SNI
SURGICAL NEUROLOGY INTERNATIONAL
SNI: Spine
OPEN ACCESS
For entire Editorial Board visit : http://www.surgicalneurologyint.com
Editor: Nancy E. Epstein, MD Winthrop Hospital, Mineola, NY, USA

Case Report

Dorsal hemangioblastoma manifesting as holocord syringomyelia

Gautam Dutta, Daljit Singh, Hukum Singh, Arvind K. Srivastava, Anita Jagetia, Atul Agrawal

Department of Neuro-Surgery, Govind Ballav Pant Institute of Postgraduate Medical Education and Research (GIPMER), New Delhi, India

E-mail: *Gautam Dutta - gautamblue@hotmail.com; Daljit Singh - drdaljit@hotmail.com; Hukum Singh - drhsingh2008@hotmail.com; Arvind K. Srivastava - aksrivastava2008@gmail.com; Anita Jagetia - anitajagetia@gmail.com; Atul Agrawal - dratul_atul369@rediffmail.com
*Corresponding author

Received: 08 February 18 Accepted: 21 February 18 Published: 05 April 18

Abstract

Background: Intramedullary spinal hemangioblastomas are known to be accompanied by syringomyelia.

Case Description: Here, we report a patient who presented with symptoms of a Chiari malformation but was found to have a D4 intramedullary hemangioblastoma with a holocord syrinx.

Conclusions: Although rare, neurosurgeons should keep in mind the possibility of an intramedullary hemangioblastoma in patients presenting with symptoms of a Chiari malformation.

Key Words: Chiari malformation, differential diagnosis, hemangioblastoma, holocord, intramedullary, spinal, syringomyelia

Access this article online
Website: www.surgicalneurologyint.com
DOI: 10.4103/sni.sni_47_18
Quick Response Code:

INTRODUCTION

Spinal hemangioblastomas compromise 1.6–2.1% of all primary spinal cord tumors. They are located in cervical or thoracic region, and are typically highly vascular solid and/or cystic lesions. Although they may be associated with syringomyelia, they only rarely present with holocord syrinxes.[2] Here, we report a patient who presented with symptoms/signs of a Chiari type 1 malformation, but whose evaluation revealed a D4 intramedullary hemangioblastoma with a holocord syrinx.

CASE REPORT

A 21-year-old female presented with progressive numbness/tingling of the face, hands and feet, along with neck pain, gait instability, and hesitancy/urge incontinence of 6 months duration. On examination she exhibited weakness of both the upper and lower extremities along with the inability to feel hot/cold, and loss of pain sensation in both upper limbs and trunk. On examination, she exhibited a spastic quadriparesis (power 4/5) with atrophy in all four limbs. Reflexes were brisk bilaterally in all four extremities and she had bilateral Babinski responses. Dissociative sensory loss was present from T1 to T12 dermatomes bilaterally. Urodynamic study revealed detrusor hyperreflexia with sphincter dysynergia, requiring an indwelling Foley's catheter.

Diagnostic studies

The cervicothoracic magnetic resonance imaging (MRI) revealed a well-defined 14 × 15 × 25 mm lesion at the D4 level. It was isointense on T1, heterogeneously hyperintense T2, and intensely enhanced within the cord at the D4 level. Additionally, it showed cystic degeneration with collateral channels on the surface [Figure 1]. An accompanying

How to cite this article: Dutta G, Singh D, Singh H, Srivastava AK, Jagetia A, Agrawal A. Dorsal hemangioblastoma manifesting as holocord syringomyelia. Surg Neurol Int 2018;9:73.
http://surgicalneurologyint.com/Dorsal-hemangioblastoma-manifesting-as-holocord-syringomyelia/

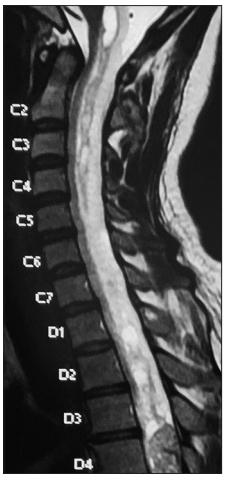

Figure 1: MRI cervico-dorsal region showing a well-defined intramedullary lesion at D4 with syrinx rostral to the lesion

holocord syrinx began at the cervicomedullary junction and extended all the way down to the termination of the spinal cord (T12 level) [Figure 2].

Surgery

The patient underwent a near total surgical excision of the lesion. The histology confirmed a WHO grade 1 hemangioblastoma. Thorough ophthalmological, abdominal, and brain evaluation documented no additional lesions that could be attributed to von Hippel–Lindau (VHL) syndrome.

Postoperative course

The patient ambulated by the end of the first postoperative week; she can now walk without support. Although urodynamic studies showed significant improvement, she still requires intermittent catheterization.

DISCUSSION

Syringomyelia is seen mainly in association with Chiari malformation, inflammatory pathologies, infarction, trauma, and intramedullary spinal cord tumors. Syrinxes with the latter are variously attributed to transudation of fluid from the tumor vessels, obstruction to cerebrospinal fluid (CSF), and/or obstruction of extracellular fluid flow. The subarachnoid and the extracellular spaces are a single fluid compartment with interrelated fluid flow; a block in one of the spaces may be reflected as increased flow in the other. Of interest, the higher the level of the tumor the greater the incidence of the accompanying syrinx.[3]

In this case, holocord syrinx formation occurred both rostral and caudal to the D4 hemangioblastoma. Here, we postulate the syrinx could be attributed both to obstruction of CSF flow and transudation of highly proteinous tumor fluid.

Although holocord syringomyelia has also been most frequently reported in conjunction with Chiari malformations,[1] a thorough evaluation of the whole spine should be performed in patients presenting with

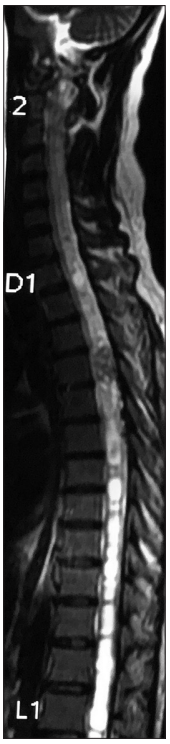

Figure 2: MRI whole spine showing associated holocord syrinx

holocord syringomyelia to rule out intramedullary tumors (e.g. as in this case, a hemangioblastoma).

CONCLUSION

Although most patients presenting with holocord symigomyelia, are thought to have Chiari malformations, others may have focal intramedullary spinal cord tumors. Therefore, it is critical to assess the entire spine to avoid missing these lesions and the opportunity to correctly treat these patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Kumar J, Kumar A, Gupta S. Neurological pictures. Chiari I malformation with holocord syrinx. J Neurol Neurosurg Psychiatry 2007;78:146
2. Pai SB, Krishna KN. Secondary holocord syringomyelia with spinal hemangioblastoma: A report of two cases. Neurol India 2003;51:67-8.
3. Wu TC, Guo WY, Lirng JF, Wong TT, Chang FC, Luo CB, *et al*. Spinal cord hemangioblastoma with extensive syringomyelia. J Chin Med Assoc 2005;68:40-4.