Case Report

Cavernoma of the cauda equina

Sotirios Apostolakis, Athanasios Mitropoulos, Kalliopi Diamantopoulou¹, Konstantinos Vlachos

Departments of Neurosurgery and ¹Pathology, KAT General Hospital of Attica, Kifisia 145 61, Greece

E-mail: *Sotirios Apostolakis - sotapostolakis@gmail.com; Athanasios Mitropoulos - mitrtha@otenet.gr; Kalliopi Diamantopoulou - popydiamond@yahoo.gr; Konstantinos Vlachos - kvlachos@gmail.com
*Corresponding author

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Abstract

Background: Cavernomas are benign malformations of the vasculature. In the central nervous system, they are mostly located supratentorially. However, in adults, cavernomas also comprise about 3% of all subdural spinal cord tumors. Notably, cavernomas of the cauda equina are extremely rare, with only 23 cases reported in the literature. Here, we report the 24th case involving a 77-year-old male.

Case Description: A 77-year-old male presented with low back pain for 3 years duration. His history included prostate cancer, skin melanoma, and a sick sinus syndrome requiring a pacemaker. An enhanced computed tomography of the lumbar spine showed an inhomogeneously enhanced, intramedullary mass, located at the L3 level. The patient underwent an L3 hemilaminectomy with gross total excision of the lesion. Macroscopically, the tumor was mulberry-shaped and well demarcated. However, it was strongly adherent to a nerve root of the cauda equina which required resection. The histologic examination was consistent with a cavernoma. The patient subsequently fully recovered without a focal neurological deficit.

Conclusions: Cavernomas of the cauda equina are extramedullary, arise on the inner aspect of the dura, and may be tightly adhered to the nerve roots. To attain gross total excision, the involved nerve may have to be sacrificed; in some cases, this may result in a permanent neurological deficit. Of interest, half of the cauda equina lesions were previously found in patients who had prior radiotherapy; this was not the case in this patient.

Key Words: Cauda equina, cavernous hemangioma, radiation therapy

INTRODUCTION

Cavernous hemangiomas or cavernomas are benign malformations of the vasculature that can occasionally be found in the central nervous system (CNS). The vast majority of CNS cavernous hemangiomas are located supratentorially (69%), primarily in males (58.9%) and in patients in their 40s.

Spinal cavernomas are reported in only 3% of CNS cases. They are typically subdural in their location and are predominantly found in the thoracolumbar spine.¹ Notably, cavernomas of the cauda equina are extremely rare, with only 23 cases reported in the literature. Here, we report the 24th case involving a 77-year-old male.

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CASE DESCRIPTION

A 77-year-old male patient presented with low back pain alone for 3 years duration. He had a history of prostate cancer, skin melanoma, and an MRI-incompatible DDI pacemaker implanted for sick sinus syndrome.

The enhanced lumbar computed tomography (CT) showed an inhomogeneously enhancing intramedullary mass, located at the L3 level, measuring approximately 1.3 × 1 × 1 cm [Figure 1a].

Surgical resection required an L3 hemilaminectomy; the lesion was fully excised. Macroscopically, the tumor was mulberry-shaped, purplish in color, and well demarcated, but tightly adhered to a nerve root of the cauda equina [Figure 2]. Intraoperative monitoring (e.g. somatosensory and motor evoked potentials) indicated that the involved nerve root innervated the anal sphincter and adduction of the toe. Nevertheless, the root had to be sacrificed as it was enveloped by tumor. The histologic examination documented dilated vessels lined by endothelial cells without atypia. Of interest, in the periphery of the mass, neurons of the peripheral nervous system were seen [Figure 3].

Immunohistochemical examination of tumor mass

Immunohistochemical examination of the specimen demonstrated that the endothelial lining was composed of a rich network of cavernous vessels (CD34 – Figure 3b, CD31 – Figure 3c, ERG – Figure 3d). The tissue stained negative for prostate cancer (PSA, PSAP), melanoma (HMB-45, S-100), lymphoma (D2-40), meningioma (EMA, progesterone receptors), and Kaposi’s sarcoma (HHV-8) markers; it also demonstrated a low mitotic index (Ki-67 2–3%, Figure 3e). Rather, findings were consistent with a cavernous hemangioma.

Postoperative course

Postoperatively, the patient’s immediate and 6-month postoperative CT scans documented complete removal of the tumor, and he remained neurologically intact [Figure 1b].

DISCUSSION

Cavernous malformations of the cauda equina are extremely rare and have been reported in only 23 cases.[2] Although most cases are sporadic, a number of gene mutations involving endothelial junctions (particularly of the CCM genes) may contribute to multiple cavernous malformations.[4]

Anatomical and pathological findings

Cavernomas are mulberry-shaped, well-defined, unencapsulated lesions. They are composed of a rich network of dilated sinusoidal spaces lined by a single layer of endothelium which anastomose with each other.[6,8] They exhibit a great infiltrative potential and rarely regress spontaneously. Differential diagnoses should include schwannomas, paragangliomas, ependymomas (including their myxopapillary variant), meningiomas, medulloblastomas, metastases, lymphomas, hemangioblastomas, astrocytomas, and gangliogliomas.[7]

Cauda equina cavernomas

Cauda equina cavernomas are intradural, extramedullary lesions that may be tightly adherent to nerve roots which in some cases may have to be sacrificed to attain a gross total resection.[8] For intramedullary cavernomas, Imagama et al.[3] and others recommended prompt surgical intervention even with mild symptoms to avoid future intratumoral hemorrhage and subsequent neurological injury.

Of interest, Drazin et al.[2] report that slightly less than half of the cauda equina cases (N = 11, 47.8% of all cases) were found in patients who had had prior radiotherapy.
CONCLUSIONS

Here, we report a cauda equina cavernoma in a 77-year-old male who presented with 3 years of low back pain alone. With a history of prostate cancer and skin melanoma, an enhanced CT of the lumbar spine was ordered. The lesion proved to be intradural, extramedullary cauda equina cavernoma which was densely adherent to a nerve root that had to be sacrificed to attain a gross total excision. Postoperatively, the immediate and 6-month enhanced CT studies confirmed no tumor recurrence, and the patient remained neurologically intact.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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