Extra-Axial Mass in the Foramen Magnum Causing Cervical Compressive Myelopathy as a Complication of Rosai-Dorfman Disease

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Dear Editor,

Sinus histiocytosis with massive lymphadenopathy, or Rosai-Dorfman disease (RDD), is a rare histiocytic proliferative disorder characterized clinically by massive adenopathy and systemic symptoms. The extranodal manifestations that occur in 43% of all RDD patients most frequently affect the skin, respiratory tract, paranasal sinuses, orbits, and bone.¹ CNS involvement is extremely rare (in <5% of cases), and usually consists of an extra-axial dura-based lesion; the most common locations are the convexity and the base of the skull.¹

We present a case of RDD with extranodal involvement at the foramen magnum extending toward the cervical spinal canal and resulting in compressive cervical myelopathy.

To the best of our knowledge, only three cases of RDD displaying spinal cord compression secondary to craniocervical junction involvement have been reported since the disease was first described in 1969.²⁻⁴

Our patient was a 28-year-old man who had been diagnosed with RDD in childhood. In September 2012, he was seen in the neurology department due to a 1-week history of loss of strength in his right arm. The neurological examination showed mild tetraparesis (4+/5 in the left limbs, with more severe right arm involvement: 3/5 proximally and 1-2/5 distally), symmetrical 2/4 deep tendon reflexes, and tactile hypoesthesia and hypoalgesia in the right C5-C8 territories.

Brain MRI revealed extensive nasal and sinus involvement (Fig. 1A). A cervical MRI scan revealed an extra-axial lesion in the posterior region of the craniocervical junction and extending from the foramen magnum to the base of the odontoid process. This mass was causing severe foramen magnum stenosis and compressive cervical myelopathy (Fig. 1B and C).

After assessment by the neurosurgery department, he underwent suboccipital craniectomy and complete tumor resection. Cytology study of the excised tissue revealed that the spinal cord parenchyma was infiltrated by histiocytes exhibiting emperipolesis (lymphophagocytosis). Immunohistochemical staining was positive for the markers CD68 and S-100 and negative for CD1a (Fig. 1D-I). The postoperative results were favorable, with the neurological symptoms resolving completely. After 4 years of follow-up, our patient is still being treated with oral corticosteroids and has presented no additional extranodal lesions in the foramen magnum.

CNS involvement is rare in RDD, and it usually presents in the form of a well-defined, solitary, extraparenchymal supratentorial dura-based lesion resembling a meningioma in neuroimaging scans.³ Lesions located in the posterior fossa and extending to the cervical spinal canal, as in our case, are rare.

Accompanying focal neurological signs depend mainly on the location of the primary lesion; these signs may be subacute, chronic, or recurrent. In all three previously reported cases

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Fig. 1. MRI images. A: Brain T2-weighted sagittal sequence showing paranasal sinus obliteration and heterogeneous signal hyperintensities. B: Noncontrast cervical T1-weighted sagittal sequence showing an extra-axial lesion located at the craniocervical junction and extending from the foramen magnum to C2. C: Postcontrast cervical T1-weighted sagittal sequence displaying intense homogeneous contrast uptake. A microscopic examination of the lesion was performed. D: The histiocytes in the infiltrate are large, with abundant, lightly eosinophilic or clear cytoplasm and vesicular nuclei. Hematoxylin and eosin; bar represents 200 μ m. E: A histiocytic cell (arrow) engulfing small lymphocytes (emperipolesis). Hematoxylin and eosin; bar represents 200 μ m. F: Peripheral nests of meningeal hyperplasia (arrows). Rosai-Dorfman histiocytic infiltrate in the right lateral border. Hematoxylin and eosin; bar represents 600 μ m. G: Abnormal histiocytes showed strong positivity for S-100 protein. H: The histiocytic nature of the cells is highlighted by the immunohistochemical marker CD68. I: The cells were immunohistochemically negative for CD1a.

of RDD with craniocervical junction involvement, the most frequent neurological symptoms were motor deficits (in two of the three patients) and cervical pain.²⁻⁴ The patients described by Ambekar et al.³ and Sandoval-Sus et al.⁴ presented prolonged insidious symptoms that had developed for more than 1 year before they were assessed by a neurosurgery department.^{3,4} In contrast, the severity of our case (tetraparesis and right-sided sensory alterations developing within 1 week) resulted in an early diagnosis of spinal cord compression; our patient was treated surgically less than 1 month after the diag-

nosis.

Treatment approaches for RDD remain controversial. Surgical resection is regarded as the best treatment option for patients with combined RDD and CNS involvement.⁴ Our patient underwent cervical spinal cord decompression with suboccipital craniectomy due to the extreme risk associated with the location of the lesion.

In conclusion, although RDD is rare and spinal cord compression is an unusual form of extranodal involvement, this disorder should be included in the differential diagnosis of

spinal cord masses.

Conflicts of Interest

The authors have no financial conflicts of interest.

REFERENCES

1. Chen H, Zhou H, Song Z. Intracranial multifocal Rosai-Dorfman disease. *Neurol Neuroimmunol Neuroinflamm* 2016;3:e293.
2. Tubbs RS, Kelly DR, Mroczek-Musulman EC, Hammers YA, Berkow RL, Oakes WJ, et al. Spinal cord compression as a result of Rosai-Dorfman disease of the upper cervical spine in a child. *Childs Nerv Syst* 2005; 21:951-954.
3. Ambekar S, Somanna S, Bhat DI, Ranjan M. Isolated cranio-spinal involvement of Rosai-Dorfman disease: case report. *Br J Neurosurg* 2011; 25:297-299.
4. Sandoval-Sus JD, Sandoval-Leon AC, Chapman JR, Velazquez-Vega J, Borja MJ, Rosenberg S, et al. Rosai-Dorfman disease of the central nervous system: report of 6 cases and review of the literature. *Medicine (Baltimore)* 2014;93:165-175.