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Pancreatic Rosai-Dorfman Disease Diagnosed Without Surgery

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Introduction

- Rosai-Dorfman disease (RDD) is a benign non-Langerhans cell histiocytosis that rarely involves the pancreas
- Prior to this case, only 18 cases of pancreatic RDD have been published to our knowledge, with 14 of these cases requiring surgical intervention often for diagnostic purposes
- Here we present a case of pancreatic RDD diagnosed by EUS-FNA, thus avoiding surgical intervention for diagnosis

Year	Age	Gender	Race	Pancreatic location	Size	Surgery
1990	N/A	F	Black	N/A	N/A	N/A
1999	48	F	Black	Body and tail	4cm	Yes
2009	63	F	Black	Head	2.6cm	Yes
2010	35	F	Hispanic	Tail	10.2cm	Yes
2012	74	F	Black	Head	2cm	Yes
2015	59	F	N/A	Head	N/A	Yes
2016	55	F	N/A	Body and tail	3.5cm	Yes
2016	65	F	N/A	Head, body and tail	1.5cm	No
2017	75	F	Black	Head	4.5cm	No
2019	71	F	Asian	Tail	3.5cm	Yes
2019	65	F	Black	Tail	1.9cm	Yes
2019	65	F	Black	Tail	2.1cm	Yes
2019	51	F	Black	Tail	2.9cm	Yes
2019	47	M	N/A	Body	4.2cm	Yes
2019	69	M	Black	Tail	2.3cm	Yes
2020	40	M	Black	Tail	1.6cm	Yes
2021	70	M	White	Head	4.8cm	Yes
2022	78	M	White	Head	N/A	No
This case	49	F	Black	Body	4.4cm	No

Table: List of the 19 known cases of pancreatic Rosai-Dorfman disease, including year of publication, age in years, gender, race, pancreatic location of mass, size of mass, and whether surgical intervention was pursued. The patient of this case report is listed in the bottom row

Case Description

- A 49-year-old female with DM and HTN initially presented with intermittent abdominal pain
- Initial CT imaging showed a 4cm pancreatic body mass and a 2.3cm retroperitoneal mass, and EUS-FNA biopsies showed findings consistent with RDD
- Patient returned in 2 years with persistent but stable symptoms
- Repeat CT imaging demonstrated progressive growth of the masses; pancreatic mass grew to 4.4cm and retroperitoneal mass grew to 3.3cm
- Pathology of these interval biopsies all demonstrated findings consistent with RDD, including histiocytosis with emperipolesis, and stains positive for S100, CD68, and CD163, and negative for CD1a
- There were no signs of malignancy or IgG4-related disease
- Patient agreed to continue routine surveillance without surgical intervention due to stability of symptoms

Discussion

- Most cases of pancreatic RDD have required surgical intervention at some point of care, oftentimes for diagnostic purposes
- However, some of these cases had biopsies initially thought to be nonspecific, but later shown to have findings of RDD on re-review
- Although there are no consensus guidelines for pancreatic RDD treatment, a wide array of nonsurgical options have been successfully employed including observation (as in our case), immunomodulatory agents, and chemotherapy
- Increased multidisciplinary awareness of pancreatic RDD can potentially avoid unnecessary surgical intervention

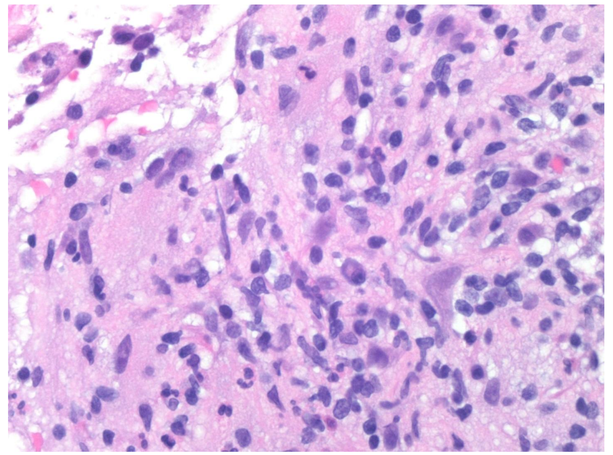

Figure: Atypical histiocytic proliferation consistent with Rosal-Dorfman disease