



HAL
open science

Clinical Image of a Spinal Ependymoma Discovered 8 Years after Initial Misdiagnosis as an Idiopathic Syringomyelia

Sam Ng, Nozar Aghakhani, Luc Bauchet

► **To cite this version:**

Sam Ng, Nozar Aghakhani, Luc Bauchet. Clinical Image of a Spinal Ependymoma Discovered 8 Years after Initial Misdiagnosis as an Idiopathic Syringomyelia. *World Neurosurgery*, 2021, 145, pp.338 - 339. 10.1016/j.wneu.2020.09.162 . hal-03493029

HAL Id: hal-03493029

<https://hal.science/hal-03493029>

Submitted on 7 Nov 2022

HAL is a multi-disciplinary open access archive for the deposit and dissemination of scientific research documents, whether they are published or not. The documents may come from teaching and research institutions in France or abroad, or from public or private research centers.

L'archive ouverte pluridisciplinaire **HAL**, est destinée au dépôt et à la diffusion de documents scientifiques de niveau recherche, publiés ou non, émanant des établissements d'enseignement et de recherche français ou étrangers, des laboratoires publics ou privés.



Distributed under a Creative Commons Attribution - NonCommercial 4.0 International License

1 Title:

2 Clinical image of a spinal ependymoma discovered 8 years after initial misdiagnosis as an
3 idiopathic syringomyelia

4
5 Authors:

6 Sam Ng,¹ MD, Nozar Aghakhani,^{2,3} MD, PhD, and Luc Bauchet,^{1,4} MD, PhD

7
8 Affiliations:

9
10 1- Department of Neurosurgery, Gui de Chauliac Hospital, Montpellier University Medical
11 Center, France

12 2- Department of Neurosurgery, Bicêtre University Hospital, Assistance Publique Hôpitaux
13 de Paris, Université Paris-Sud, France

14 3- Reference Center for Rare Diseases: Chiari-Syringomyelia and rare malformations of the
15 spine and spinal cord, Assistance Publique Hôpitaux de Paris, Université Paris-Sud,
16 France

17 4- INSERM U1051, Montpellier Neuroscience Institute, France

18
19 Corresponding Author's name and current institution:

20 Sam Ng, MD

21 Department of Neurosurgery, Hôpital Gui de Chauliac,

22 80 avenue Augustin Fliche, 34295 Montpellier, France

23 s-ng@chu-montpellier.fr

24

25 Key Words: Intramedullary spinal ependymoma, spinal cord cyst, syringomyelia, incidental
26 findings

27

28 Manuscript type: Clinical Image

29 Abstract word count: 114/150

30 Text word count: 316/350

31 Number of references: 5

32 Number of tables and/or figures: 1

33 Number of videos: none

34 Figure legends: 49/150

35

36

37

38

39 Abstract:

40 With the increasing frequency of neuroimaging, incidental intramedullary cavities are diagnosed
41 more frequently. We present a case of asymptomatic incidental intramedullary cervical cavity
42 diagnosed as an idiopathic syringomyelia as initial MRI showed an isolated cystic image without
43 contrasted component. The patient had no subsequent MRI follow-up but eventually showed
44 symptoms 8 years later. By this stage of the disease, the MRI appearance had changed, showing a
45 solid and enhanced component. The patient underwent surgical resection and histopathology
46 concluded a papillary ependymoma (grade 2). This case illustrates how asymptomatic
47 intramedullary cavities may hide an underlying tumoral process and why these cavities should
48 not be considered as idiopathic syringomyelia by default, except after prolonged MRI follow-up.

49

50 Main text:

51 In July 2011, a 57-year-old woman without symptoms was diagnosed with an incidental
52 intramedullary cavity after a mild cervical trauma. MRI, with and without gadolinium infusion,
53 demonstrated an isolated cervical cystic image. No solid and/or contrasted component was
54 observed (Figure 1A and 1B). Craniospinal explorations reported no other abnormalities (i.e. no
55 Chiari malformation, other cystic formation along the spinal cord or brain tumoral process). This
56 image was interpreted as an incidental idiopathic syringomyelia. The patient remained
57 asymptomatic and had clinical follow-up for 5 years, without MRI reevaluation.

