Spinal intramedullary glioependymal cyst presented with paraplegia in an adult

Pankaj Kumar Gupta, R. S. Mittal, Pankaj Gupta, Ashok Gandhi, Varsha Kumar[1]

Department of Neurosurgery, Sawai Man Singh Medical College, Jaipur, Rajasthan. [1]Department of Pathology, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, Uttar Pradesh, India

ABSTRACT

We report a case of intramedullary glioependymal cyst at the level of D11-D12 vertebra in 38 year old male patient, who presented to our hospital with progressive spastic paraplegia. The preoperative magnetic resonance image spine revealed a well-defined intramedullary cystic lesion at D11-D12 level, which was hypointense on T1W1, hyperintense on T2W1 and showing ring type enhancement on post contrast images. The patient underwent D11-D12 laminectomy, durotomy, dorsal midline myelotomy with fenestration and decompression of intramedullary cystic lesion. Biopsy was taken from the cyst wall. The histopathological study of specimen was suggestive of glio-ependymal cyst. Postoperatively patient improved neurologically but there was residual paraparesis at the time of discharge. To the best of our knowledge, only 20 cases have been reported so far. Considering this rarity and reviewing the literature, we present a case of spinal cord glioependymal cyst along with radiological, surgical, pathological and immunohistochemistry findings.

Key words: Glioependymal cyst, intramedullary, immunohistochemistry

INTRODUCTION

Spinal intramedullary glioependymal cysts are extremely rare and pathologically proven cases are few in literature.[1-3] Their association with other congenital lesions (e.g, tethered cord) is still rarer.[4,5] It originates from the ectopic ependymal cells situated parallel to the central canal during the development of the spinal axis along the spinal column.[6,7] In the brain, the common location of the cyst is paraventricular white matter of frontal and parietal lobes.[7,8] The different varieties of developmental intradural spinal cord cysts include enterogenous cyst, bronchogenic cyst, teratomatous cyst, arachnoid cyst, neurenteric cyst, foregut cyst, epithelial cyst, colloid cyst and ependymal cysts. Glioependymal cysts differ from other congenital enterogenous cysts which results from displaced elements of alimentary canal and do not exhibit goblet cell differentiation.

Access this article online	
Quick Response Code:	Website: www.ijns.in
	DOI: 10.4103/2277-9167.146834

CASE REPORT

A 38 year old male patient presented with progressive weakness of both lower limbs with bladder and bowel dysfunction since last 1.5 months. On examination, bulk was normal, hypertonia was there in both lower limbs, power was 0/5 at hip, knee and ankle, deep tendon reflexes were brisk with patellar and ankle clonus present. Babinsky was present bilaterally and there was a loss of fine touch, crude touch, pain and temperature sensation below D12 level.

Magnetic resonance image showed well defined cystic lesion of size 10mm × 8mm, present at D11-D12 level, appearing hypointense on T1W1 and hyperintense on T2W1 [Figures 1 and 2]. Contrast study revealed smooth peripheral enhancement and perilesional edema from D8 to D12 level. During the surgery, D11-D12 laminectomy was done and dura opened in the midline. Adhesions were found between dura and cord. Midline dorsal myelotomy was done. An intramedullary, greyish white, partly cystic mass found, which was soft to firm in consistency, moderately vascular, nonsuckable and densely adhered to cord parenchyma. There was no definite plane of cleavage found between cystic mass and cord. Fenestration and decompression of the cyst was done and biopsy taken from the cyst wall [Figure 3]. Dura was closed in a water tight fashion.

Address for correspondence: Prof. R. S. Mittal,
Department of Neurosurgery, Sawai Man Singh Medical College, Jaipur - 302 004, Rajasthan, India. E-mail: drpankaj.gupta@yahoo.co.in

This document was downloaded for personal use only. Unauthorized distribution is strictly prohibited.

Histopathological examination [Figure 4] of specimen showed cyst wall composed of a single layer of ependymal cells resting directly upon a densely gliotic stroma. There was no intervening basement membrane or connective tissue between the ependymal cell lining and gliotic supporting cell line. Immunohistochemistry [Figure 5] was showing glial fibrillary acidic protein (GFAP) reactivity in the glial element and S-100 reactivity in ependymal element of cyst. No reactivity to cytokeratin was present. These findings are suggestive of glioependymal cyst. Postoperative recovery was good and patient improved with power grade 3/5 along with improved sensations of all modalities. But while discharge from hospital the bladder and bowel dysfunction remained same.

