Case 5599

Clear cell meningioma of the conus medullaris: a rare tumor in an uncommon location
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Patient: 13 years, female

Clinical History:

We report a case of a 13-year-old female who presented with low back pain and weakness in both lower limbs. MRI scans revealed an intradural minimally enhancing lesion extending from L1-3. On surgical excision, the tumor had no apparent attachment to the dura. Histopathology a clear cell meningioma.

Imaging Findings:

A 13-year-old female presented to us with complaints of low back pain since one year. There was history of gradual onset of bilateral lower limb weakness since six months. Her power was grade three in the left lower limb & grade four in the right lower limb. The Extensor Hallucis Longus was found to be weak on the left side. There was decreased perception of sensations in L2 to S2 Dermatomes. Straight leg raising test was within normal range. M.R.I. Scan of the dorso-lumbar spine revealed an intradural extramedullary lesion extending from the L1 to L3 that is homogenous iso to hypointense on T1 weighted images and inhomogenous iso-hypointense on T2 weighted images. There was minimal inhomogenous enhancement on gadolinium contrast. The patient underwent a D12 to L4 laminectomy with excision of the intradural lesion. The lesion was yellow, firm, well capsulated, vascular 6.3x1.5x1.5 cm in size, at the level of L1 to L3 more towards the right. Another grayish red vascular firm, well capsulated tumour 4.5x1.5x1.5 cm was found at the level of L2 & L3. Both these tumours were found to be entrapped in the nerve roots. There was no apparent attachment to the dura mater. A histopathological diagnosis of a clear cell meningioma as reached. Post operatively the patient showed good recovery with full power in both the lower limbs. Sensations gradually returned to normal and the patient was discharged on the 10th day. Follow up examination after six months revealed no neurological deficit and MRI Scan was normal.
Discussion:

Tumors of the cauda equina and conus medullaris are uncommon. Meningiomas in the lumbosacral region are very rare. They represent only 2% of all the spinal cord meningiomas. Thoracic region is involved far more often followed by the cervical region. The new WHO classification of tumors of the central nervous system divides meningiomas into 11 histological variants including the recently described clear cell meningiomas. Clear cell meningiomas are one of the rarest histological forms representing 0.2% of all meningiomas. Fewer than 20 cases of clear cell meningiomas have been reported in literature, the only series being by Zorludemir et al. Unlike other meningiomas, which are very uncommon in the lumbosacral region, clear cell meningiomas may have a predilection for the lumbar region and patients often present with the cauda equina syndrome of back pain and radiculopathy. Zorludemir et al reviewed 14 cases of clear cell meningiomas of which six were lumbar and three at the cerebellopontine angle. Intraspinal meningiomas are rare in the young age group particularly children. They also occur more commonly in females. However the clear cell meningiomas show no predilection for any sex, children being commonly affected. Clear cell meningiomas tend to be aggressive tumors with frequent recurrence post operatively. They may also spread locally or disseminate within the nervous system. The younger the patient, higher the risk of recurrence. Our case was a young female with a conus-cauda meningioma, which on ultra structural studies was found to be a clear cell meningioma. Thus although this is an uncommon age and location for a meningioma, the age and location are typical for the rare morphologic variant of the clear cell meningiomas. Another atypical feature of our case was the tumor's non-attachment to the dura. Nondural attachment of intradural meningiomas is quite uncommon. All the cases reported so far, as in our case, have involved tumor located in the lumbosacral region. Only two cases of a non-dural based clear cell meningioma have been described, both of which were in the lumbar region. Contrast enhanced MRI remains the imaging modality of choice. Treatment of intradural spinal meningiomas is surgical with excision of the tumor and excision or coagulation of the dural attachment. However the importance of the clear cell variant is in its tendency to recur. Therefore complete excision is necessary. Some authors recommend postoperative adjuvant radiotherapy. Routine follow up with serial imaging studies every 3 months may also be necessary. We report this rare morphologic variant, typically occurring in a lumbar location, which is rare for meningiomas and also for the uncommon occurrence of its non-attachment to the dura. Thus the diagnosis of a clear cell meningioma should be kept in mind for intradural spinal tumors occurring in the conus-cauda region especially in the young. The aggressive nature & possibility of recurrence of this tumor also calls for meticulous and complete excision with regular follow up.

Differential Diagnosis List: Intradural Extramedullary Clear Cell Meningioma extending from L1 to L3

Final Diagnosis: Intradural Extramedullary Clear Cell Meningioma extending from L1 to L3

References:


Zorludemir S, Scheithauer BW, Hirose T, Van Houten C, Miller G, Meyer FB. Clear cell meningioma. A
Description: Figure 1: MRI Sagittal images Origin:
Figure 2

Description: Figure 2: Axial Images

Origin:
Figure 3

Description: Figure 3: Hematoxylin Eosin Stain

Origin:
Figure 4

Description: Figure 4: PAS stain
Origin: