

Rosai Dorfman Testis: A Rare Case Report

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Abstract:

Rosai dorfman disease was defined by Rosai and Dorfman as an idiopathic histiocytic proliferative disorder afflicting nodes in the year 1969. The disease presents in the 10-20 year age group mainly as fever, painless lymphadenopathy, and hypergammaglobulinaemia, however older ages can also be affected. Although it is mainly a node involving disease, 25% extranodal cases have been reported worldwide. The eyes and ocular adnexa, head and neck region upper respiratory tract, skin and subcutaneous tissue, and central nervous system are the commonly involved extranodal sites. We present a case of extranodal testicular Rosai Dorfman in a 40 year old male with right testicular lump which radiologically mimicked a malignancy but on histopathology was diagnosed to be a benign Testicular Rosai Dorfman.

Keywords: Idiopathic histiocytic pathology; Respiratory tract; Differential diagnosis

Abbreviations: OPD: Out Patient Department; HCG: Human Chorionic Gonadotropin; RDD: Rosai Dorfman Disease; XO: Xanthogranulomatous Orchitis

Case presentation

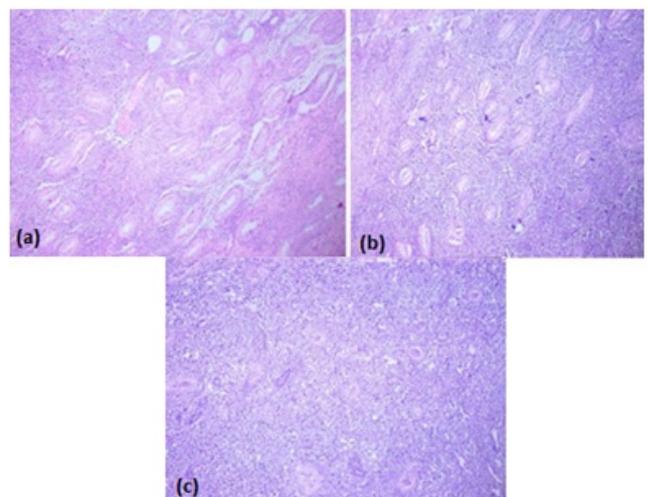
A 40 year old man visited the surgical OPD [Out patient department] of Eras Lucknow medical college and hospital with complains of right side testicular lump for the past 45 days. The lump was not associated with any pain or fever. No lymphadenopathy was recorded and serum beta HCG (Human chorionic gonadotropin) was within normal limits. Ultrasound of testis revealed a mass measuring 6 × 5 cm with increased vascularity suggestive of a testicular neoplasm [1-3]. Orchidectomy was performed and lump sent for histopathology.



Figure 1: Gross image.

Grossly the testicular tissue measured 11 × 9 × 8 cm. Outer surface was smooth and soft. The cut surface showed grey white cystic to solid areas with recognizable uninvolved testicular parenchyma measuring 5 × 4 cm. Testicular cord was recognized and representative sections

taken. On microscopy the sections show sheets of inflammatory cells comprising of plasma cells and lymphocytes with numerous histiocytes showing emperipolesis infiltrating in between the atrophic seminiferous tubules. Also seen was a foci of normal to atrophic tubules at the periphery of the section. Few congested blood vessels and lymphatics were also seen. A diagnosis of Testicular Rosai Dorfman was made finally [Figures 1-4].



Figures 2(a,b,c): [10X Objective] showing inflammatory cells and macrophages with emperipolesis infiltrating in between atrophic tubules.

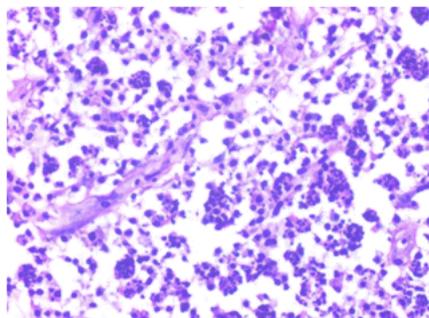


Figure 3: [40X Objective] shows macrophages showing emperipolesis.

