Case Report

Rosai-Dorfman disease with paravertebral and epidural thoracic spine involvement: A case report and literature review

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Abstract

Rosai-Dorfman disease (RDD) with spinal cord involvement is a rare clinical entity. We report a case of RDD with paravertebral and intraspinal epidural involvement in a 24-year-old male Bangladeshi patient who presented with progressive bilateral lower limb weakness for 20 days duration associated with spasticity and muscle spasm. MRI demonstrated an enhancing paravertebral soft tissue lesion extending from C7 through T4 with intraspinal epidural extension encasing the spinal cord with focal cord oedema. Histopathology of the paraspinal-epidural lesion reported a finding consistent with RDD. The patient was initiated on high-dose steroids. Follow up after 2 months demonstrated symptomatic improvement as the patient was able to move on the crutch and repeated MRI showed lesion regression.

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Introduction

Rosai and Dorfman in 1969 described Rosai-Dorfman disease (RDD), a rare histiocytic disorder as a separate entity under the term sinus histiocytosis with massive lymphadenopathy (SHML) [1]. Subsequently, RDD is described as a benign, idiopathic and proliferative disease of phagocytic histiocytes mainly affecting children and young adults. There are conflicting data regarding gender and race preponderance. RDD is characterized by massive, painless, bilateral and symmetrical cervical lymphadenopathy, fever, leukocytosis, and high sedimentation rate. Other lymph nodes such as inguinal, axillary, mediastinal and upper paraaortic lymph nodes may also be involved [2].

Extra-nodal disease that frequently affects the respiratory tract, paranasal sinuses, visceral organs, skin, bones, breast, genitourinary tract, and orbits has been documented in 43% of cases [3]. The involvement of the central nervous system (CNS) in RDD is uncommon and cerebral convexities, parasagittal, suprasellar, cavernous sinuses and petroclival regions are the most frequently involved locations [4]. Isolated Rosai-Dorfman spinal cord disease is extremely rare, with only a few cases reported in the literature [5–8].

We hereby report a case of Rosai-Dorfman spinal cord disease in a 24-year-old Bangladeshi man. The main purpose of this case report is to provide health care professionals with the scientific framework to gain a better understanding of the tumor biology, clinical features, pathology, and treatment for RDD.

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24-year-old male Bangladeshi patient presented with progressive bilateral lower limb weakness for 20 days duration associated with spasticity. On physical examination, there was bilateral weakness of the lower extremities (more on the left side) associated with spasticity. The bilateral dorsiflexion power at both ankles was grade II and plantar flexion was grade IV. The power of bilateral external hallucus was grade IV. Flexion and hip extension power of right lower limb was grade IV, while on the left side it was grade II. On both knees, the extension was grade IV. The sensory level was just below the nipple at the T5 level.

An initial CT scan of thoracic spine was performed, which was reported as noncontributory. Therefore, an emergency MRI of cervicothoracic spine and brain was performed to rule out demyelination, which showed paravertebral soft tissue with intraspinal epidural extension encasing the spinal cord with focal cord edema, see Fig. 1 (A, B, C, D, E, and F). A retrospective analysis of CT scan showed focal cortical destruction of right posterolateral aspect of T2 vertebra. Paravertebral and epidural soft tissue lesion was iso to hyperdense compared with the muscle, see Fig. 2 (A and B).

Considering the MRI and CT findings a possibility of an infectious etiology possibly tuberculosis and lymphoma were considered in the differential diagnosis. Patient was admitted by the neurosurgery team and underwent T1-T2 Lamino-plasty and excision of epidural mass. Histopathology of the paraspinal-epidural lesion reported a finding consistent with RDD. Neuro-oncology multidisciplinary team meeting advised PET-CT and referral to hematology team. PET/CT shows paravertebral moderate uptake Fig. 3. The patient was initiated on high-dose steroid. At hematology clinic the patient’s cell blood count, peripheral smear and lumbar puncture were normal. Bone marrow biopsy was hypocellular with histiocytes stuffed with intact hemopoietic cells.

Follow up after 2 months demonstrated symptomatic improvement as the patient was able to move on the crutch and repeated MRI showed lesion regression, see Fig. 4.

