

Spastic Quadripareisis and Communicating Hydrocephalus as Late Sequel of Rosai–Dorfman Disease: A Case Report and Review of Literature

Abstract

Rosai–Dorfman disease (RDD) predominantly affects cervical lymph nodes and presents with fever and pancytopenia. Central nervous system involvement though uncommon is often reported. Hydrocephalus and paraparesis as a consequence of RDD is an extremely rare entity. We present a 58-year-old male, diagnosed and treated for RDD with cervical lymphadenopathy, who now presented with spastic paraparesis and on evaluation was found to have communicating hydrocephalus that resolved after ventriculoperitoneal shunt surgery.

Keywords: *Hydrocephalus, lymphocyte phagocytosis, Rosai–Dorfman disease, spastic quadripareisis*

**Ankur Kapoor,
Pravin Salunke,
Chirag K. Ahuja¹,
Debjyoti Chatterjee²**

*Departments of Neurosurgery
and ¹Radiodiagnosis,
²Histopathology, PGIMER,
Chandigarh, India*

Introduction

Sinus histiocytosis with massive lymphadenopathy, also known as Rosai–Dorfman disease (RDD) described in 1969, is an idiopathic histiocytic proliferation affecting lymph nodes.^[1] Although extranodal involvement has been reported in diverse sites, central nervous system (CNS) involvement is quite rare.^[2–5] There are only a few isolated reports of intraventricular or subarachnoid involvement in this entity. We present a similar rare case, where 8 years after successful treatment for systemic RDD, a 52-year-old male presented with spastic quadripareisis and had communicating hydrocephalus that resolved after shunt surgery.

Case Report

A 58-year-old male presented with a history of generalized weakness, weight loss, and multiple swellings all over the body for 1 year. The evaluation revealed normochromic normocytic anemia, pancytopenia, polyclonal hypergammaglobulinemia, and widespread generalized lymphadenopathy. Erythrocyte sedimentation rate was 48. Fine needle aspiration cytology from cervical lymph node was suggestive of RDD [Figure 1]. He was started on 6-mercaptopurine and prednisolone and responded well. Symptoms resolved completely in few days

and patient remained well for next 6 years when presented again, this time with stiffness bilateral lower limbs, difficulty in urination in the form of poor stream and increased frequency. Examination revealed mild spasticity in all four limbs with no sensory deficit. There were no residual signs of the previous disease. Gadolinium magnetic resonance imaging (MRI) of brain and spine revealed no abnormality other than slightly dilated ventricles. The disease maintained static course for the next 5 years, and then symptoms worsened in the past 1 year in the form of increasing stiffness, weakness in the hand grip, spastic gait, and spastic dysfunction of bladder. Repeat MRI brain revealed communicating hydrocephalus. Spine MRI revealed no abnormality [Figure 1]. The patient was taken up for ventriculoperitoneal shunt. After shunt surgery, the patient noticed an improvement in gait and spasticity. Repeat MRI of brain and spine 8 weeks later revealed no additional lesion and resolved hydrocephalus.

Discussion

RDD classically presents in children and adolescents with massive painless cervical lymphadenopathy, often with associated fever, anemia, polyclonal hypergammaglobulinemia, and an elevated sedimentation rate.^[2] The majority of patients are younger than 20 years of age. Ninety percent of patients with this

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Address for correspondence:

*Dr. Ankur Kapoor,
Flat No. E-33, GHS 94,
Sector 20, Panchkula - 134 116,
Haryana, India.
E-mail: dr.ankurkapoor@gmail.
com*

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condition have bilateral cervical adenopathy that is massive but painless. The illness typically lasts for several months to years, but usually, results in complete recovery.^[3]

Extranodal RDD is a known entity, seen in 43% of cases, with common sites being the skin, orbit, and upper respiratory tract. When intracranial, the lesions are usually dural based, are extraparenchymal and so can mimic meningioma. Commonly affected intracranial sites are cerebral convexities, the parasagittal, suprasellar, cavernous sinuses, and petroclival regions^[2] while spinal involvement is commonly based on dura, though rarely intramedullary location is also reported. On MRI, the lesions may appear slightly hyperintense on T1-weighted (T1W) images, enhance after gadolinium administration, and show hypointensity on T2W studies. The hypointensity seen on T2W MRI is quite peculiar and helps in differentiating from similar looking meningioma.^[1] Tumefactive sarcoidosis may simulate the radiological picture by being hypointense on T2W image.^[2]