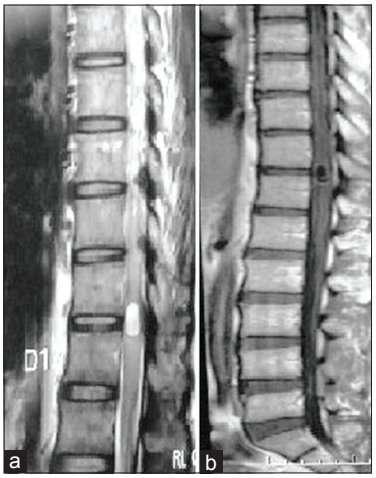

Figure 1: (a) T2W1 magnetic resonance image (MRI) showing hyperintense cystic intramedullary lesion at D11-D12 disc level. (b)-Contrast T1W1 MRI showing hypointense cystic intramedullary lesion with peripheral rim enhancement on sagittal cut

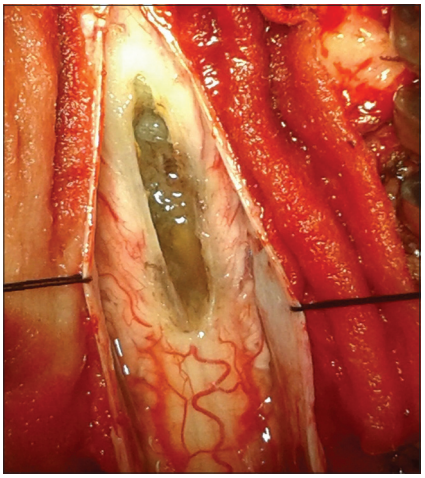

Figure 3: Residual intramedullary cystic mass lesion densely adhered to the cord structure. No definite plane of cleavage is visible and good amount of decompression achieved by fenestration and biopsy of the cyst wall

DISCUSSION

Intradural spinal glioependymal cysts are rare and comprise 0.4% of all primary spinal cord tumors and even more uncommon in pediatric age group (less than 20 years).[6,9] Almost all cases are embryonic in origin and may be found with associated tethered cord.[4,5] The most common site of intramedullary glioependymal cyst is dorsal spine.[7] The most widely accepted hypothesis regarding the formation of glioependymal cysts hold that the floor plate of the neural tube is evaginated on the ventral side and becomes isolated to form an ependymal cyst.[4,8] The location of these isolated ependymal cells determines whether the cyst presentation is intramedullary or extramedullary but intramedullary spinal ependymal cysts are rarer. Sometimes, neurenteric cysts have been confused with glioependymal cyst, though the latter has characteristic absence of basement membrane and they lack mucin.[1,2] Immunohistochemistry examination of the cyst wall shows positivity for GFAP in the glial component and S-100 reactivity in ependymal

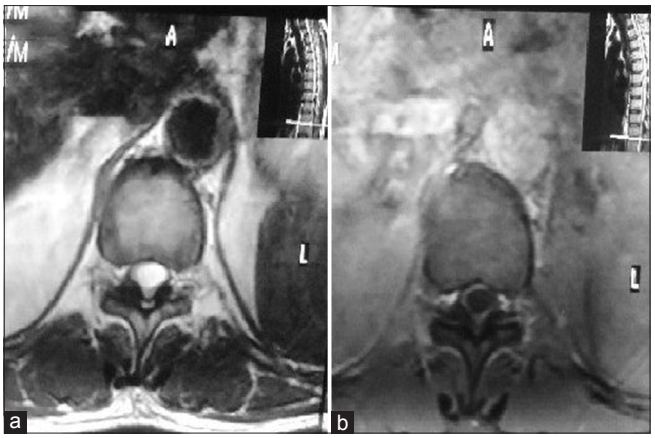

Figure 2: (a) Contrast axial magnetic resonance image showing cystic intramedullary lesion which is hypointense on T1W1 and hyperintense on T2W1 with thin peripheral ring enhancement (b)

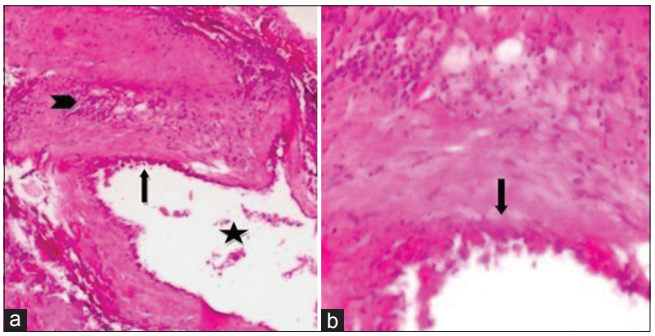

Figure 4: (a) Photomicrograph at ×10 showing cystic lesion (asterisks) lined by ependymal cells (arrow) surrounded by glial cells (arrow head). (b) photomicrograph at ×40 showing complete absence of basement membrane (arrow) between ependymal cell lining and glial component, which is a characteristic feature of glioependymal cyst

This document was downloaded for personal use only. Unauthorized distribution is strictly prohibited.

