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Article

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The many faces of primary cauda equina myxopapillary ependymoma: clinicoradiological manifestations of two cases and review of the literature

Mohammad Zare Mehrjardi[1,2], Samira Mirzaei[3], Hamid Reza Haghighatkhah[1]

[1]Department of Radiology, Shohada Tajrish Hospital, School of Medicine, Shahid Beheshti University of Medical Sciences, Tehran, IRAN
[2]Section of Neuroimaging, Division of Clinical Research, Climax Radiology Education Foundation, Tehran, IRAN
[3]Department of Radiology, Imam Khomeini Hospital, School of Medicine, Tehran University of Medical Sciences, Tehran, IRAN

Abstract: *Introduction*: Myxopapillary ependymoma (MPE) is a benign and slow growing tumor that originates exclusively from the conus medullaris and cauda equina nervous tissue. It occurs more commonly in young patients. In addition, clinical presentations are non-specific and may mimic benign conditions. Magnetic resonance imaging (MRI) is the imaging modality of choice for evaluating this tumor. We hereby report on two young patients with long-lasting non-specific symptoms, which were clinically attributed to benign conditions. However, cauda equina tumor was revealed on MRI in both patients. *Case report*: Two cases of cauda equina MPE, a 19-year-old female and a 38-year-old male, with different clinical manifestations are reported. The first patient presented with paresis and paresthesia of the lower limbs, and the second patient's complaint was chronic low-back pain and urinary incontinence. In both cases, MRI detected a tumor originated from the cauda equina, and histopathologic examination was compatible with MPE. *Conclusion*: Cauda equina tumors should be considered as a differential diagnosis in the young patients presented with neurologic deficits in the lower limbs, sphincter or erectile dysfunction, and even a simple low-back pain (particularly when symptoms are long-lasting and/or progressive).

Key words: cauda equina, ependymoma, magnetic resonance imaging, myxopapillary ependymoma, radiology

Introduction

Ependymoma is the most common spinal cord tumor in adults, which is more commonly presented during the fourth and fifth decades of life [1, 2]. However, it comprises about 30% of pediatric intramedullary tumors as well [3]. In one study, the mean age of patients with spinal ependymoma was 34.6±16 years-old, and it was slightly more common in males (male-to-female ratio, 4:3) [4].

Ependymomas originate from ependymal cells of the central spinal canal. World Health Organization (WHO) has classified spinal ependymomas into three discrete types, including myxopapillary type or subependymoma (grade-I), classic type (grade-II), and anaplastic type (grade-III) [5].

Although primary tumors of lumbosacral spinal nervous tissue (i.e., conus medullaris, and cauda equina nerve roots) are not common, myxopapillary ependymomas (MPE) originate almost exclusively from this region and comprise about 90% of all tumoral lesions in the lumbosacral canal [6, 7]. Clinical presentations of cauda equina tumors (including MPE) are not specific, and they may mimic benign conditions such as spondylosis or a prolapsed intervertebral disk [8].

MPE often has a peripheral capsule and a lobulated margin on gross examination. Histopathologically, MPE is a heterogeneous tumor due to its mucinous background, and presence of cellular and papillary areas [9, 10]. On immunohistochemical study, tumor cells usually show positive immunoreactivity for glial fibrillary acidic protein (GFAP), cytokeratin AE1/AE3, S100 protein, and D2-40; but they are negative for epithelial membrane antigen (EMA), and brachyury [11, 12].

The mainstay treatment of MPE is complete resection of the lesion, because partial removal may lead to tumor recurrence. Adjuvant chemotherapy and/or radiation therapy are recommended on occasions [4, 8]. Early diagnosis and surgical resection of MPE significantly ameliorates the prognosis of this tumor, because there is the chance of tumor cell dissemination in the cerebrospinal fluid (CSF) leading to multiple disseminated lesions [5].

Magnetic resonance imaging (MRI) is the imaging modality of choice for the diagnosis and evaluation of the cauda equina tumors. However, it is still difficult to determine exact nature and type of the tumor based on the imaging findings [1, 9].

We, hereby, present two cases of cauda equina MPE, and discuss the clinical and neuroimaging findings of MPE. It is important for the clinicians and radiologists to have a detailed knowledge of the clinical and radiological presentations of the cauda equina tumors for making a correct diagnosis, and managing the patient properly.

Case report

Informed written consent was obtained from both patients for reporting and disclosing their clinical data with respect to ethical issues.

