Intradural Extramedullary Epidermoid Cyst at the Conus Medullaris level with Thoracic Syringomyelia: a Case Report

Bekir Akgun¹*, Ahmet Cemil Ergun¹, Ibrahim Hanifi Ozercan², Selman Kok¹

ABSTRACT
Spinal epidermoid cysts are benign tumors. Syringomyelia secondary to intramedullary tumors are frequently observed. However, the association between syringomyelia and spinal intradural extramedullary epidermoid cyst in the conus medullaris region is extremely rare. We present the case of a 3-year-old male who was admitted with paraparesis and urinary retention. Magnetic resonance imaging (MRI) of the spine demonstrated intradural extramedullary lesion, compatible with epidermoid cyst, that at the conus medullaris level and a large syringomyelia extending from T4 to L1 vertebrae. Total microsurgical excision of the cyst was performed. No additional drainage was carried out for the syringomyelic cavity. Histopathological examination verified the diagnosis of the epidermoid cyst. Total excision of the cyst and disappearance of the syringomyelia were observed on MRI at 15 days postoperatively. We have clarified the etiology, clinical, histopathological and radiological features, differential diagnosis, and treatment modalities of spinal epidermoid cysts. In addition, we have discussed the possible mechanisms of syringomyelia formation in spinal intradural lesions.

KEYWORDS
conus medullaris; epidermoid cyst; syringomyelia; spine

AUTHOR AFFILIATIONS
¹ Firat University Hospital, Departments of Neurosurgery Elazig, Turkey
² Firat University Hospital, Clinical Pathology, Elazig, Turkey
* Corresponding author: Firat Universitesi Hastanesi, Beyin Cerrahisi Klinigi, 23119, Elazig, Turkey; e-mail: bekirakgun@yahoo.com

Received: 12 July 2018
Accepted: 10 November 2018
Published online: 1 April 2019

https://doi.org/10.14712/18059694.2019.45
© 2019 The Authors. This is an open-access article distributed under the terms of the Creative Commons Attribution License (http://creativecommons.org/licenses/by/4.0), which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.
INTRODUCTION

The term “inclusion cysts” includes dermoid and epidermoid tumors/cysts. They are derived from cutaneous ectoderm (1). Spinal epidermoid cysts are rare and slow-growing benign lesions. They constitute <1% of all spinal tumors (2, 3). Most of the spinal epidermoid cysts are located in the lumbosacral and thoracic regions (4). Conus medullaris epidermoid cysts are extremely rare entities.

Syringomyelia is characterized by the presence of cystic cavities inside the spinal cord. It is a secondary process with many etiologies. Chiari malformations, neoplasms, arteriovenous malformations, arachnoiditis, and spinal dysraphisms are some examples (5). Syringomyelia associated with intramedullary tumors are frequently observed. The most common tumors associated with syringomyelia are astrocytomas and ependymomas. In addition, their most common locations are the lower cervical and upper thoracic regions (6, 7). However, extramedullary tumors at the conus medullaris level associated with syringomyelia are uncommon coexistences. Here we describe and discuss the case of a patient with conus medullaris epidermoid cyst associated with a large thoracic syringomyelia.

CASE REPORT

A 3-year-old male was admitted with increasingly leg weakness that had persisted for 2 months. Neurologic examination revealed that the proximal and distal muscular forces in his right and left lower extremities were 3/5 and 2/5, respectively. A large syringomyelia, extending from T4 to L1 vertebrae, was detected on thoracic magnetic resonance imaging (MRI). In lumbar MRI, T1-weighted imaging demonstrated isointense expansion at the L1-2 vertebrae level. T2-weighted imaging revealed an intradural extramedullary regular lesion that had hyperintense capsule-like formation with isointense content and that excessively compressed the conus medullaris. It was approximately 15 × 15 mm in size. T1-gadolinium-enhanced imaging showed that dense cystic mass had circumferential contrast enhancement. Besides, some cauda equina fibers had contrast enhancement (Figure 1). In addition, renal ultrasonography was performed before and after micturition. Significant post-micturition residue was determined. He underwent surgery. Total microsurgical excision of the lesion was achieved with the help of neuromonitoring after L1 and L2 laminotomies. Also, the integrity of the facet joints was protected. After dural opening, the lower end of the spinal cord (conus medullaris) and the beginning of the cauda equina fibers appeared compressed by an anterior extramedullary lesion. Dense pearly content and whitish cyst capsule were completely excised. No additional drainage procedure for the syringomyelic cavity was considered at this surgical intervention. Besides, CSF leakage was not observed from the neighbouring syringomyelic cavity after cyst removal. In the closure, L1 and L2 laminoplasties were performed. In the early postoperative period, muscular force in his right and left lower extremities had improved to 4/5 and 3/5, respectively. Histological examination of the specimens demonstrated that the cyst walls were lined with stratified squamous epithelium without skin adnexa. Furthermore, desquamation of keratin from the epithelial lining was detected. This examination revealed epidermoid cyst (Figure 2). There was no finding of residual tumor on MRI at 15 days postoperatively. In addition, disappearance of the syringomyelia was seen. However hyperintensity at T2 weighted image compatible with myelomalacia, existed after the cyst resection, was observed (Figure 3A,B,C). At 20 days postoperatively, renal ultrasonography was performed before and after micturition, which showed decreased post-micturition residue. Physical therapy and rehabilitation were indicated at 1 month postoperatively. In the second month control his examination revealed no neurological deficits. Besides, significant decrease of myelomalacia was seen at the second month follow-up MRI. (Figure 3D).

