 8 sites

- Minimal toxicity was noted with radiation therapy at these radiation doses
  - There were **no grade 3+** toxicities

Discussion

- Radiotherapy to involved disease sites may be beneficial for **local control** in ECD
- This is particularly true for disease sites not responding adequately to systemic therapy, such as CNS disease
- With advances in technology, we can deliver radiation **safely** with minimal side effects
- Unlike Langerhans cell histiocytosis which can typically be controlled with **10 Gy**, it appears that doses **of 20 Gy or higher** may be needed for durable local control in ECD
- Limitations: retrospective, single institution, cohort size, followup of 13.5 years (<18Gy) vs 3.5 years (>18Gy)