58

59 Eight years later, she reported progressive dysesthesias in both hands. At this time, clinical
60 examination revealed 4 limb hyperreflexia and motor weakness. Craniospinal MRI with
61 Gadolinium enhancement showed a C2-C5 lesion (Figure 1C and 1D). Gross total resection was
62 achieved and neuropathology confirmed a papillary ependymoma, WHO grade 2.¹ At 6 months

63 postoperatively, the patient had recovered from transient neurological deficits thanks to intensive
64 rehabilitation. She received no adjuvant therapy but did have regular clinical and craniospinal
65 MRI follow-up.²

66
67 Intramedullary spinal ependymomas are frequently described with a cystic component,³ however,
68 it should be noted that recent surgical series have reported a solid component in all cases.⁴
69 Nevertheless, most studies have reported morphological aspects of symptomatic ependymomas
70 and the present case suggests that MRI findings of intramedullary ependymoma at a
71 presymptomatic stage may be associated with smaller volumes, inconsistent solid components
72 and non-enhanced lesions. Thus, the authors propose that asymptomatic intramedullary cavities
73 may hide an underlying tumoral process and should not be considered as idiopathic
74 syringomyelia by default, except after a prolonged MRI follow-up.⁵ Slow-growing tumors, such
75 as ependymal tumors, may become symptomatic many years after their incidental discovery and
76 give rise to significant therapeutic/surgical issues, while careful and repeated MRI assessment
77 may help to determine a better timing for surgical management. Therefore, even though they
78 have been incidentally diagnosed, long-term clinical and MRI follow-up of intramedullary
79 cavities is recommended.

80

81 Disclosures:

82
83 All authors (Sam Ng, Nozar Aghakhani, and Luc Bauchet) report no disclosures relevant to the
84 manuscript. No targeted funding reported.

85

86 Authors' contributions:

87 SN: Drafted the manuscript for intellectual content

88 NA: revised the manuscript for intellectual content

89 LB: Conceptualization of the study, acquisition of the data, revised the manuscript for intellectual
90 content

91

92 References

93

- 94 1. Louis DN, Perry A, Reifenberger G, et al. The 2016 World Health Organization
95 Classification of Tumors of the Central Nervous System: a summary. *Acta Neuropathol (Berl)*.
96 2016;131(6):803-820. doi:10.1007/s00401-016-1545-1
- 97
- 98 2. Rudà R, Reifenberger G, Frappaz D, et al. EANO guidelines for the diagnosis and
99 treatment of ependymal tumors. *Neuro-Oncol*. 2018;20(4):445-456. doi:10.1093/neuonc/nox166
- 100
- 101 3. Dauleac C, Messerer R, Obadia-Andre N, Afathi M, Barrey CY. Cysts associated with
102 intramedullary ependymomas of the spinal cord: clinical, MRI and oncological features. *J*
103 *Neurooncol*. 2019;144(2):385-391. doi:10.1007/s11060-019-03241-9
- 104
- 105 4. Aghakhani N, David P, Parker F, Lacroix C, Benoudiba F, Tadie M. Intramedullary spinal
106 ependymomas: analysis of a consecutive series of 82 adult cases with particular attention to
107 patients with no preoperative neurological deficit. *Neurosurgery*. 2008;62(6):1279-1285;
108 discussion 1285-1286. doi:10.1227/01.neu.0000333299.26566.15
- 109
- 110 5. Roy AK, Slimack NP, Ganju A. Idiopathic syringomyelia: retrospective case series,
111 comprehensive review, and update on management. *Neurosurg Focus*. 2011;31(6):E15.
112 doi:10.3171/2011.9.FOCUS11198

113

114

115

116 Figure 1

117 MRI of the cervical spine, T2 (left) and T1 postcontrast sequences (right).

118 (A) and (B) sequences were performed in 2011: an intramedullary cavity without solid
119 component was interpreted as an idiopathic syringomyelia.

120 (C) and (D) sequences demonstrate an intramedullary solid and cystic tumor with enhanced
121 components in segments C2-C5.

122