Fig. 1 In the preoperative (A) thoracic MRI, a syringohydromyelia (arrow), extending from T4 to L1 vertebrae, was observed. Lumbar MRI (B) T1-weighted sagittal image showed hypointense expansion (arrowhead) at the L1-2 vertebrae level. (C) T2-weighted image showed a lesion that had hyperintense capsular formation with isointense content and that excessively compressed the conus medullaris. (D) T1-gadolinium-enhanced image demonstrated that dense cystic mass had circumferential contrast enhancement. Besides, some cauda equina fibers had contrast enhancement.
DISCUSSION

Spinal epidermoid cysts may be acquired or congenital. Acquired epidermoid cysts, due to iatrogenic penetration of skin fragments, have been frequently reported after lumbar puncture or meningomyelocele repair with years of latency (1, 8, 9). Congenital spinal epidermoid cyst is thought to be an inclusion of ectodermal tissues during closure of the neural tube between the third and fourth weeks of embryonic development (3). Occult spinal dysraphisms, spina bifida aperta, hemivertebra, and dermoid tracts are often associated with congenital cysts (1, 3, 8). Our patient had no known history of meningomyelocele repair such as open wound trauma or lumbar puncture; therefore, the tumor was thought to be congenital.

Signs and symptoms arising from epidermoid cysts vary with the extent of involvement and are not different from other spinal space-occupying lesions. Paraparesis, sensory loss, urological manifestations, pathologic reflex appearance, deep tendon reflex changes, and back extremity pain are usually found at presentation. The slow growth of epidermoid cysts often leads to a delay in their diagnosis (2).

MRI is useful for diagnosis, particularly contrast-enhanced studies, which also show the exact location and extension of the lesion. MRI demonstrates that epidermoid cyst is hypointense on T1-weighted and hyperintense on T2-weighted images (8). Peripheral postgadolinium enhancement with well-defined limits without perilesional edema contributes to diagnosis. Some authors emphasize that peripheral enhancement represents normal peripheral surrounding tissue reaction, whereas others consider it as an outer capsule composed of tumor cells (9). In this patient, circumferential contrast enhancement was due to capsular formation. However, contrast enhancement of the some cauda equina fibers might be reactional. During

Fig. 2 In the histopathological examination, (A) cystic lesion with stratified squamous epithelium (black arrow) (hematoxylin–eosin, ×100). (B) Inflammatory cells (arrowhead), stained by leukocyte common antigen (LCA), around the cystic lesion with stratified squamous epithelium (white arrow) (LCA, ×100). (C) Stratified squamous epithelium stained with pancytokeratin. Desquamation of keratin from the epithelial lining can be observed (pancytokeratin, ×100).

Fig. 3 Postoperative MRIs. Disappearance of the syringohydromyelia was observed. There was no finding of residual tumor. Decompression of the conus medullaris and cauda equina fibers is shown. (A) T1-weighted, (B) T2-weighted (arrow: hyperintense image thought to be myelomalacia due to chronic compression, which existed after the cyst resection), and (C) contrasted sagittal section at 15 days postoperatively. (D) T2-weighted sagittal section also showed significant decrease of myelomalacic aspect at 60 days postoperatively.
the operation, we observed conspicuous capsular forma-
tion; therefore, we obtained tissue samples as well as the
capsule-like formation and dense content of the cyst for
histopathological examination.

Ependymomas are the most common space-occupy-
ing lesions at the conus medullaris-cauda equina level. Lymphomas, astrocytomas, gangliogliomas, and gan-
glioneuromas can be considered during differential di-
agnosis (7, 10, 11). They can be frequently identified and
distinguished by MRI studies. Astrocytomas, ganglioglio-
as, and ganglioneuromas have heterogeneous contrast
patterns. Ependymomas have homogenous contrast en-
hancement, and ependymoid cysts have peripheral con-
trast enhancement (7, 9, 11). Nevertheless, radiological dif-
ferentiation is sometimes difficult for these spinal lesions,
but they can be histopathologically diagnosed.

Histopathological examination is critical for diagnos-
ing of ependymoid cysts. Differentiation between the two
forms of inclusion tumors (dermoid/epidermoid) is based
on the presence of adnexal skin structures, such as seba-
ceous glands, eccrine glands, and hair follicles. They are
present only in dermoid cysts (1, 2). A specific histological
feature of the epidermoid cyst is a lined stratified squa-
amous keratinizing epithelium surrounded by an outer lay-
er of collagenous tissue and/or inflammatory cells without
skin adnexa. Desquamation of keratin from the epithelial
lining generates cholesterol crystals (2, 9).