Case 1:

A 19-year-old female presented with a five month history of gradually progressive paresis and paresthesia of the lower limbs. She did not have any sphincter dysfunction. On physical examination, both lower extremities muscle strength and patellar tendon reflex had been reduced to the grade 3/5 and 1+, respectively. Muscle tone of both legs was slightly reduced. Achilles and plantar reflexes as well as proprioceptive sensation were normal. Biochemical examinations revealed no abnormality. Based on the clinical data, the first impression was a prolapsed intervertebral disk, and the patient was referred for lumbosacral spine MRI.

On MRI (Figure 1), there was an intradural extramedullary mass in the lumbar spinal canal at the level of L2–L3 intervertebral disk, which was originated from the cauda equina. It was isointense compared to the normal cord and slightly hyperintense compared to the CSF on T1-weighted, and markedly hyperintense on T2-weighted images. The tumor revealed significant homogeneous post-contrast enhancement following gadolinium-based contrast agent injection. Based on MRI findings, differential diagnoses were nerve sheath tumors, MPE, and less likely meningioma. Histopathological examination following complete surgical tumor removal demonstrated benign ependymal cells with pseudopapillae formation and redundant mucin production in a myxoid stroma, suggestive of MPE.

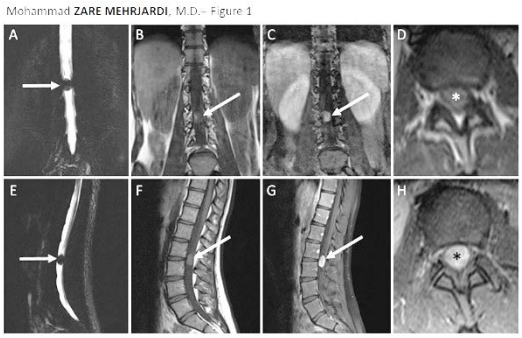

Figure 1 - Lumbosacral spine MRI of a 19-year-old female. On coronal (A to C) and sagittal (E to G) sections, a well-defined oval mass is noted within the lumbar spinal canal on the cauda equina nerve roots, at the level of L2–L3 intervertebral disk. It is seen as a filling defect on heavily T2-weighted myelograms (arrows in A and E), and has slightly high signal intensity compared to the surrounding cerebrospinal fluid, and iso-signal intensity compared to the normal cord on T1-weighted images (arrows in B and F). The lesion is homogeneously enhanced on post-contrast T1-weighted with fat saturation images (arrows in C and G). Post-contrast enhancement is also evident comparing axial pre- (asterisk in D), and post- (asterisk in H) contrast injection fat saturated T1-weighted images

Case 2:

A 38-year-old male presented with a 2 year history of low-back pain and mild paresthesia of the lower limbs, as well as a recent urinary incontinence. On physical examination, lower limb muscle strength and tone, as well as deep tendon reflexes were normal on either sides. The initial clinical impressions were congenital spinal canal stenosis, intervertebral disk herniation, and spondylolisthesis. The patient was referred for additional evaluation by lumbosacral spine MRI.

MRI study (Figure 2) demonstrated an intradural extramedullary mass in the sacral canal, which had slightly expanded the canal at

S1–S3, and remodeled posterior aspects of the corresponding vertebral bodies. It was hypointense compared to the normal cord and isointense compared to the CSF on T1-weighted, and markedly hyperintense with internal foci of low signal intensity representing previous episodes of hemorrhage on T2-weighted images. The tumor showed extensive post-contrast enhancement following gadolinium-based contrast agent injection. According to these imaging findings, differential considerations were MPE, paraganglioma, and astrocytoma. Histopathological examination after surgical resection was compatible with MPE.

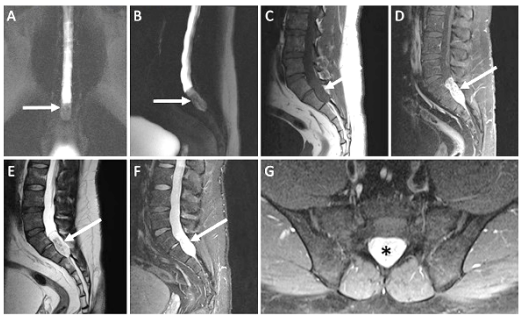

Figure 2 - Lumbosacral spine MRI of a 38-year-old male. On coronal (A), and sagittal (B) myelograms, a filling defect (arrow) with obstruction of the cerebrospinal fluid flow, and mild canal expansion, is noted at the sacral level (S1–S3). The mass is hypointense on sagittal T1-weighted image (arrow in C), and shows heterogeneous post-contrast enhancement on sagittal fat saturated T1-weighted image (arrow in D). The lesion is hyperintense with internal foci of hypointensity (due to previous hemorrhage) on sagittal T2-weightd (arrow in E), and turbo inversion recovery magnitude (arrow in F) images. Heterogeneous post-contrast enhancement with mild canal expansion is also noted on axial contrast-enhanced T1-weighted with fat saturation image (asterisk in G)