Typical histopathological features are marked dilated nodal sinuses filled with mature histiocytes showing phagocytosis of blood cells and proliferation of plasma cells.^[3,4] Cytology is remarkable for histiocytes intermixed with plasma cells and lymphocytes and positivity for S-100 protein and CD68. Staining for CD20 and CD3 may show a mixed population of B- and T-lymphocytes in the background. One can often find evidence of emperipolesis (lymphocyte phagocytosis). The etiology of RDD is not well understood.^[5] The unusual response of the hematolymphoid system against an immunologic disorder is one presumed cause while Epstein–Barr virus and human herpes virus six is the other.^[2,3]

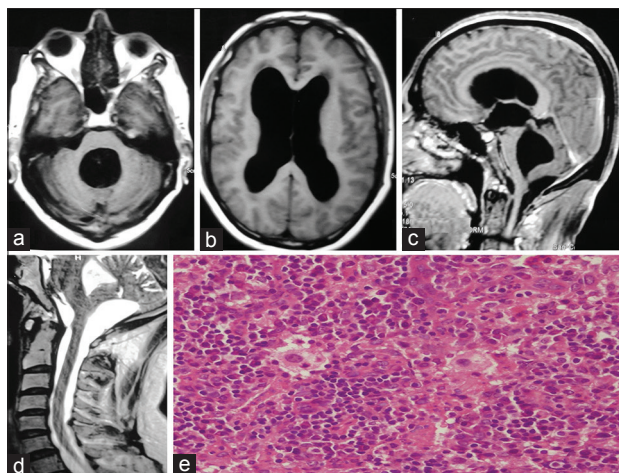


Figure 1: (a-c) Preoperative magnetic resonance images showing tetra-ventricular hydrocephalus with distorted fourth ventricle compressing brainstem as seen on plain T1-weighted axial images (a and b) and on sagittal T1-weighted contrast (c). On sagittal T2-weighted image of magnetic resonance, cervical spine dilated the fourth ventricle, and normal thecal sac is appreciated (d). (e) Photomicrograph showing histiocytes with emperipolesis in a background of plasma cells and mature lymphocytes (H and E, ×400)

Spinal affection is extremely rare. Foucar *et al.* had noticed spinal disease in four of their 200 patients, and the symptoms pertained to weakness due to spinal cord compression.^[6] Spinal epidural infiltration causing paraparesis has been documented on two previous occasions. In one the disease responded to surgery and in the other to immunosuppressants.^[7,8] Most previous reports describe intra- or extra-dural lesions when spinal compression is present, but significant spastic weakness with only hydrocephalus on imaging is extremely rare. We could find only one prior report in literature mentioning isolated fourth ventricular affection in RDD. The improvement in spasticity after shunt surgery indicates toward tetra-ventricular hydrocephalus as the cause for spastic quadriplegia.

Growth rates of such lesions are unpredictable. The disease may remain quiescent for months to years and then recur. Our patient became symptomatic 6 years after successful treatment of systemic disease by immunosuppressants. Abou-Zeid *et al.* mentioned about thoracic involvement in RDD. After initial histological diagnosis by laminectomy, the patient responded to immunosuppressants but later had to be taken up for surgery for rapid progress of cord compression in 6 months' time.^[9]

The management principles in such cases with intracranial or intraspinal affection remain controversial. Although the majority of dural-based lesions are excised, there are several reports of satisfactory disease control even with subtotal excision or biopsy followed by steroids. Our patient presented with late CNS manifestation of RDD in the form of communicating hydrocephalus and spastic quadriplegia. The distortion of the fourth ventricle and extraventricular cisternal anatomy with apparent stenosis of the fourth ventricular outflow tract suggested the development of meningeal adhesions due to prior subclinical meningeal disease. This was possibly overlooked at the time of initial diagnosis of RDD as no neuroimaging was performed. However, the meningeal adhesions might have developed sequentially and manifested later with decompensated hydrocephalus. As all other causes of hydrocephalus were excluded by appropriate investigations, this phenomenon seems the most likely explanation of hydrocephalus. We, thus, emphasize on long-term follow-up of patients with RDD and suspicion of CNS involvement in those with clinical features of spasticity and raised intracranial pressure (ICP).

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Conflicts of interest

There are no conflicts of interest.

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