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Case Report

Spinal clear cell meningioma without dural attachment: a case report and literature review ☆

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ABSTRACT

Clear cell meningiomas (CCM) are a very rare histologic subtype of meningioma usually affecting younger patients. The reported data on spinal CCM are extremely rare. Until today, only 89 cases have been reported. Furthermore, CCM without dural attachment is even rarer since only 19 cases have been reported in English literature. In this article, we present the twentieth case of a spinal CCM without dural attachment. Our patient was a 58-year-old female who was presented with pain in her lower back and bilateral sciatica for 6 months. Magnetic resonance imaging showed an intra-dural well-demarcated lesion at L3. Via a posterior approach, total resection was possible due to the lack of dural adhesion of the tumor. Histologic diagnosis was clear cell meningioma.

Introduction

Clear cell meningioma (CCM) is a rare subtype of meningioma. It accounts for less than 1% of all meningiomas [1]. Compared to ordinary meningiomas, CCMs have a higher recurrence rate (~50%) and a higher tendency to metastasize (4.1%) [2,3]. Therefore, the World Health Organization (WHO) classified them as grade II tumors.

Previously reported cases of CCMs have been mostly intracranial. Spinal CCMs are even rarer since only less than 100 cases of spinal CCMs have been reported until today [4]. Although it is known that meningiomas are attached to the dura mater, they seldom grow without dural attachment. Our literature review only found 19 previous cases of non-dura-based spinal CCM until today [4].

This article presents an unusual case of a spinal clear cell meningioma characterized by the absence of dural attachment.

Case report

A 58-year-old female was admitted with complaints of pain in her lower back and bilateral sciatica. Her symptoms started 6 months ago and worsened over time. Symptomatic treatment was not efficient. The neurologic examination did not reveal

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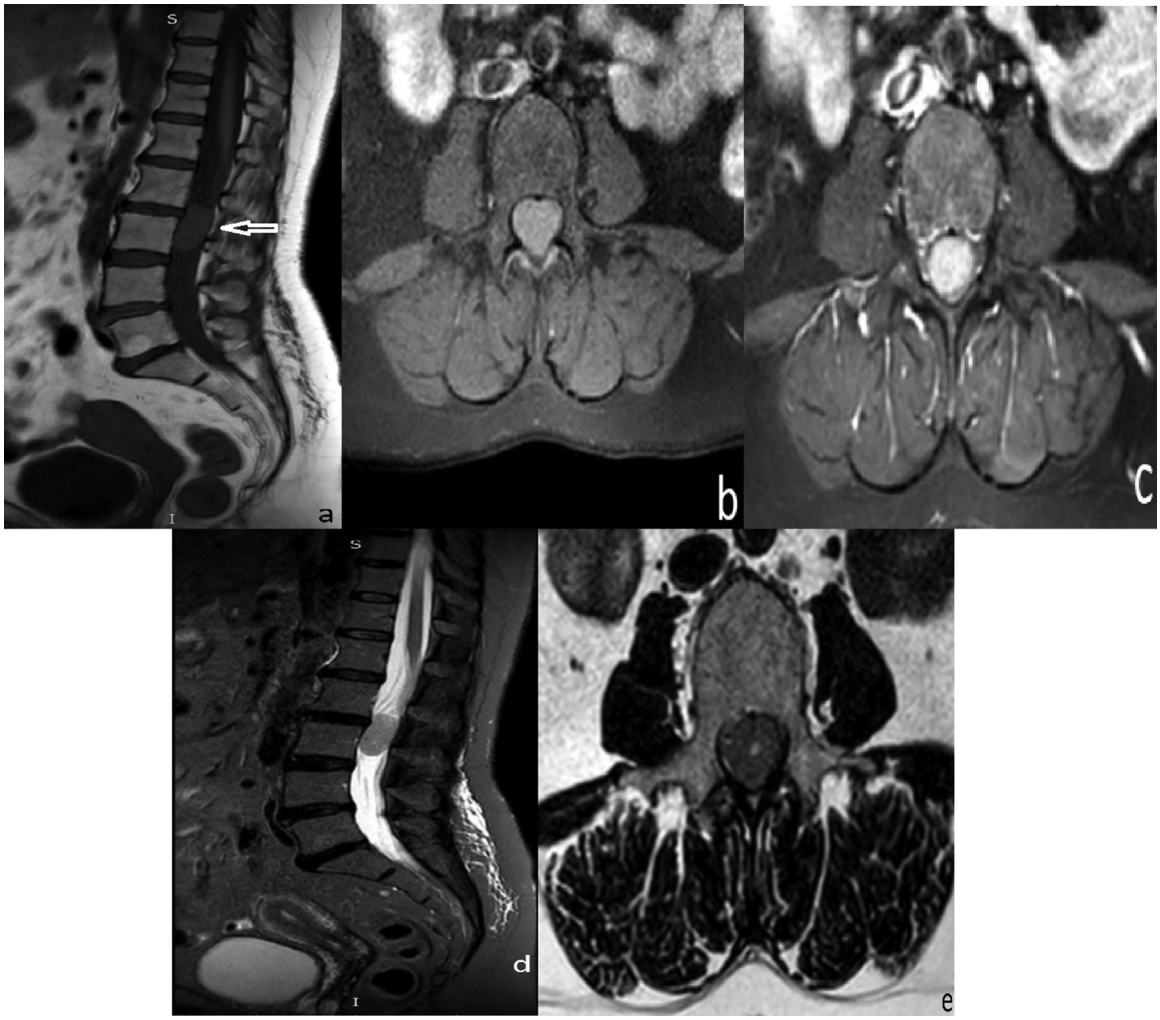

Fig. 1 – Intra-dural mass at L3 isointense on T1 on sagittal and axial section (A, B) and after injection of gadolinium (C) and hyperintense on T2 on sagittal and axial section (D, E).

any specific findings other than a classical lumbar spinal syndrome. There was no motor or sensitive deficit of her lower limbs and no abnormalities of the anal tone.

Magnetic resonance imaging (MRI) showed an intra-dural lesion at L3 measuring 31 mm of height. The mass was well-demarcated and had an isointense signal on both T1-and T2-weighted images with intense and homogeneous gadolinium enhancement (Fig. 1). There was no foraminal extension and no associated bone destruction.

The operation started with an L2-L3 laminectomy. The intra-dural exposure revealed an elliptic, encapsulated yellow mass of firm consistency. The tumor was found to be draped by and adhered to the nerve roots without dural attachment. The tumor's major arterial blood supply seems to be coming from a lumbar radicular artery. After an easy dissection, we managed to perform a total resection.

On microscopic examination (Fig. 2), the tumor was composed of sheets of polygonal cells with clear glycogen-rich cytoplasm and monomorphic, non-mitotic nuclei. There was prominent perivascular and interstitial collagen. Whorl formation and psammoma bodies were absent. The pathological diagnosis was a CCM (WHO II).

Patient's recovery course after the operation was uneventful. All of the patient's preoperative symptoms improved after surgery. She was able to get down the next day.

Discussion

First reported by Manivel and Sung [5] in 1990, CCM is one of the rarest subtypes of meningiomas, representing 0.2%-0.8% [6–8]. Zorludemir et al [6]. and Oviedo et al [7]. found that this type of meningiomas had a higher local recurrence rate and a more aggressive clinical course compared to ordinary meningiomas. For these reasons, the World Health Organization (WHO) classified CCMs as grade II tumors in 2016 [9].

Although around 300 cases of CCMs have been reported in the English language studies, most have been intracranial cases. The reported data on spinal CCMs are extremely rare. Our literature review only found 88 cases of spinal CCMs reported since 1996, 18 of them had no dural attachment (Table).

