PARATESTİCULAR ROSAI DORFMAN DİSEASE: A PHENOMENON NOT PREVIOUSLY REPORTED

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ABSTRACT
Rosai–Dorfman disease is a rare extranodal involvement without systemic findings it is also less common, particularly in the paratesticular region. A 28-year-old male presented with a several month history of scrotal swelling without inguinal lymphadenopathy and any associated systemic symptoms. On physical and ultrasound examination revealed a tumor of the right epididim in the scrotum. Histologic examination revealed a dense infiltrate of histiocytic cells. The morphologic and immunophenotypic features were diagnostic for RDD.

KEYWORDS: To our knowledge, this is the first reported case in the literature of scrotal RDD which was located in epididim.

INTRODUCTION
Rosai–Dorfman disease (RDD) is a rare condition characterized by painless massive lymphadenopathy in the cervical region (1,2). While extranodal involvement occurs in up to 41% of systemic cases, with skin being the most common site, extranodal involvement without systemic findings is less common, particularly in the paratesticular region. We present a case of extranodal RDD of the paratesticular region and discuss the imaging and pathologic findings as well as the treatment and outcomes of the entity.

CASE
A 28-year-old male presented with a several month history of scrotal swelling without inguinal lymphadenopathy and any associated systemic symptoms. On physical examination, a 2.5 cm, lesion on the right scrotum was appreciated. Palpation and ultrasound examination revealed a tumor of the right epididim in the scrotum. Head of the right epididim had an izoekoik lobulated mass, measured 14x16 mm. His preoperative blood tests (including tumor markers such as LDH, beta-HGC and AFP) were normal. The abdominal and thoracic CT scans did not show any significant lymph node enlargements or distant metastases. Surgical excision was decided. The preoperative clinical diagnosis was epididim tumor. Intraoperative frozen reasults were benign and the mass was subsequently excised without complication. The surgical specimen measuring 2.5 x 2.5 x 0.7 cm. (Fig. 1).

Histologic examination revealed a dense infiltrate of histiocytic cells. Most of these cells had abundant lightly eosinophilic cytoplasm, round nuclear contours, and single, distinct nucleoli. Admixed lymphoid aggregates with conspicuous plasma cells and scattered granulocytes were present surrounding the sheets of histiocytes (Figs. 2(a–d)). The histiocytic cells were uniformly strongly positive for S100 protein and CD45 (leukocyte common antigen) and variably positive for CD68 by immunohistochemistry; they were negative for CD1a, smooth muscle actin, and pan-cytokeratin (Figs 3(a-d)). The S100 protein immunohistochemical stain highlighted numerous emperipoletic histiocytes. The morphologic and immunophenotypic features were diagnostic for RDD. Postoperatively, the scrotal mass had not recurred, and the patient was stable. Radiographic findings of the patient had no evidence of any additional systemic RDD involvement. He has received no additional therapy and is being followed with observation alone.

Figure Legends

Figure 1. The surgical specimen, measuring 2.5 x 2.5 x 0.7 cm, lobule mass.
DISCUSSION
The characteristic presentation and pathology of RDD were first described by Rosai et al. in 1969.[1] Subsequent reports noted that extranodal involvement occurred in a
significant percentage of patients in various anatomic locations including the respiratory tract, the genitourinary tract, the skin, the bones and soft tissues, and the head/neck and central nervous system among others.\(^\text{2,3}\)

To our knowledge, this is the first reported case in the literature of scrotal RDD which was located in epididim. Although testicular involvement by RDD has been documented occasionally.\(^\text{3,4,5}\) paratesticular involvement of RDD has not yet been described in the literature.

Histologically, RDD classically has a nodular or sheet-like infiltrate of large polygonal histiocytes with large, vesicular, round to oval nuclei and abundant eosinophilic cytoplasm with identification of the hallmark histologic feature of emperipolesis, and the presence of accompanying lymphoid aggregates and scattered plasma cells.\(^\text{7}\) The histiocytes express S100 protein and CD68 and are negative for CD1a, which helps to differentiate these cells phenotypically from Langerhans cells. Although RDD and Langerhans cell histiocytosis are often discussed together because of their shared S100 positivity, the morphologic features are usually sufficient to distinguish between the two entities.\(^\text{7}\)

Regardless of the etiology, RDD is almost always self-limiting and usually spontaneously resolves, although resolution may take years and recurrence is possible.\(^\text{8}\)

Scrotal involvement by RDD raises a broad clinical differential diagnosis that includes both malignant and infectious disorders, as our case demonstrates.

REFERENCES