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Intramedullary epidermoid cyst of spinal cord: Experience of 24 Cases at a Tertiary Care Institute

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Abstract

Introduction: Epidermoid cyst is the sequel of developmental aberration often intriguing the neurosurgeons. Intramedullary epidermoid cysts are rare entities with delay and variability in the clinical presentation. The aim of present study is to analyze clinical presentation and management issues of this entity of neurological pertinence.

Materials and Method: The study is retrospective analysis of cases operated for intramedullary epidermoid cysts of spinal cord at King Edward Memorial Hospital, Mumbai over a period of 16 years from 2001 to 2016. All relevant hospital records and follow-up were analyzed.

Results: The study revealed that this entity more commonly manifests in males of second and third decades. The age at the time of presentation ranged from 3 to 51 years (mean age-24.2 years). There were 15 male and 9 female patients. The average duration of complaints was 7 months. The backache and stiffness of both lower limbs were most common presentations followed by sphincter disturbances and dysaesthesia. Our follow up ranged from a period of 6 months to 15 years with a mean of 9.6 years. Patients had excellent clinical recovery with safe excision of cyst contents with recurrence in three patients.

Conclusions: Our study highlights that patients of intramedullary epidermoids have satisfactory outcome with safe resection of cyst content leaving behind the densely adherent capsule.

Keywords: Epidermoid cyst, Intramedullary lesions, Spinal cord, Inclusion cysts

Introduction

Epidermoid tumors are benign developmental lesions. Intracranial epidermoid cyst occurs more commonly with a ratio of cranial to spinal epidermoids reported at 14:1.[1,2,3,4] The incidence of epidermoid cysts among intracranial tumors in most large series is estimated to be 0.2%-1%. Whereas, epidermoid cysts account for 0.7% of all intraspinal tumours.[1,2,3,4] Most spinal epidermoids are intradural extramedullary in location and are usually associated with spinal dysraphism.[3,4]

Materials and Method

This was a retrospective study of cases operated for intramedullary epidermoid cysts of spinal cord at King Edward Memorial Hospital, Mumbai over a period of 16 years from 2001 to 2016. The case histories, operative details and radiological imaging were obtained from hospital records. All patients were called for follow up with clinical evaluation and fresh radiological imaging.

Preoperatively all patients were evaluated clinically followed by magnetic resonance imaging (MRI). The operative procedure was standardized. Standard laminectomy, extending from one level above to one level below the extent of lesion, was performed. The dura was opened in the midline. A midline myelotomy with total excision of the pearly content leaving behind the capsule was done. Only loose capsular fragments were removed, whereas capsular fragments adherent to cord parenchyma were left behind. The patients were evaluated for immediate neurological status and a delayed neurological examination was performed at regular intervals. Post operative stay at the institute ranged from a period of 2 days to 30 days with an average of 11.8 days. Histology was suggestive of epidermoid cyst. One lesion showed presence of calcification in the wall. The patients were subjected to delayed post operative imaging at 3 months interval and one at long term follow up.

Results

Analysis of our data (Table 1 & 2) revealed that males were more commonly involved. The second and third decades were the commonest age group involved. The youngest was 3 years old and the oldest patient was 51 years old. (Mean age of 24.2 years). Back pain and difficulty in walking were the commonest presenting symptoms followed by bladder disturbances and sensory symptoms. Examination revealed gait disturbances in 9 patients and spinal deformity in 3 patients. There was hypertonia of lower limbs in 11 patients with associated exaggerated reflexes and extensor type of plantar reflexes. The commonest bladder disturbance was incontinence of urine followed by hesitancy and retention. The duration of symptom ranged from a period of 2 months to 5 years with a mean of 17 months. Two of these patients had been operated previously for tethered cord and one was operated for extramedullary lipoma before presentation to our centre. One patient had features of neurofibromatosis and one patient had chronic renal failure on presentation. One patient had associated

spina bifida and dermal sinus in upper dorsal region (Fig. 1).

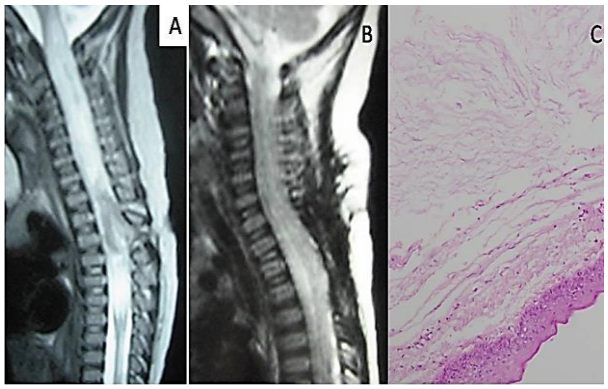

Fig. 1

1A: T2-weighted sagital image of Magnetic resonance imaging (MRI) of cervicodorsal spine showing intramedullary lesion with tethering of cord to overlying dermal sinus.

1B: Postoperative T2-weighted sagital image of MRI of cervicodorsal spine showing excision of intramedullary epidermoid and excision of tethering band of cord.

1C: Histological picture showing desquamated amorphous keratin and lining of stratified squamous epithelium characteristic of epidermoid cyst.

