Lumbar spinal cystic schwannoma

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Though cystic changes in schwannomas are well described, predominantly cystic schwannomas are uncommon lesions and form a different spectrum of conditions as compared with the commonly seen intradural extramedullary spinal tumors.

Savardekar et al. present a case series of six patients with spinal intradural extramedullary cystic schwannomas. Two patients had uniloculated cystic schwannomas, two patients had multi-loculated cystic lesions with thick walls and intralesional septations, and two patients had giant cystic schwannomas, one of which had an extradural extension. We report two cases in which preoperative radiological dilemma was encountered and discuss the differential diagnoses of this uncommon entity.

Cystic spinal schwannomas may be confused with other cystic lesions in the spine, differentiating them preoperatively is important and in this regard, contrast-enhanced magnetic resonance imaging plays a vital role. Frozen section histopathology should be used to identify them at surgery. It is important to detect these lesions at surgery, as total excision is possible and almost always results in good long-term neurological outcome [1)].

Case reports

A rare case of a long completely cystic sciatic schwannoma in the left foraminal L5-S1 zone extending to the left ischial groove with chronic sciatica that was diagnosed radiologically with a combination of conventional MRI and MR neurography and confirmed histopathologically by surgical resection. The patient previously had conservative therapy, but the complaints were not reduced. Nonsurgical therapy is considered the first choice, and surgical therapy is indicated in cases that do not respond to conservative therapy, with recurrent cysts, severe pain, or neurological deficits [2)].

A 66-year-old man presented with leg pain, paresthesia, and weakness for 2 years. Magnetic resonance imaging demonstrated a large mass lesion involving a continuous multi-lobulated bead-like mass and a cystic portion from L1 to S3. The lesion was iso-intense on T2-weighted images (WI), iso- to slightly low-intense on T1-WI, and heterogeneous enhancement on contrast-enhanced T1-WI. The large mass lesion had three portions, including a cystic mass at L1, continuous multi-lobulated bead-like mass with a cystic portion from L2 to S1, and multi-lobulated mass from S2 to S3, which were identified with severe adhesions with cauda equina on operative assessment. Grossly total extirpation was achieved at the lumbar spine, and remained three round shaped small masses at the lumbar area and a multi-lobulated round masses from S2 to S3 involving nerves related with motor function of the lower extremities and anal sphincter, respectively. Histological examination revealed multinodular or plexiform growth pattern composed of spindle-shaped tumor cells, which were diffusely and strongly positive for S100 protein with KI67 < 1%. There were no recurrence of preoperative symptoms and changes of the remained masses over a 2-year follow-up period.

Subtotal extirpation to minimize neural deficits and close observation can be considered an appropriate treatment strategy for a giant spinal PS considering its benign prognosis and histological features, with a high risk of neurological damage during surgery [3)].

A 66-year-old male had an incidental left-sided paraspinal mass discovered while undergoing workup for cholecystitis. On examination, the patient was neurologically intact. Imaging revealed the presence of a contrast-enhanced, partially cystic mass arising from the L3-4 intervertebral foramen and causing left psoas muscle displacement. A minimally invasive left L3-4 posterior extracavitary resection was done. Histopathologic examination revealed a partly unencapsulated tumor with higher than usual cellular density and nuclear atypia, resulting in a diagnosis of “atypical schwannoma.” Imaging at 6 months' follow-up showed stable postsurgical changes and residual tumor with no evidence of progression/recurrence.