However, it is noteworthy that the proportion of spinal to total meningiomas is higher for CCMs (45%) than for other

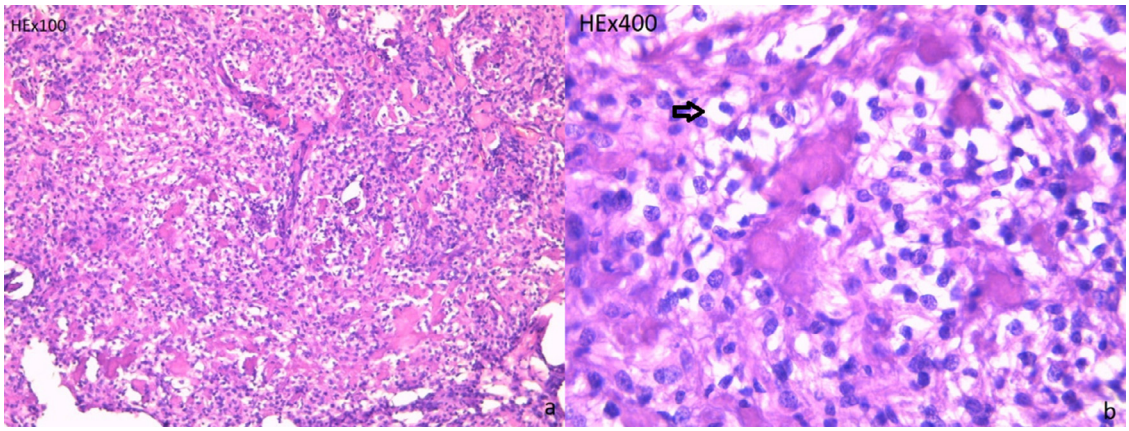

Fig. 2 – Pathological features of spinal clear cell meningioma: Tumor cells in classical CCM area with distinct cellular outlines and abundant clear cytoplasm (Black arrow) with no mitosis and no necrosis area (A: H&E staining with original magnifications x 100; B: H&E staining with x400).

Table 1 – Information summary of the reported intra-spinal CCMs without dural attachment in English literature.

Reference	Report year	Age (years)	Sex	Location	Dural attachment	Surgical treatment	Adjuvant therapy	Time of recurrence (months)
Present case	2021	58	F	L3	No	GTR	None	No
Inoue et al. [24]	2018	5	M	L5-S1	No	GTR	None	No
Kawasaki et al. [25]	2018	8	F	L2	No	GTR	None	No
Li et al. [1]	2016	7	F	L2-L4	No	GTR	None	No
Zhang et al. [26]	2013	26	F	Temporal lobe + T12-L1	No	GTR	None	No
Kobayashi et al. [27]	2013	43	M	L2-L3	No	GTR	None	No
Ko et al. [28]	2011	34	F	L2-L3	No	GTR	None	No
Park et al. [10]	2005	65	F	T9-T10	No	GTR	None	No
Epstein et al. [29]	2005	41	F	L3-L4	No	GTR	None	No
Oviedo et al.[15]	2005	7	M	L2-L3	No	GTR	None	No
Chen et al. [18]	2004	41	F	L4-L5	No	GTR	None	No
Payano et al. [14]	2004	24	M	L3-L4	No	GTR	None	No
		19	F	L3	No	GTR	None	No
Carrà et al. [30]	2003	1.8	M	T11-L4	No	GTR	None	60
Jallo et al. [2]	2001	8	F	L3-L4	No	GTR	None	6
		1.8	F	C3-C5	No	STR	None	2.3
Dubois et al. [31]	1998	10	F	L1-L4	No	GTR	None	6
Matsui et al. [32]	1998	9	F	L2	No	GTR	None	4
Holtzman et al. [33]	1996	32	M	L3-L4	No	GTR	None	No
Zorludemir et al. [6]	1996	17	F	L4-L5	No	GTR	None	No

F: Female, M: Male, NA: Not Available, GTR: Gross Total Resection, STR: Subtotal Resection, RT: Radiotherapy

meningiomas (the highest being 20% of the reported cases) [10].