Total excision including the capsule is the curative treat-
ment for spinal epidermoid cyst because it is a be-
nign condition. Sometimes, because of its anatomical fea-
tures, intramedullary tumor shows tight adherence to the
arachnoid membrane and spinal cord, thereby causing
some surgical difficulties for safe and complete resection.
Therefore, subtotal resection is also commonly performed
to avoid neurological deficits (1, 2). Local recurrence is re-
ported especially after subtotal excision. Incomplete exci-
sion of basal germinal cells of the tumor is suggested to be
the cause of recurrence (12). Long-term follow-up is nec-
essary for tumor recurrence risk. Surgeons should keep in
mind that cyst content includes fat and cholesterol. These
causes an inflammatory reaction that leads to aseptic
chemical meningitis (Mollaret’s meningitis) (8). Plugging
of proximal and distal arachnoid spaces and irrigating the
site with normal saline prior to dural closure reduces the
risk of chemical meningitis (9). We performed total exci-
sion in this case. Besides, the patient did not develop any
inflammatory reactions and neurological deterioration
postoperatively.

The exact pathogenesis of syringomyelia accompanied
with spinal cord tumors is unclear. Different theories have
been proposed. Some of them are intramedullary soften-
ing due to an impaired blood circulation, stasis of tissue
fluid resulting from occlusion of the drainage pathways,
effect of edema, and spontaneous autolytic reactions of the
tumor (7, 13). Another suggested mechanism of syrinx for-
mation was obstruction of cerebrospinal fluid (CSF) flow
(13, 10, 14). Given these possible reasons, syringomyelia
associated with intramedullary tumors can be frequent-
ly observed. However, extramedullary tumors associated
with syringomyelia are very rare. In our case, syringomy-
elia most likely developed secondary to the mass effect of
the epidermoid cyst. It has been postulated that cyst may
cause blockage of CSF flow dynamics. Accordingly, exci-
sion of the cyst can remove the mass effect and regenerate
these CSF flow alterations. Hence, after the total resection
of the cyst, disappearance/reduction of the syrinx cavi-
ty was achieved, in spite of CSF leakage was not observed
from the neighbouring syringomyelic cavity. Besides, no
additional drainage procedure for the syringomyelic cav-
ity was considered.

In conclusion, spinal intradural extramedullary epi-
dermoid cyst at the conus medullaris level associated with
thoracic syringomyelia is a very rare entity. MRI is very
useful for diagnosis, identifying the exact location of the
cyst, and detecting comorbidities. The clinical course is
dependent on the neurological condition of the patient
at presentation and any delay in the diagnosis. Moreover,
appropriate early surgical intervention with total exci-
sion results in good prognosis. Syringomyelia most like-
dly developed secondary to the mass effect of the cyst and
blockage of the CSF flow dynamics. Therefore, excision of
the cyst can ensure the disappearance or reduction of the
syringomyelia.

REFERENCES
1. Thompson DNP. Spinal inclusion cysts. Childs Nerv Syst 2013; 29:
1647–55.
2. Gotecha S, Ranade D, Sharma S, Punia P, Kotecha M. Giant intradu-
ral intramedullary ependymoma case report of two cases with varied
presenting with abnormal urological manifestations. Spinal Cord
5. Su DK, Ebenezer S, Avellino AM. Symptomatic Spinal Cord Compres-
sion from an Intradural Arachnoid Cyst with Associated Syrinx in a
6. Quencer RM, el Gammal T, Cohen G. Syringomyelia associated with
intradural extramedullary masses of the spinal canal. AJNR Am J
7. Sarkeraya S, Ackgog B, Tekkok IH, Gungun Y. Conus ependymoma
with holocord syringohydromyelia and syringobulbia. Journal of
tradural ependymoma cyst after repetitive epidural anesthesia:
A case report and review of the literature. World Journal of Surgical
9. Mishra SS, Satapathy MC, Deo RC, Tripathy SR, Senapati SB. Isolated
thoracic (D6) intramedullary ependymoma without spinal dysra-
10. Nagahiro S, Matsukado Y, Kuratsu J, Saito Y, Takamura S. Syringomy-
elia and syringobulbia associated with an ependymoma of the cauda
equina involving the conus medullaris: case report. Neurosurgery
11. Wang K, Dai J. Conus medullaris ganglioneuroma with syringomye-
elia radiologically mimicking ependymoma: A case report. Oncology
cyst evaluated by computed tomographic scan and magnetic reso-
13. Sami M, Klekamp J. Surgical results of 100 intramedullary tumors
in relation to accompanying syringomyelia. Neurosurgery 1994; 35:
865–73.
14. Blaylock RL. Hydroxyxirgomyelia of the conus medullaris associated