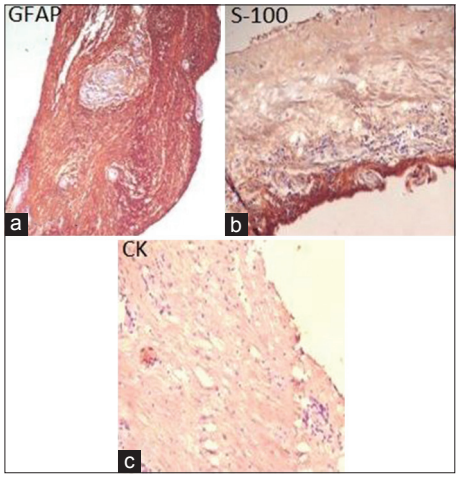

Figure 5: (a) Immunohistochemistry study showing glial fibrillary acidic protein positivity in the glial element of cyst, (b) S-100 reactivity in ependymal cell lining, (c) No reaction to cytokeratin

component of cyst. It shows no reactivity to epithelial membrane antigen and Cytokeratin, which differentiates the glioependymal cyst from other neuroenteric cysts.[6,8] In addition, cyst wall stain is positive for CAM 5.2, AE1/AE3 keratin.[1,5] The borders of cyst appear to be smooth and well defined. It is a benign cyst, and there is no evidence of neoplastic transformation.[2] Symptomatic intramedullary spinal cysts need surgical treatment with complete excision and closure of its communication with the subarachnoid space, although complete surgical excision of intramedullary glioependymal cyst is difficult due to adhesions with cord structure.[2,3,6] Other options of treatment are marsupialization of cyst, fenestration and resection of the cyst wall, placement of the shunt between cyst to the pleural space or subarachnoid space and biopsy.[8,9] Total excision of the cyst should be avoided if there is no definite plane of cleavage.[2,3]

CONCLUSION

We report a very rare spinal intramedullary glioependymal cyst. These cysts are benign in nature. They have good functional recovery and associated with low recurrence rates. We emphasize complete cyst excision only if there is a plane of cleavage present otherwise cyst fenestration, biopsy of cyst wall, and marsupialization are the safest surgical method.

REFERENCES

1. Sharma BS, Banerjee AK, Khosla VK, Kak VK. Congenital intramedullary spinal ependymal cyst. Surg Neurol 1987;27:476-80.
2. Chhabra R, Bansal S, Radotra BD, Mathuriya SN. Recurrent intramedullary cervical ependymal cyst. Neurol India 2003;51:111-3.
3. Dharker SR, Kanhere S, Dharker RS. Intramedullary epithelial cyst of the spinal cord. Surg Neurol 1979;12:443-4.
4. Balasubramaniam C, Balasubramaniam V, Santosh V. Intramedullary glioependymal cyst and tethered cord in an infant. Childs Nerv Syst 2004;20:496-8.
5. Takci E, Sengul G, Keles M. Spinal intramedullary ependymal cyst and tethered cord in an adult. Case report. J Neurosurg Spine 2006;4:506-8.
6. Findler G, Hadani M, Tadmor R, Bubis JJ, Shaked I, Sahar A. Spinal intradural ependymal cyst: A case report and review of the literature. Neurosurgery 1985;17:484-6.
7. Kumar R, Nayak SR, Krishnani N, Chhabra DK. Spinal intramedullary ependymal cyst. Report of two cases and review of the literature. Pediatr Neurosurg 2001;35:29-34.
8. Pagni CA, Canavero S, Vinattieri A, Forni M. Intramedullary spinal ependymal cyst: Case report. Surg Neurol 1991;35:325-8.
9. Iwahashi H, Kawai S, Watabe Y, Chitoku S, Akita N, Fuji T, *et al.* Spinal intramedullary ependymal cyst: A case report. Surg Neurol 1999;52:357-61.

How to cite this article: Gupta PK, Mittal RS, Gupta P, Gandhi A, Kumar V. Spinal intramedullary glioependymal cyst presented with paraplegia in an adult. Indian J Neurosurg 2014;3:163-5.

Source of Support: Nil, **Conflict of Interest:** None declared.

This document was downloaded for personal use only. Unauthorized distribution is strictly prohibited.