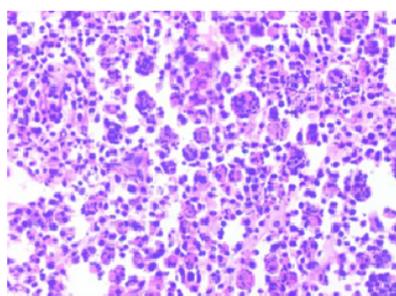


Figure 4: 63X [Objective] showing macrophages with emperipolesis.

Discussion

Despite an extensive search for such cases we could gather very little data about testicular Rosai Dorfman disease [RDD] and ours is the 4th case to be reported to the best of my knowledge. Only 2 cases of testicular Rosai Dorfman have been reported in literature [4]. Although this entity has been described as non-tumorous testicular lesion in books there have been 3 reports of testicular RDD. One in a patient who after 12 years of suffering from lymphoma developed testicular RDD [5]. 2nd report of RDD was of the breast. In 1966 a 3rd case of histiocytic proliferative disease of testis with enlargement, severe lymphadenopathy, transient hepatomegaly, recurrent infections, and rheumatoid arthritis was seen in a boy [5,6]. Extranodal Rosai can occur in other sites like the gastrointestinal tract, salivary glands, genitourinary tract, thyroid, and breast [35]. The spleen and bone marrow are usually not affected. In nodal RDD patients, about 43% also demonstrate at least one site of extranodal disease [7]. The pathological features of RDD have been well described and the microscopic hallmark of Rosai testis is presence of S100 positivity in histiocytes with [8,9]. Xanthogranulomatous orchitis (XO) occurs more frequently as compared to a testicular RDD and hence forms an important differential for this entity [8]. It is thought to be etiologically related to immunological defects, chronic infection and abnormal phagocytosis.

It's a rare entity and usually occurs around 40-59 years of age with sudden onset of tender testicular mass, variable fever. It could be a response to acid fast products of disintegrated sperm, post infectious or due to trauma or sarcoidosis. Microscopically it shows Lymphocytes

and plasma cells infiltrating the interstitium and surrounding seminiferous tubules. Giant cells and histiocytes that resemble (but are not) actual granulomas. Malakoplakia is the other histological differential diagnosis, although the absence of Michaelis-Guttman bodies essentially excluded this likelihood. Grossly the testis is enlarged, tan-yellowbrown and can be associated with abscesses and thrombosed blood vessels. Microscopically tubular atrophy, Histiocytes sheets and Michaelis-Gutmann bodies (intracellular and extracellular round structures containing iron and calcium) can be seen [9,10]. It is very important to exclude a testicular neoplasm, specially burnt out seminomas or seminomas with prominent accompanying inflammatory infiltrates which may disclose few neoplastic cells obscured by inflammation and mimic a RDD [10]. Use of immunostaining for PLAP and CD117 helps in diagnosis. The lack of intratubular germ cell neoplasia is also a helpful feature [5]. Although the pathogenesis of RDD is still not fully understood, infective or immune mediated causes are plausible [11]. The clinical course of RDD is generally benign, though there are reports of progressive disease. Studies have reported that RDD involving the GU system is rare with it most commonly involves the kidney followed by testis and that testicular RDD need IHC for S100 to verify the diagnosis [12].

Conclusion

Testicular RDD is difficult to diagnose preoperatively, and it is worrisome to the clinician as it mimics a testicular neoplasm clinicoradiologically. On microscopy, XO, Burnt out seminoma and Malakoplakia are important differentials. A high index of suspicion is needed to identify typical histiocytes with emperipolesis. The main aim to publish this case report is to not only bring forth the rarity of such a lesion but also to be careful that radiologically these mimic tumors and can become a cause of concern for the surgeon and patient as well and that microscopically they have many benign mimickers as mentioned above which need to be ruled out before giving this diagnosis.

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