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Recurrent Clear Cell Meningioma of Cauda Equina in a Middle-Aged Gentleman

***Abstract* —** Clear cell meningioma of the cauda equina is an infrequent occurrence. It falls under the Central Nervous System WHO Grade 2, 5th Edition WHO Classification. This type of meningioma is known for its aggressive nature, frequent recurrence, and instances of leptomeningeal metastasis. The radiological findings resemble typical meningioma, but specific indicators of clear cell meningioma include leptomeningeal enhancement, although this feature is not exclusive to it. Therefore, the diagnosis primarily relies on histopathology. Here, we present a case of a 58-year-old local male with a history of laminectomy and excision of clear cell meningioma of the cauda equina.

***Keywords* —** Clear cell meningioma, cauda equina, laminectomy, meningioma

1 INTRODUCTION

Spinal meningiomas account for 12% of all meningiomas [1]. However, only 4% of spinal meningiomas are found in the lumbar regions, with a greater prevalence in the thoracic and cervical regions [2]. The clear cell meningioma (CCM) subtype, characterized by round or polygonal cells with clear and glycogen-rich cytoplasm, is a scarce variant among all meningiomas [3]. This particular subtype primarily affects children and young adults [3]. The World Health Organization (WHO) has classified CCM as a Grade 2 tumor of the central nervous system (CNS), and it is known for its aggressive nature, high recurrence rate, and instances of leptomeningeal metastasis [3]. In our case, the patient was diagnosed in middle age with clear cell meningioma, which is a rare type of meningioma and the disease at the atypical location, cauda equina.

2 CASE REPORT

A 58-year-old local male with no known medical illness initially presented to a nearby referral center with progressive left foot weakness and numbness associated with lower back pain for six months. There was no bowel or bladder involvement, symptoms, or deficits over the right lower limb. A magnetic resonance imaging (MRI) of the lumbosacral spine revealed a well-defined extramedullary elongated lesion over the superior border of L4 till the superior border of S1 measuring approximately 6 cm in craniocaudal, which was seen arising from the cauda equina, favoring for a myxopapillary ependymoma (Figure 1).

The L4-S1 laminectomy and tumor excision were performed. The intraoperative finding was fibrous tissue measuring 5.5cm in length arising from filum terminale extending to the left neural foramen, which was completely excised. Histopathological analysis revealed a pattern-less arrangement of clear cells interspersed with distinct cell borders with prominent perivascular and interstitial thick collagen.

The nuclei were small and round, with no mitotic activity. Immunohistochemically, the stain was positive for vimentin, Epithelial membrane antigen (EMA), and Periodic acid-Schiff (PAS), whereas negative for PAS D, CD10, Glial fibrillary acidic protein (GFAP), Cytokeratin (PanCK), and S100. The final impression was CNS WHO Grade 2 CCM **(**Figure 2).

Post-operatively, his symptoms improved until six months later, when he developed a recurrence of back pain associated with worsening numbness over the left foot and leg. He also started to have numbness of the right foot. On examination, the straight leg raising test was positive bilaterally. The motor powers were normal except for left toe flexion of Medical Research Council (4) grade 4/5. Sensation over the right side was reduced from L5 and below bilaterally.

A follow-up lumbosacral MRI revealed the previous laminectomy of L4-S1. A lobulated intradural lesion at level L5 till S1 measuring approximately 3.7x2.6cm showed a similar imaging appearance as per the initial MRI. The mass was also extended into the left neural foramen (Figure 3). The patient was subjected to a second surgery. Intraoperatively, there was a well-defined brownish nonvascular intradural extramedullary tumor dorsal to the spinal cord, measuring 3.5cm in length, extending into the left neural foramen, for which near total excision was performed. The histology remained unchanged. Postoperatively, the back pain resolved, and he was left with residual numbness over the left L5 dermatome. A follow-up MRI at one month postoperatively revealed a small residual tumor with L5 exiting nerve root impingement. After a multidisciplinary discussion, the patient was planned for radiotherapy for treatment of the residual lesion.

