Extranodal Rosai Dorfman Disease Mimicking Paraspinal Neoplasm - A Diagnostic Dilemma

Mukhopadhyay G1, Chakrabarti S1 and Shilpi Kundu2*
1Department of Surgical Oncology, Ruby Cancer Center, India
2Department of Pathology, Ruby General Hospital, India

Abstract

Rosai Dorfman disease is a self-limiting rare histiocytic proliferative disorder of unknown aetiology. It classically presents with painless bilateral cervical lymphadenopathy. Extranodal presentation has been documented in the head neck region. We present a rare case Rosai Dorfman disease in 20 years old woman who presented with a painless paraspinal mass for close to a year. This case is being reported for is rarity and unusual anatomic presentation, which to our best knowledge has not been reported in literature.

Keywords: Rosai Dorfman disease; Paraspinal mass; Histiocytic proliferative disorder

Introduction

Rosai-Dorfman disease (RDD) is a rare and benign histiocytic proliferative disorder of unknown aetiology. It usually presents with painless lymphadenopathy with or without extra nodal manifestations.

Case Presentation

A 20 years old lady with history of a gradually progressive swelling over the right lower back for over a year. She had no history of similar swelling elsewhere over her body or any history of trauma over the back. No history similar complaints in first degree relatives. On examination there was a solitary, smooth, ovoid swelling 4 cm × 3 cm, mobile, non-fluctuant, painless, no impulse on coughing over right lower back in the para spinal region. Examination of local and regional lymph node was negative. CT scan of the Lower spine: Revealed a solitary 5.8 cm × 5.78 cm × 2.78 cm solitary hyperdense and enhancing lesion in the subcutaneous plane in the right posterior para spinal region without any extension or bony erosion or calcification or necrosis (Figure 1).

FNAC of the swelling was suggestive of chronic inflammatory Rosai Dorfman disease (Figure 2). Preoperative investigations were performed and patient was taken up for surgery with due consent. Specimen excised was a solitary 6 cm × 6 cm × 3.5 cm with gray-white firm homogenous firm lesion. Microscopy revealed sheets of histiocytes with ovoid, round nuclei and abundant cytoplasm, some of which contained lipid. Nodular collections of small lymphocytes and plasma cells were noted. Some of the histiocytes showed hemophagocytosis and emperipolesis. Immunohistochemistry showed cells positive S-100, CD 68 and negative for CD1a, SMA, CK. Desmin and a final diagnosis of Extranodal Rosai Dorfman Disease was confirmed.

Discussion

Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, was originally described by Destombes in 1965 [1]. Subsequently, it was characterized as a distinct clinicopathological disorder in 1969 by Rosai and Dorfman [2]. It is a benign self-limiting histiocytic proliferative disorder of unknown aetiology, which usually occurs in the first two decades of life. It is believed to be a reactive process and infection or an undefined immunological defect initiated by some other organism is believed to be responsible for the disease. RDD usually presents with painless massive cervical lymphadenopathy. Extranodal involvement is common and is found mainly in the head and neck regions [2]. RDD can also present in any extra nodal site, with common sites including the skin and soft tissue, the Central Nervous System (CNS), and, less commonly, the gastrointestinal tract [3]. Cutaneous Rosai-Dorfman disease (CRDD) is a rare proliferative disorder of histiocytes with unknown etiology, broadly different from systemic Rosai-Dorfman disease. The cutaneous-only form of RDD (CRDD) is a clinically distinct entity from RDD, and some researchers suggest that it may be a different clinicopathological entity from nodal RDD.
Our patient presented with a solitary painless swelling in the right paraspinal area which was increasing in size and causing a lot of mental agony as she had been provisionally diagnosed as a neoplasm elsewhere and counseled for the same. Despite confirmation of diagnosis and reassurance about the non malignant nature of RDD, patient in order to allay her fear she wanted to get operated and reconfirm the diagnosis. RDD is self-limiting, 20% of cases show spontaneous regression without therapy [7]. Relapsing and remitting RDD without treatment has been reported in another 70% of patients in literature, thereby complicating the timing of when to use therapy. Patients requiring treatment, surgery is an appropriate option for disease that can be excised, including single nodal areas, primary CNS involvement, or localized primary RDD [7,8] which was in our case. We have advised regular follow up for our patient. Radiotherapy, cryotherapy, topical chemotherapies adjuvant therapy have been described in literature but with varying success [9,10].

### Conclusion

Rosai–Dorfman disease is a histiocytic disorder with no malignant potential that classically presents with massive, painless lymphadenopathy, fever, and an elevated erythrocyte sedimentation rate. Common extra nodal sites include the skin and the central nervous system. A high degree of clinical suspicion is needed to make the diagnosis because the differential diagnosis includes both malignancy and other histiocytic disorders. For patients who require therapy, surgical resection is the mainstay of treatment. Health care professionals are requested to explain and counsel patients for the same.

### References