Discussion and conclusion

Four cases of a disease were described by Rosai and Dorfman in 1969, which they called SHML. 30 additional cases were analyzed in 1972, establishing SHML as a clinico-pathologic entity [9]. The exact etiology of RDD is not yet known, but various factors are thought to cause RDD such as infectious agents (ie, herpes virus, Brucella or Klebsiella, Varicella zoster virus, Epstein-Barr virus), hematopoietic malignancies, and even a genetic predisposition due to its reported occurrence in pairs of siblings [10-19].
Fig. 2 – G and H: CT thoracic spine in bone and soft tissue window, show a focal cortical destruction of right posterolateral aspect of T2 vertebra (black arrow) and paravertebral and epidural soft tissue lesion was iso to hyperdense compared with the muscle (white arrows).

Fig. 3 – PET/CT shows paravertebral moderate uptake (white arrow).

Children and young adults most commonly get affected by this disease, while there are conflicting data regarding. However, male population and most commonly the individuals of African descent are more commonly affected. Clinically this disease is characterized by painless, bilateral massive cervical lymphadenopathy frequently accompanied by various symptoms such as tonsillitis, malaise, night sweats, fever, weight loss, rhinorrhea, and hepatosplenomegaly, which are nonspecific making the diagnosis difficult [1,9,12,13,20]. Extramedal involvement have some documented sites including bone, skin, genitourinary system, respiratory tract, oral cavity, eyes/orbit/ocular adnexa, CNS, tonsil, breast, salivary gland, soft tissue, and heart.

In 1 study, multiple lymph node swellings of various sites involved were observed in RDD patient [21]. According to this study, low-grade fever is usually present along with hyperglobulinemia, elevated ESR, normochromic normocytic anemia and leukocytosis, which are nonspecific clinical findings and the patient’s response was good to high doses of steroids [9]. In our case, there was no associated fever, nor was there any associated anemia, elevated ESR, leucocytosis, or hyperglobulinaemia while there was bilateral lower limb weakness and spasticity; left-sided weakness was more than right associated with spasticity.

Radiological manifestation of Rosai-Dorfman disease can be extremely variable due to magnitude of organ systems which can be involved. Most common imaging finding on conventional and cross-sectional imaging is lymphadenopathy. Most common group of lymph nodes involved are the cervical nodes. Intracranial and spinal involvement in Rosai-Dorfman disease is uncommon. In the central nervous system, the disease may manifest as meningeal-based masses which appears as hyperattenuating on CT due to high cellular content. On MRI the lesion appears isointense to spinal cord on T1, is to hypointense on T2 with homogenous avid postcontrast enhancement. T2 hypointensity related to high cellular content can suggest the diagnosis of RDD. In the spine the disease may manifest as epidural or intradural extramedullary mass. The lesion often shows increased uptake with gallium scan and increased metabolic uptake with FDG-PET.
Rosai-Dorfman disease confined to the epidural thoracic spine is extremely rare [4–8,22]. Constitutional symptoms are notably absent in patients with CNS RDD [23]. Similarly, our case showed no constitutional symptoms. While CT was not performed or not documented in the case reports of isolated spinal RDD before, this patient had vertebral body erosion and paravertebral lesion which made us to think of an infectious etiology. Many of the authors reported heterogenous signal intensity on DWI and increased signal intensity on apparent diffusion coefficient (ADC) mapping which was almost keeping with this patient.

Differential diagnoses of isolated spinal RDD include tuberculosis, lymphoma and meningioma. In tuberculosis there is endplate and disc destruction with paravertebral collection which usually show restriction on ADC. Lymphomas are isointense on T1 and iso to hyperintense on T2 with avid post contrast enhancement. Hypercellular lymphomas can be hypointense on T2 similar to RDD. Majority of the meningiomas are intradural rather than epidural [24]. Meningiomas are isointense on T1, iso to hyperintense on T2 with avid post contrast enhancement.

The diagnosis of RDD is a challenge, a thorough radiological review and a high degree of suspicion are necessary to diagnose this rare clinical entity. Some of the patients may experience spontaneous resolution, while others may require steroid therapy or surgical resection alone or in conjunction with chemotherapy or radiotherapy.

In conclusion, spinal cord involvement in DDR is a rare clinical entity that mimics lymphoma and tuberculosis. The diagnosis is challenging and needs high index of suspiscious. In proper clinical setting T2 hypointensity and avid homogenous post contrast enhancement can suggest the diagnosis of RDD.

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