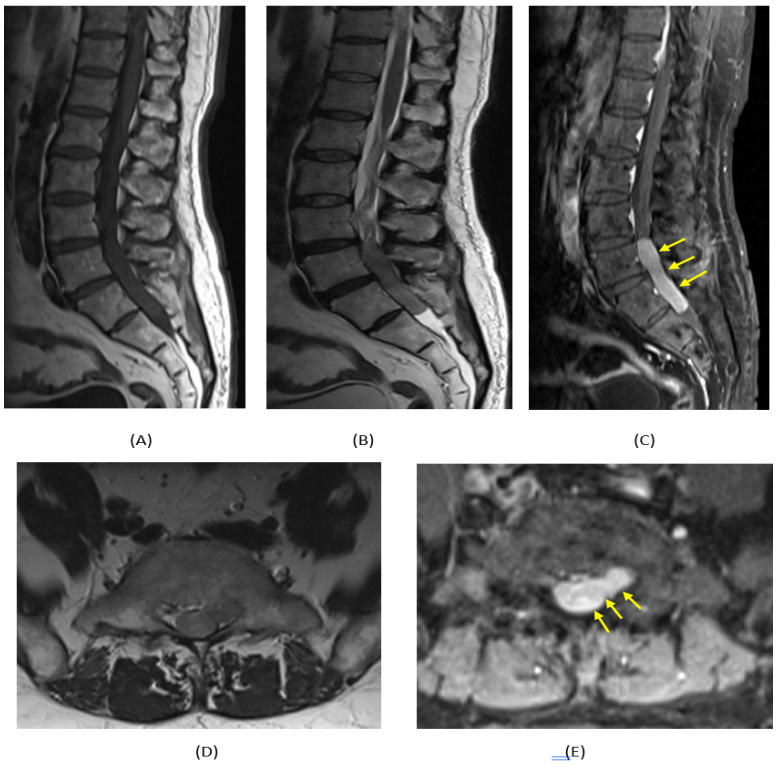

Figure 1. Magnetic resonance imaging of the lumbosacral (A) Sagittal T1WI, (B) Sagittal T2WI, (C) Sagittal Post-contrast T1, (D) Axial T2 and (E) Axial Post-contrast T1, shows well-defined intradural extramedullary elongated lesion within the spinal canal which is located over the superior border of L4 till the border of S1(arrow). The mass is iso- to hypointense on T1, iso- to hyperintense on T2 and homogenously enhances following the contrast media administration. It has extension into the left neural foramen

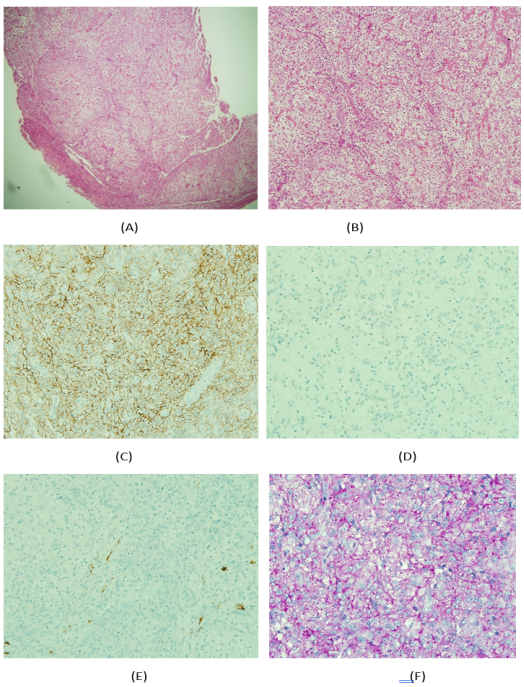

Figure 2. (A) Haematoxylin and Eosin (H&E) stain (4x magnification), (B) (H&E) (10x magnification), (C) Epithelial membrane antigen (EMA) (X20) positivity, (D) Glial fibrillary acidic protein (GFAP) (X40) negativity, (E) S100 stain (X20) negativity and (F) Periodic acid-Schiff (PAS) stain (X40) positivity revealed a pattern-less arrangement of clear cells interspersed with distinct cell borders with prominent perivascular and interstitial thick collagen, with small, round to oval nucleus with no mitotic activity