In 2019, Zhang et al [4]. found that CCMs mostly attack young patients with a mean age of 24 years old. Amazingly, 36 (42.9%) suffered from spinal CCM at an age of under 18 years. As it is for other meningiomas, CCMs have a slight female predominance with a female to male ratio at 1.7:1 (53 vs 31).

In our review, 37 (42%) patients were under 18 years old and the mean age at resection was 24.5 years 57 (64%) patients were female, 32 (36%) were male and the female-to-male ratio was 1.78:1.

Remarkably different from conventional meningiomas, for CCMs, the most affected location was the lumbar region (66.7%), rather than the thoracic region [4], which is the most affected location for ordinary spinal meningiomas [11–13].

Histologically, clear cell meningioma contains sheets of clear, glycogen-rich (Periodic acid-Schiff positive, diastase-labile), polygonal cells forming only a few vague whorls [6]. Its abundant glycogen is the reason why it's called "clear cell" meningiomas [14]. These cells are almost immunoreactive to epithelial membrane antigen (EMA) and vimentin, while negative to GFAP, S-100, CK, and SMA, with their ki67 index varying from negative to 40%. The overexpression of EGFR, PDGF-receptor, and VEGF in CCMs promotes meningioma cell proliferation, a key process in meningioma angiogenesis, the formation of peritumoral edema, as well as tumor aggression [15,16].

The imaging features of clear cell meningiomas are very similar to those of ordinary meningiomas [16]. MRI of CCMs often reveals an intradural-extramedullary, well-demarcated, homogeneously enhanced mass. It is isointense on both T1- and T2-weighted images and demonstrates fairly homogeneous enhancement after the injection of gadolinium [8,17]. Some cases showed foraminal extension and lacking dural attachment, just like schwannomas [18]. Even an intramedullary clear cell meningioma has been reported [10]. Because of radiologic and gross morphologic similarities between the spinal meningiomas and other intradural extramedullary spinal tumors just as schwannomas, neurofibromas, and ependymomas, an accurate histological diagnosis is important. It is important to mention that some CCMs are not attached to the dura but the neural sheath, as it is in our case. Thirteen such non-dura-based intra-spinal CCM have been reported to date (Table 1).

As an intra-spinal lesion, bone destruction of CCM is rare, reported only 3 times: 2 cases reported by Jian Yang et al. in 2018 [19] and a case reported by Alsadiq et al. in 2021 [20]. The rarity of bone destruction may be explained by the fact that neurological symptoms usually appear before bone involvement due to the limited space of the spine canal, especially in the lumbosacral region.

The potential aggressiveness and recurrence are the main reason for the very challenging management of this tumor [21]. Up to date, for spinal CCMs, total resection should be considered the optimal treatment. In our review of literature, 79 (89.77%) cases received gross total resection (GTR) as the first treatment. However, 20 cases of spinal CCMs (22.72%) still experienced one or more recurrences until the last follow-up.

The value of postoperative radiotherapy has been increasingly affirmed and highlighted for the treatment of intracranial CCMs [3,22]. However, because of its rarity, the efficacy of postoperative radiotherapy for spinal CCM could not be affirmatively concluded and requires further investigation. Tao et al [22]. reported that radiotherapy should not be performed immediately after the first operation for spinal CCMs, because the recurrence rate is lower than that for intracranial CCM, which can be explained by the higher rate of GTR in spinal CCMs.

According to the reported data, it is evident that spinal CCMs are more aggressive, with a much greater progression rate (38.0%) after initial treatment compared with ordinary spinal meningiomas (~3%) [13]. Jiu Hong Li et al. showed in their study that younger patients had better evolution after treatment (54.2%) compared with older ones (23.1%) [23].

Conclusion

Spinal CCM, a rare subtype of meningiomas is an aggressive neoplasm with a high rate of local recurrence. They have a predilection to affect younger patients and the lumbar spine.

