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EXTRANODAL ROSAI-DORFMAN DISEASE IN THE PANCREAS: FIRST REPORT OF MRI GUIDED STEREOTACTIC BODY RADIATION THERAPY

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Abstract
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Abstract

Purpose: Rosai-Dorfman disease (RDD) is a rare accumulation of activated histiocytes in the lymph nodes and various other tissues with unknown etiology. There remains no consensus of clinical management or treatment of the disease.1 Areas of histiocytosis mostly include lymph nodes and less often includes cutaneous (10%), nasal sinus (11%), and CNS manifestations (<5%). Pancreatic or hepatic involvement is extremely rare.1 Although Radiotherapy (RT) can be used in patients who are not candidates for surgery, no literature exists for RT for pancreatic RDD. In this report, we describe the case of a 75-year-old male with a diagnosis of RDD involving a pancreatic neck mass that has been successfully irradiated with Stereotactic Body Radiation Therapy (SBRT).

Methodology: The patient's case was reviewed as part of an IRB exempt retrospective study. His clinical record, imaging, dosimetry records, treatment plan, and treatment images were reviewed by the study team.

Results: The patient was diagnosed in 2009 after presenting with a cutaneous site on his shoulder that did not respond to Targretin, Rituxan, or prednisone. He then received palliative radiation of 27 Gy in 9-10 fractions in 2011 for 4 symptomatic skin lesions with resolution. In 2020, yearly surveillance CT scan demonstrated a new mass in the pancreatic neck. Workup with upper endoscopic ultrasound (EUS) confirmed a 2.8 x 2.3 cm mass abutting the splenoportal confluence, with Fine Needle Aspiration (FNA) confirming RDD. Given his age and potential surgical morbidity, the patient elected for MRI-guided SBRT (MRgRT) over a Whipple procedure.

He completed 32 Gy in 4 fractions in August 2021 with no side effects. Serial follow-up CT imaging showed a decreased size in the lesion until a nadir of 0.8 cm, which has been stable. At nearly 2 years post-SBRT, he maintains an excellent quality of life with no long- term sequelae.

Conclusions: The patient’s pancreatic RDD responded completely, similar to the response for his other extranodal sites, demonstrating how a prior history of successfully treated RDD may predict a patient’s response to future treatments. There are reports of RT effectively putting RDD into long-term remission and preserving vital organ function.2 However, depending on the variant of extranodal RDD, the RT technique, dose, and fractionation can vary, with no consensus yet on optimization. This is the first report of a patient with RDD being successfully treated with MRgRT to an extranodal site in the pancreas.

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