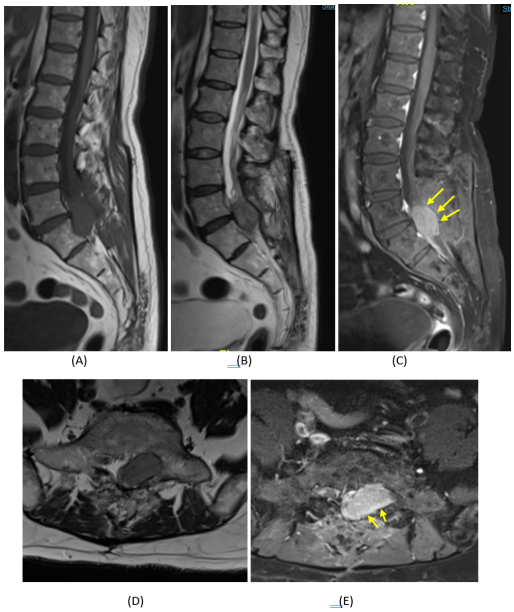

Figure 3. Magnetic resonance imaging of the lumbosacral (A) Sagittal T1WI, (B) Sagittal T2WI, (C) Sagittal Post-contrast T1WI, (D) Axial T2WI and (E) Axial Post-contrast T1WI show evidence of the previous laminectomy at L4-S1. There is a lobulated intradural lesion within the spinal canal which is located at level L5 till S1(arrow). The mass is isointense on T1, iso- to hyperintense on T2, and homogenously enhances following the contrast media administration. There is also an extension of the mass into the left neural foramen

3 DISCUSSION

Spinal meningiomas constitute 12% of all meningiomas [3] and approximately 25% of all primary spinal cord tumours, with a male-to-female ratio of 1:4 [4]. The majority of spinal meningiomas are located within the intradural space, while cases with extradural extension are uncommon. Thoracic spinal region is the most common site (80%), followed by the cervical region (16%), whereas the lumbar and cauda equina regions are the least common (4%) [2]. CCM is an exceedingly rare subtype of meningioma characterized by sheets of round or polygonal cells with clear glycogen-rich cytoplasm, along with prominent perivascular and interstitial collagen [3]. CCM predominantly affects children and young adults [3]. It is associated with an aggressive clinical course and occasional metastasis via the cerebrospinal fluid [3,5]. The existing literature on CCM is limited to case reports and small case series, and due to its rarity, the epidemiological and clinical characteristics of CCMs have not been fully elucidated.

The first reported case of CCM was documented by Harkin et al. in 1998 [6]. Electron microscopic analysis of the tumour revealed broad zones with large amianthoid collagen fibres. Initially classified as Grade 1, CCM's classification was later revised to Grade 2 due to its high recurrence rate and aggressive clinical behaviour [5,7]. According to available literature [8,9], CCM is touted as among the rarest subtypes, accounting for approximately 0.2–0.8% of all meningiomas. Limited genetic aberrations have been identified, including mutations in the neurofibromatosis gene (NF-2) [9,10] and SMARCE1 [11-13]. However, the precise aetiology and defining genomic mutations underlying CCM are still unclear.

Zhang et al. conducted separate reviews of reported cases of intracranial CCMs [14] and spinal CCMs [15], revealing a significant female predominance in spinal CCMs. Furthermore, Louis et al. [16] reported that CCMs are more commonly observed in younger patients, including children and young adults [4]. In Zhang's review, it was found that 42.9% of patients with spinal CCMs were younger than 18 years old [17], and the mean age at the time of surgery was 24 years old for spinal CCMs and 32 years old for intracranial CCMs [14,15]. However, it is worth noting that some reports did not show a pronounced preference for the younger population [17].

CCMs are predominantly reported to occur at the cerebellopontine angle, supratentorial and spine, especially in the cauda equina region [3]. Radiologically, CCMs typically exhibit similar characteristics to ordinary meningiomas. They appear isointense on T1-weighted imaging and iso- or hyperintense on T2-weighted imaging, with intense and homogeneous enhancement following the administration of gadolinium [18]. There was also one case report of solid cystic appearance of clear cell meningioma within cerebellopontine angle [19] and another reported case of multifocal intradural extramedullary lesions within thoracic and lumbar spine [20]. Clues suggestive of CCM include leptomeningeal enhancement, even though leptomeningeal enhancement has also been reported in other types of ordinary meningioma [21,22].

4 CONCLUSION

CCM is a histologically rare subtype of meningioma that has a higher recurrence rate, compared to typical meningiomas. Recurrence occurs as either local recurrence or as CSF seeding. CCM must hence be followed up carefully because of its aggressive behaviour, even when benign histologic features are reported. As seen in our reported case, it can affect even middle-aged gentleman for which recurrence can occur as early as 6 months postoperatively despite complete excision.

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