GTR is recommended in treating spinal CCM whenever possible. However, radiotherapy could be considered for patients who have undergone STR or younger patients, regardless of the extent of resection. Younger patient age was associated with a significantly shorter progression-free survival, and patients who undergo STR might experience a greater.

Informed consent

The patient was informed of this manuscript and gave us his consent.

REFERENCES

[1] Li P, Yang Z, Wang Z, Zhou Q, Li S, Wang X, et al. Clinical features of clear cell meningioma: a retrospective study of 36 cases among 10,529 patients in a single institution. Acta Neurochir (Wien) 2016;158(1):67–76.

[2] Jallo GI, Kothbauer KF, Silvera VM, Epstein FJ. Intraspinal clear cell meningioma: diagnosis and management: report of two cases. Neurosurgery 2001;48(1):218–21 discussion 221-222.

[3] Zhang H, Ma L, Wang Y-B, Shu C, Kuang W, Huang Y-A, et al. Intracranial clear cell meningiomas: study on clinical features and predictors of recurrence. World Neurosurg 2017;97:693–700 e11.

[4] Zhang H, Ma L, Shu C, Dong L-Q, Ma Y-Q, Zhou Y. Spinal clear cell meningiomas: clinical features and factors predicting recurrence. World Neurosurg 2019;134:e1062–76.

[5] Manivel JC, Sung JH. Pathology of meningiomas. Pathol Annu 1990:159–92 25 Pt 2.

[6] Zorludemir S, Scheithauer BW, Hirose T, Van Houten C, Miller G, Meyer FB. Clear cell meningioma. A clinicopathologic study of a potentially aggressive variant of meningioma. Am J Surg Pathol. 1995;19(5):493–505.

[7] Ohba S, Sasaki H, Kimura T, Ikeda E, Kawase T. Clear cell meningiomas: three case reports with genetic characterization and review of the literature. Neurosurgery 2010;67(3):E870–1 discussion E871.

[8] Lee W, Chang KH, Choe G, Chi JG, Chung CK, Kim IH, et al. MR imaging features of clear-cell meningioma with diffuse leptomeningeal seeding. AJNR Am J Neuroradiol 2000;21(1):130–2.

[9] Louis DN, Perry A, Reifenberger G, von Deimling A, Figarella-Branger D, Cavenee WK, et al. The 2016 World Health Organization classification of tumors of the central nervous system: a summary. Acta Neuropathol (Berl) 2016;131(6):803–20.

[10] Park S-H, Hwang S-K, Park Y-M. Intramedullary clear cell meningioma. Acta Neurochir (Wien) 2006;148(4):463–6.

[11] Solero CL, Fornari M, Giombini S, Lasio G, Oliveri G, Cimino C, et al. Spinal meningiomas: review of 174 operated cases. Neurosurgery 1989;25(2):153–60.

[12] Spinal meningiomas surgical management and outcome. - PubMed - NCBI [Internet]. [cité 16 juill; 2019.]Disponible sur: https://www.ncbi.nlm.nih.gov/pubmed/15669787.

[13] Raco A, Pesce A, Toccaceli G, Domenicucci M, Miscusi M, Delfini R. Factors leading to a poor functional outcome in spinal meningioma surgery: remarks on 173 cases. Neurosurgery 2017;80(4):602–9.

[14] Payano M, Kondo Y, Kashima K, Daa T, Yatsuka T, Kida H, et al. Two cases of nondura-based clear cell meningioma of the cauda equina. APMIS Acta Pathol Microbiol Immunol Scand 2004;112(2):141–7.