Discussion

MPE is a benign and slow growing tumor that exclusively originates from cauda equina nerve roots and/or conus medullaris. It affects young adults and males more common than children and females [1, 10]. Common clinical manifestations of this tumor include chronic low-back pain, paresthesia and flaccid paralysis of the lower limbs, and dysfunction of the rectal and urinary sphincters in both genders, as well as erectile dysfunction in males. Because the clinical presentations are non-specific and the patients are commonly young, there is usually delay in the diagnosis and treatment [7, 8]. Although low back pain etiologies are usually benign in nature and neoplastic conditions comprise less than 1% of all etiologies, neoplasms should be considered as a differential diagnosis because they are associated with significant morbidity and mortality [13].

Ngo et al. [13] reported a 19-year-old male with a 3 year history of low back pain, which was attributed to a soft tissue injury and he was prescribed anti-inflammatory drugs. Two years later, he developed intermittent erectile dysfunction and difficulty in defecation. Finally, a large lumbosacral spinal tumor was diagnosed on MRI four years after his first presentation at the age of 23 years. Surgical excision was performed and histopathologic examination was compatible with MPE. Similarly, Demir et al. [14] reported MPE as the cause of chronic low back pain in a 28-year-old female.

MRI is the imaging modality of choice for precise evaluation of these patients. MPE is

commonly iso to hypointense on T1-weighted, and hyperintense on T2-weighted images [9, 15]. In a study on 20 cases with MPE, MRI features of the tumor were not specific. Tumors were isointense (75%), hypointense (19%), or hyperintense (6%) compared to the normal cord on T1-weightd images, while they were hyperintense (80%) or isointense (20%) on proton-density, and hyperintense (100%) on T2-weighted images [7]. Although this tumor is almost always hyperintense on T2-weighted images (due to the mucinous content), areas of low signal intensity secondary to previous hemorrhage may be evident within the tumor. Almost all MPEs enhance avidly following contrast agent injection. Post-contrast enhancement is usually homogenous, but heterogeneous enhancement due to previous hemorrhage and cystic changes is not uncommon [9, 15].

Although MRI can accurately demonstrate presence of a mass in the spinal canal, it has currently limitations in determining the exact type of the tumor. Other benign as well as malignant tumors that may occur in this region include nerve sheath tumors such as schwannoma and neurofibroma, paraganglioma, hemangioblastoma, astrocytoma, primitive neuroectodermal tumor (PNET), ganglioglioma, meningioma, metastasis, lipoma, dermoid, and epidermoid.

Bavbek et al. [16] reported a 46-year-old man with a 2 year history of progressive paresthesia and paresis of the right leg. Lumbar spinal MRI showed an intradural extramedullary mass with homogenous post-contrast enhancement at L1–L2. Initial radiological differential considerations were schwannoma, neurofibroma, and meningioma. However, postsurgical histopathological examination identified the tumor as a MPE. In another report by Kim et al. [17], a 30-year-old male with a 1 year history of low back pain and leg pain was evaluated by MR imaging. MRI revealed a well-defined intradural extramedullary tumor with significant homogenous post-contrast enhancement and cystic changes at L3–L4 level, which was initially diagnosed as a nerve sheath tumor based on imaging findings. Histopathological examination after resection was, however, compatible with MPE. On the other hand, Thoriya et al. [18] reported a case of primary PNET of lumbosacral spine in a 31-year-old female with a 4 week history of progressive low back pain and weakness of lower limbs, which was primarily diagnosed as MPE based on MRI findings.

Conclusion

Although MPE is a rare tumor, it consists the most common neoplasm of the cauda equina nerve roots. It occurs more commonly in the young patients and causes non-specific clinical manifestations; therefore, it should be considered as a differential diagnosis in any young patient with a history of chronic low-back pain, paresis or paresthesia of the lower limbs and sphincter or erectile dysfunction, particularly when the symptoms are progressive and do not respond to the conservative management. MRI is the imaging modality of choice for further evaluation of these patients. It may be of help to make differentiation between benign and malignant conditions, but there is currently a significant

overlap between the imaging findings of cauda equina tumors. MRI is also helpful to determine boundaries and extensions of the tumor for preoperative planning.

Correspondence
Mohammad Zare Mehrjardi
Contact no.: +989133530656
E-mail: zare@sbmu.ac.ir
Mailing address: Department of Radiology, Shohada Tajrish Hospital, Tajrish SQ., Tehran 1989934148, Iran

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