Table 1: Demographic and clinical profile

Age Range	3 -51 years
Mean Age	24.2 years
Gender Ratio(M:F)	15:9
Duration of Symptoms	2 months to 5 years
Mean Duration of Symptoms	7 months
Frequency of Presenting Symptoms	Back pain > difficulty in walking > bladder disturbances and dysesthesia
Presentation after Previous surgery for spinal dysraphism	3
Presence of Bladder Symptoms	11
Presence of Upper Motor Neuron Signs, Extensor Plantar Reflex	11
Presence of associated features/comorbidities	
Neurofibromatosis	1
Chronic renal failure	1
Site of lesion	
Brainstem to lower cervical region	1
Cervical	2
Upper dorsal	10
Lower dorsal	6
Intramedullary of conus region with variable involvement of cauda equina	5

Table 2: Clinical outcome

Parameter	Frequency
Transient deterioration of power	4
Post –op improvement in power	
Improvement	8
Maintained as pre operative status(paraplegic)	1
Post-op improvement in bladder disturbance	5
Recurrence of lesion	3
Death	1

M R Imaging revealed intramedullary lesion which was hypo intense on T1W and hyper intense on T2W images with no or mild heterogeneous post contrast enhancement. There was evidence of spinal bifida and tethered cord in 2 cases, one patient had evidence of subcutaneous dermoid and extra medullary lipoma. The commonest segment involved was the dorsal spine followed by the lumbar segments and cervical region.

Our follow up ranged from a period of 6 months to 15 years with a mean on 9.6 years. In the immediate post operative period four patients had transient deterioration of power and one had transient radicular pain in the right thigh. Those patients improved at interval of 3 to 6 weeks. One of our patient, who had giant multi-segmental intramedullary epidermoid extending from brainstem to dorsal spinal cord, developed respiratory depression in postoperative period and died due to septicemia. This patient had severe quadriparesis and poor general condition at time of admission due to extensive disease. Of the 10 patients who had lower limb weakness 8 improved in powers, one patient who was paraplegic did not improve. Of the 10 patients who had bladder disturbances 5 did not improve whereas the rest improved and could pass urine independently. Three patients had symptomatic recurrent tumor who responded well to resurgery.

Discussion

Epidermoid tumors were first described by Cruveilhier in 1835, who called them “tumor perlees” (pearly tumors). Hans Chiari in 1883 was the first to describe an intramedullary epidermoid cyst. In 1934, Gross reported the first successful surgical treatment of an intramedullary epidermoid cyst.[1,2,3,4]

Thoracic region is the most common site of the intramedullary epidermoid cysts (especially between D4-D8 levels. The lumbar cord is the next common area; however the cervical cord is the least common site. The association of these tumors with defect of the overlying bone is very rare and less frequent than in dermoid or extra medullary epidermoids.[5,6,7,8]

The epidermoid cysts can be either congenital or acquired.[1,2,3,9] Congenital epidermoid cysts are believed to originate from displaced nests of ectoderm

inclusions or epidermal cells in early fetal life between 3rd and 4th weeks of gestation. Defective closure of the dural tube or neural tube may lead to a patent communication with exterior and subsequent infection or intramedullary abscess in few cases. Acquired epidermoid cysts have been found years after single or multiple lumbar spinal punctures and are thought to result from iatrogenic penetration of skin fragments. Such implantation can occur during a variety of procedures and events such as bullet wounds, surgery, myelography or punctures for diagnosis, anesthesia or treatment.[8,9]

Intraspinal tumors are rare in infancy and childhood, but spinal epidermoid and dermoid cysts occur more frequently in this age group. Intramedullary epidermoid cysts are very slow-growing tumors, usually becoming apparent after the 2nd decade of life.[8,10,11] The slow growth of epidermoid cysts often leads to a delay in their diagnosis. Signs and symptoms at presentation are usually neurological, such as progressive paraparesis, sensory loss and back/leg pain that can be variable according to the specific tumor localization. Urological manifestations result due to stretching of fibers of central cord.[12,13,14] The giant intramedullary epidermoids have been reported with worse prognosis.[15,16]

Biochemical analysis of the cyst material shows considerable fat but negligible cholesterol. Free fatty acids and cholesterol can be carried into the mass by inflammatory cells in response to necrosis of the degenerating squamous cells. The escape of epidermoid-cyst contents into the subarachnoid space may produce a granulomatous or chemical meningitis.[1,2,3,4]

MRI has revolutionized the diagnosis of intramedullary tumors, reducing the interval between onset of symptoms and diagnosis. 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, well-defined limits, and the presence of calcifications. An intravenous injection of gadolinium demonstrates peripheral enhancement. The disparity in signal intensity most likely reflects variable lipid and protein composition in these lesions.[4,7,14]

Histologically, epidermoid are lined by stratified squamous epithelium supported by an outer layer of collagenous tissue; progressive desquamation of keratin from epithelial lining toward the interior of the cyst produces a soft white material- Epidermoid cyst differ from dermoid cyst in conspicuous absence of dermal appendages.[1,4,8]

The primary management of epidermoid cysts is surgical. The basic philosophy of treatment should be excision of cyst content and leaving behind the capsule adherent to the surrounding eloquent parenchyma in view of benign nature of lesion and low potential for clinical recurrence.[4,7,8] Attempts at complete capsular resection are unnecessary and carry a serious risk of substantial neurological deficit. If recurrence does occur, another surgery may relieve the symptoms again. The prognosis for epidermoid tumors of the spine is good, since these have histologically benign nature. Patients with neurological findings should undergo early surgery. Preoperative symptoms may resolve completely, even after partial resection of the cyst.[4,7,8]

Conclusion

Surgery of intramedullary epidermoids carries innate difficulties due to dense adhesions of capsule to the eloquent cord parenchyma. These patients have excellent outcome with safe resection of cyst content leaving behind the capsule.

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