[15] Oviedo A, Pang D, Zovickian J, Smith M. Clear cell meningioma: case report and review of the literature. Pediatr

Dev Pathol Off J Soc Pediatr Pathol Paediatr Pathol Soc 2005;8(3):386–90.
[16] Dhall SS, Tumialán LM, Brat DJ, Barrow DL. Spinal intradural clear cell meningioma following resection of a suprasellar clear cell meningioma. Case report and recommendations for management. J Neurosurg 2005;103(3):559–63.
[17] Wang X-Q, Huang M-Z, Zhang H, Sun F-B, Tao B-B, Feng B-H, et al. Clear cell meningioma: clinical features, CT, and MR imaging findings in 23 patients. J Comput Assist Tomogr 2014;38(2):200–8.
[18] Chen M-H, Chen S-J, Lin S-M, Chen M-H. A lumbar clear cell meningioma with foraminal extension in a renal transplant recipient. J Clin Neurosci Off J Neurosurg Soc Australas 2004;11(6):665–7.
[19] Two novel cases of spinal clear cell meningioma with vertebra involvement. - PubMed - NCBI [Internet]; 2019. [cité 16 juill]. Disponible sur: https://www.ncbi.nlm.nih.gov/pubmed/30545721.
[20] Alsadiq MN, Albarbari ZS, Alshakhs F, Alduayji MA, Al-Umran S, Alenzi A. Spinal clear cell meningioma: atypical clinical and radiological manifestations. Case Rep Surg 2021:9998399.
[21] Liu P-I, Liu G-C, Tsai K-B, Lin C-L, Hsu J-S. Intraspinal clear-cell meningioma: case report and review of literature. Surg Neurol 2005;63(3):285–8 discussion 288-289.
[22] Tao X, Dong J, Hou Z, Hao S, Zhang J, Wu Z, et al. Clinical features, treatment, and prognostic factors of 56 intracranial and intraspinal clear cell meningiomas. World Neurosurg 2018;111:e880–7.
[23] Li J, Zhang S, Wang Q, Cheng J, Deng X, Wang Y, et al. Spinal clear cell meningioma: clinical study with long-term follow-up in 12 patients. World Neurosurg 2019;122:e415–26.
[24] Inoue T, Shitara S, Ozeki M, Nozawa A, Fukao T, Fukushima T. Hereditary clear cell meningiomas in a single family: three-cases report. Acta Neurochir (Wien) 2018;160(12):2321–5.
[25] Kawasaki Y, Uchida S, Onishi K, Okanari K, Fujiki M. Pediatric nondura-based clear cell meningioma of the cauda equina: case report and review of literature. Br J Neurosurg 2018:1–4.
[26] Zhang J, Shrestha R, Li J, Shu J. An intracranial and intraspinal clear cell meningioma. Clin Neurol Neurosurg 2013;115(3):371–4.
[27] Kobayashi Y, Nakamura M, Tsuji O, Iwanami A, Ishii K, Watanabe K, et al. Nondura-based clear cell meningioma of the cauda equina in an adult. J Orthop Sci Off J Jpn Orthop Assoc 2013;18(5):861–5.
[28] Ko JK, Choi BK, Cho WH, Choi CH. Non-dura based intaspinal clear cell meningioma. J Korean Neurosurg Soc 2011;49(1):71–4.
[29] Epstein NE, Drexler S, Schneider J. Clear cell meningioma of the cauda equina in an adult: case report and literature review. J Spinal Disord Tech 2005;18(6):539–43.
[30] Carrà S, Drigo P, Gardiman M, Perilongo G, Rigobello L. Clear cell meningioma in a 22-month-old male: update after five years. Pediatr Neurosurg 2003;38(3):162–3.
[31] Dubois A, Sévely A, Boetto S, Delisle MB, Manelfe C. Clear-cell meningioma of the cauda equina. Neuroradiology 1998;40(11):743–7.
[32] Matsui H, Kanamori M, Abe Y, Sakai T, Wakaki K. Multifocal clear cell meningioma in the spine: a case report. Neurosurg Rev 1998;21(2–3):171–3.
[33] Holtzman RN, Jormark SC. Nondural-based lumbar clear cell meningioma. Case report. J Neurosurg 1996;84(2):264–6.