International Journal of Scientific Reports
Gollwitzer M et al. Int J Sci Rep. 2019 Mar;5(3):75-77
http://www.sci-rep.com
pISSN 2454-2156 | eISSN 2454-2164

DOI: http://dx.doi.org/10.18203/issn.2454-2156.IntJSciRep20190643

Case Report

Recurrent ominous spinal mass

Maria Gollwitzer*, Wolfgang Hasenauer, Anja Grimmer, Andreas Gruber

Department of Neurosurgery, Kepler University Hospital, Neuromed Campus, Linz, Upper Austria, Austria

Received: 15 January 2019
Revised: 05 February 2019
Accepted: 06 February 2019

***Correspondence:**
Dr. Maria Gollwitzer,
E-mail: mariagollwitzer@gmx.de

ABSTRACT

Hypertrophic pachymeningitis is a very rare form of diffuse inflammatory dural fibrosis. It is typically encountered intracranially rather than spinally and can have a progressive and recurrent course as encountered in our patient. The aetiology of this disorder is multifactorial. While acute management of hypertrophic pachymeningitis consists of immediate decompression in conjunction with steroid medication and treatment of the underlying disease, the management of chronic and recurrent disease is controversially debated in the current literature. A previously healthy 49-year-old woman presented with progressive back pain and acute onset of incomplete paraplegia. Spinal MRI showed a large ventral semicircular mass with intense contrast enhancement from Th3-8. Acute patient management consisted of immediate decompression and debulking the space-occupying lesion in conjunction with antibiotic and steroid therapy. Histopathologic examination of the surgical specimens revealed a chronic inflammatory process. The patient fortunately recovered from the severe paraparesis and upon discharged, spinal MRI disclosed a further regressing residual spinal mass. Over time, our patient suffered two times a relapse and was finally treated with Methotrexate and low-dose steroids. Under this medication she has made a nearly complete neurological recovery. This case report highlights a rare disease with challenging management, which should be considered diagnostically in patients with intraspinal space-occupying lesions. In our experience combination of Methotrexate and low-dose steroids might be an effective and safe treatment.

Keywords: Idiopathic meningitis, Pachymeningitis, Spine, Treatment

INTRODUCTION

Hypertrophic pachymeningitis is a very rare form of diffuse inflammatory dural fibrosis. It is typically encountered intracranially rather than spinally and can have a progressive and recurrent course as encountered in our patient.[1-3] The aetiology of this disorder is multifactorial, including infectious diseases (tuberculosis, lues, neurocysticercosis), rheumatic diseases (sarcoidosis), metabolic diseases, as well as vasculitis, autoimmune disorders, intrathecal injections or - as in our case - idiopathic.[4-7]

While acute management of hypertrophic pachy-meningitis consists of immediate decompression in conjunction with steroid medication and treatment of the underlying disease, the management of chronic and recurrent disease is controversially debated in the current literature.[8,9]

CASE REPORT

A previously healthy 49-year-old woman presented with progressive back pain and acute onset of incomplete paraplegia. On examination, her right leg was plegic and the strength in her left leg was severely reduced. The tendon reflexes of her lower extremities were pathologically increased and sensation was bilaterally impaired. Laboratory parameters and inflammatory markers were within normal ranges.

Spinal MRI showed a large ventral semicircular mass with intense contrast enhancement from Th3-8 (Figure 1). Acute patient management consisted of immediate decompression and debulking the space-occupying lesion in conjunction with antibiotic and steroid therapy. Histopathologic examination of the surgical specimens revealed a chronic inflammatory process and failed to demonstrate a tumorous lesion as initially suspected. Intraoperative bacteriologic examinations were negative as well. Since tumorous, infectious, rheumatic and autoimmune disease could be subsequently excluded, the aethiology of the lesion remained uncertain. Over time, the patient fortunately recovered from the severe paraparesis to a motor strength of 4/5. Upon discharged, spinal MRI disclosed a further regressing residual spinal mass (Figure 2).

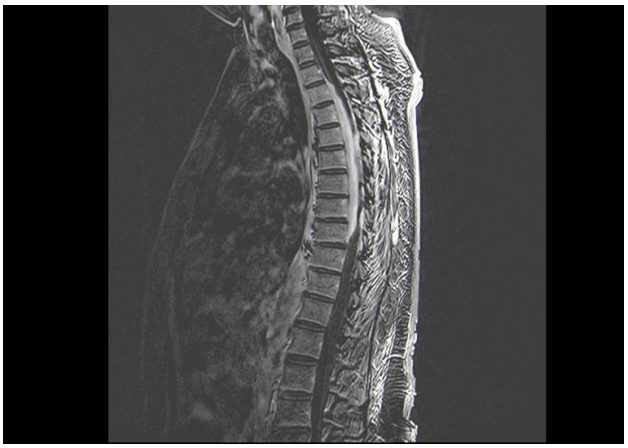

Figure 1: Preoperative intraspinal mass: Sagittal T1-weighted image with gadolinium reveal ventral semicircular intraspinal mass between T3 and T8 compressing the spinal cord.

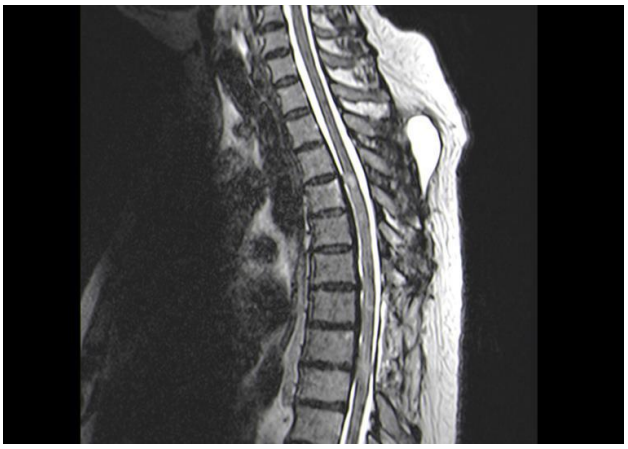

Figure 2: Postoperative control MRI of the spine: Sagittal T2-weighted images disclose a further regressing residual spinal mass after laminectomy.

