CASE REPORT

Spinal Intramedullary Epidermoid – A rare occurrence

MANZOOR AHMAD, RIZWAN BUTT, NAZIR AHMAD

SUMMARY

Intramedullary epidermoid cysts of the spinal cord are rare tumors, especially those not associated with spinal dysraphism. We report a case of spinal intramedullary epidermoid cyst. It was associated with spina bifida. It was located within the conus medullaris. The clinical features, MR imaging characteristics and surgical treatment are discussed and the relevant literature reviewed.

INTRODUCTION

Intraspinal epidermoid cysts represent less than one percent of all intraspinal tumors in adults. Intramedullary epidermoids are still rarer. We present a case of intramedullary epidermoid tumor and review the relevant literature.

CASE REPORT

A 23 years old girl presented with history of pain in the left thigh for four years. She developed progressively worsening difficulty in walking associated with stiffness and paraesthesias of the legs for the last two years. There was no history of trauma or any surgical procedure on the spine. Clinical examination revealed spastic paraparesis with power of grade 4 (MRC grading) on the left side and 3-4/5 on the right side. Sensory examination revealed a sensory loss to pain in lumbar segments. Deep tendon reflexes were exaggerated in both the lower limbs.

MRI showed tethered cord and diastematomyelia with associated bony spur and low-lying conus. There was evidence of an intramedullary tumor in the form of widening of the cord at that level, the lesion being hypointense on T1 weighted sequence and hyperintense on T2 sequence.

The patient underwent L1-L5 laminectomy. At surgery, there was a bony spur extending from the lamina of L4 to the posterior surface of the body of that vertebra. There was diastematomyelia as well. The cord was wide & distorted. The bony spur was excised with the help of bone reongers & drill. The anterior dura was repaired. Following a midline dorsal myelotomy, a pearly white and flacky lesion within the cord substance was seen. Total excision of the lesion was performed using microneurosurgical technique. Small portions of the capsule densely adherent to the cord parenchyma were left behind.

Department of Neurosurgery, Services Hospital, Lahore
Correspondence: Dr. Manzoor Ahmad, Assistant Professor Neurosurgery, Email; *manzoor63@gmail.com*

The patient gradually improved and at six months of follow-up, she had power of grade 4+/5 with mild spasticity and was able to walk without support

DISCUSSION

The incidence of epidermoid cysts among intracranial tumors in most large series is estimated to be 0.2%-1%. It is much less amongst the spinal tumors. Guidetti and Gagliardi reported a series of 3844 intracranial and 452 intraspinal tumors in patients of all ages and found incidence of 0.8% for cranial epidermoid cysts and 0.7% for spinal epidermoid cysts. The incidence of intramedullary epidermoid cysts is still rarer. These tend to occur at the cauda equina[3]. The epidermoid cysts can be either congenital or acquired. Manno et al reported a series of 90 intraspinal epidermoid cysts collected from the literature, of which 39 were acquired and 51 congenital. Acquired epidermoid cysts have been found years after single or multiple lumbar punctures and are thought to result from iatrogenic penetration of skin fragments. It is generally believed that congenital epidermoid cysts originate from displaced ectoderm inclusions arising in early fetal life and possibly may be associated with defective closure of the dural tube[1,2,4,5,6,7].

The frequency of intramedullary epidermoid tumors is low. Chiari reported the first case in 1833. Roux et al in 1989 reviewed and collected 46 patients from the literature and added one of their own. Since then four other cases have been reported. Thoracic region is the favorite site of the intramedullary epidermoid cysts (especially between D4-D8 levels). The lumbar cord is the next common area. There are no reports of occurrence of intramedullary epidermoid cysts within the cervical canal. The association of these tumors with defect of the overlying bone is possible but is less frequent than in dermoid or some extra medullary epidermoids[1,6].

Histologically the epidermoid cyst contains desquamated epithelium surrounded by keratin-producing squamous epithelium[5,6,7].

They may be associated with a dermal sinus or occur in isolation. When not associated with dermal sinus, they may present with progressive compressive myelopathy or acute onset chemical meningitis due to the rupture of the cyst and the spread of cholesterol crystals in the CSF. The thoracic, lumbar and sacral spine are affected, with a slight increase in incidence in the craniocaudal direction[1,5,6,7].

CT and MRI characteristics are similar to those seen in epidermoids at other sites. The CT scan of an epidermoid cyst is characterized by a low density lesion that does not enhance[1,5,6,8].

Epidermoid cysts are generally characterized on MR images by an important variability of signal intensity between the different cases and at times between the different parts of the same cyst; other features include the absence of edema in surrounding tissue, fairly well defined limits and peripheral enhancement on injection of gadolinium. The disparity in signal intensity most likely reflects variable lipid and protein composition in these lesions. Moreover, the margins of these lesions are 'shaggy', possibly because of chronic inflammatory response to the squamous tissue 'leak' through the capsule and variable gliosis along the margin extending into the cord. This feature may be of help in differentiating these lesions from other intramedullary tumors[1].

Most authors do not attempt total removal of the capsule when it is intimately attached to the spinal cord or located within its confines. Attempts to remove the cyst wall completely under these circumstances are unnecessary and carry a high risk of neurological deficit. The risk of recurrence exists. However, in most cases, even partial removal of the intramedullary epidermoid resulted in total remission of symptoms. If recurrence does occur, another surgery may relieve the symptoms again[1,5,6,9].

In addition to the general complications of any surgical procedure, the operative complications include damage to the neural and vascular structures and sphincter disturbance but an operative complication unique to epidermoid is aseptic/chemical meningitis which can last for a number of weeks. If it is severe and long-lasting, it can lead to a “granulomatous” type of arachnoiditis[9].

CONCLUSIONS

1. Spinal epidermoids are very rare while intramedullary epidermoid cysts are still rarer.
2. These can be either congenital or acquired.
3. The patient may present with neurological deficits or occasionally with chemical meningitis.
4. MRI is the investigation of choice.
5. Complete excision should be attempted under microscope by an experienced neurosurgeon.
6. To avoid acquired epidermoids, lumbar puncture should be done very carefully preferably with a fine bore needle.

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