Atypical schwannoma has higher cellular density and nuclear atypia and lacks encapsulation. A review of the literature suggests an increased risk of recurrence when compared with typical variants, and complete tumor removal should be attempted [4)]

A 48-year-old female presented with dysuria and right leg pain. Initial magnetic resonance imaging (MRI) revealed a well-delineated intradural cystic lesion at the level of L4-S1 vertebrae that was isointense with cerebrospinal fluid on both T1- and T2-weighted images. A follow-up MRI 6 months later showed that the tumor had moved to the level of L2-L4; it also revealed tortuous configuration of nerve roots of the cauda equina. The tumor was resected, and a diagnosis of schwannoma with extensive cystic degeneration was pathologically confirmed. CONCLUSIONS Various possible mechanisms have been suggested for the mobility of extramedullary tumors. In the present case, MRI findings indicated the cause of the tumor movement might be attributed to the laxity of nerve roots. Besides, it is highly atypical for a schwannoma to present an entirely cystic appearance, and the combination of the 2 extraordinary features made preoperative diagnosis difficult. However, 16 out of 22 (73%) of previously reported mobile spinal tumors were schwannomas, so the differential diagnosis for a mobile spinal tumor should include schwannoma, even when the lesion seems entirely cystic on MRI. To minimize the risk of complications and additional surgical dissection, physicians should acknowledge that spinal tumors can migrate [5)].

A 36-year-old woman experienced progressive, severe lumbar radicular pain in the second trimester of pregnancy which became intractable soon after delivery. Magnetic resonance imaging revealed a complex heterogeneous hypointense mass lesion around the conus. There were two small punctate lesions in the cauda equina suggestive of myxopapillary ependymoma with 'drop metastases.' The patient underwent surgical resection of two cystic compressive conus masses. Her low back pain improved after surgery. The masses were consistent with cystic/cellular schwannomas. An incidental finding was of a small, low-grade spinal ependymoma which lacked the characteristic histologic features of myxopapillary ependymoma. Multiple, large cystic schwannomas are not uncommon in schwannomatosis, however, the co-occurrence of low-grade ependymoma strongly suggests a clinical diagnosis of new, sporadic neurofibromatosis type 2. Although cranial nerve schwannomas (orbital, auditory) have been reported to enlarge during pregnancy, to our knowledge, this is the first report of multiple cystic/cellular schwannomas causing severe pain (due to conus compression) during and immediately after pregnancy [6)]

A case of a giant cystic intradural schwannoma of the lumbosacral region in a 30- year-old man who presented with a 2-year history of non-specific lower back pain. Lateral radiographs demonstrated scalloping of the posterior wall of L5 and the upper sacrum. Magnetic resonance imaging revealed a 12 x 2.3-cm intradural multi-septated cystic lesion extending from L3 to S2 with predominant hypointense signal on T1-weighted images and a mixed signal on T2-weighted images. There was heterogeneous rim enhancement of the retrosacral portion of lesions following the administration of gadolinium contrast. The tumour was completely excised. Histological investigation confirmed the diagnosis of cystic schwannoma with alternating hypercellular (Antoni A) and hypocellular (Antoni B) areas in a fibrillar background. The patient had complete relief of symptoms and remained asymptomatic after 2 years of follow-up [7)].

A pauci-symptomatic 55 year-old male patient whose complaint was solely a non specific lumbar pain. Investigation revealed a large cystic lesion comprising the lower lumbar intradural space. He was then treated with microneurosurgical technique involving complete removal of the tumor and reconstruction of the dura mater. Histological and immunohistochemical diagnosis were consistent with cystic schwannoma. The patient presented with complete recovery of his symptom [8)].

Case report from the HGUA

62 year old male patient with clinical symptoms of paresthesias/dysesthesias on the left L3 with suggestive findings of a probable intradural and predominantly extraforaminal schwannoma, with cystic content, exhibiting growth since last MRI.

Left foraminal tumor at the L3-L4 level related to the left L3 nerve root, of cystic nature with well-defined margins and peripheral enhancement predominantly in the lower margin. It occupies the left L3 neural foramen, presenting a preferentially epidural location with a small intradural extension at that level. It remodels the cortical bone of the left posterior wall of L3 and the vertebral pedicle.

Consistent with cystic schwannoma.

Measures 2.3 x 2.8 x 3 cm (APxTx CC).

Other lumbar intersomatic levels show no relevant alterations with moderate mechanical changes due to incipient disc dehydration.

T1

75% are isointense, 25% are hypointense.

T2

More than 95% are hyperintense, often with mixed signal

T1 C+

Virtually 100% enhance

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