Eight months later, our patient suffered a relaps with dysaesthesia and worsened paraparesis. The morphologic substrate for her clinical worsening was a recurrent spinal mass Th1–Th9. The patient was subsequently operated again. The spinal mass was debulked and the spinal cord decompressed.

Upon pathologic work–up of the surgical specimen, the tissue resected was grey, of solid-elastic consistency. Histopathologic examination revealed signs of chronic inflammation and granulation tissue formation but again failed to disclose a tumorous lesion. A second histological opinion was obtained, which basically came to the same results. Antibiotic and steroid therapy was implemented again. Over time, the residual spinal mass was again decreasing under medical treatment and the patient improved clinically again.

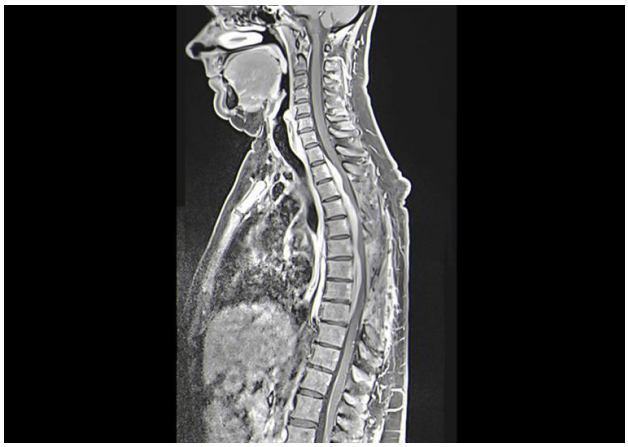

Figure 3: Relapse eight months after surgery: Sagittal T1-weighted image with gadolinium show recurrent spinal mass of the ventral dura from Th1–Th9.

Three months later, she suffered another relaps and was re-admitted with dysaesthesia, and worsened paraparesis (Figure 3). The recurrent and insufficiently understood clinical course of this patient led to an extended literature research on part of the treating physicians. We were able to find publications on hyperthrophic pachymeningitis.

DISCUSSION

In view of the paucity of published experience with this rare disease, management guidelines do not exist and published case reports describe different treatment algorithms on an individual basis. Recent case reports have described good therapeutic results after a regimen of low-dose steroid medication.[1] Other authors have recommend methotrexate as medical first line treatment.[2] Another case report, describing a case of recurrent idiopathic hypertrophic spinal pachymeningitis refractory to first line steroid treatment, describes the successful use of Rituximab as second-line treatment.[3] Moreover, strategies combining these three medical treatment approaches are also possible. In the present case, our patient is currently on methotrexate (15 mg/week) and a regimen of low-dose-steroids (prednisolone 10 mg/day) and has made a nearly complete neurological recovery under this medication.

CONCLUSION

This case report highlights a rare disease with challenging management, which should be considered diagnostically in patients with intraspinal space-occupying lesions. Management guidelines do not exist and published case reports describe different treatment algorithms on an individual basis. In our experience combination of Methotrexate and low-dose steroids might be an effective and safe treatment.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Qin LX, Wang CY, Hu ZP, Zeng LW, Tan LM, Zhang HN. Idiopathic hypertrophic spinal pachymeningitis: a case report and review of literature. Eur Spine J Suppl. 2015;4:636-43.
2. Park TJ, Seo WD, Kim SY, Cho JH, Kim DH, Kim KH. Effective Response of Methotrexate for Recurrent Idiopathic Hypertrophic Spinal Pachymeningitis. Korean J Spine. 2016;13:200-3.
3. Jang Y, Lee ST, Jung KH, Chu K, Lee SK. Rituximab Treatment for Idiopathic Hypertrophic Pachymeningitis. J Clin Neurol. 2017;13:155-61.
4. Kim JH, Park YM, Chin DK. Idiopathic hypertrophic spinal pachymeningitis: Report of two cases and Review of the Literature. J Korean Neurosurg Soc. 2011;50:392-5.
5. Bureta C, Abematsu M, Tominaga H, Saitoh Y, Kawamura I, Nagano S, et al. Hypertrophic spinal pachymeningitis associated with human T-cell lymphotrophic virus-1 infection and Sjogren's syndrome: A case report and brief literature review. Int J Surg Case Rep. 2018;45:22-8.
6. Kupersmith MJ, Martin V, Heller G, Shah A, Mitnick HJ. Idiopathic hypertrophic pachymeningitis. Neurology. 2014;62(5):686–94.
7. Sylaja PN, Cherian P, Das CK, Radhakrishnan V, Radhakrishnan K. Idiopathic hypertrophic cranial pachymeningitis. Neurol India. 2002;50(1):53–9.
8. Reining-Festa A, Flament H, Hoberstorfer M. Hypertrophe chronische Pachymeningitis. J. Neurol. Neurochir. Psychiatr. 2002;41-47.
9. Fukuda A, Punaro E, Rogerio F, de Souza Queiroz L, Reis F. Idiopathic hypertrophic pachymeningitis as a rare cause of cervical compressive myelopathy. J Craniovertebr Junction Spine. 2017;8(4):387-9.

Cite this article as: Gollwitzer M, Hasenauer W, Grimmer A, Gruber A. Recurrent ominous spinal mass. Int J Sci Rep 2019;5(3):75-7.