Myxopapillary Ependymoma (MPE)

Astrid Sacasa MD, MPH; Susana Ferra M.D.

Pathology, HCA Florida Westside Hospital

FLORIDA SOCIETY OF PATHOLOGISTS

HCA Florida Westside Hospital

Introduction

Myxopapillary ependymoma is a slow growing central nervous system tumor located almost exclusively in the conus medullaris and filum terminalis. It is a rare tumor with an incidence rate of 0.6 – 1.0 cases per 1 million person-years. Adults are more commonly affected but can occur in all ages. The most common clinical features are lower back pain, sometimes associated with sciatica, sensorimotor deficits, and fecal/urinary incontinence.

Here, we describe a case of Myxopapillary Ependymoma that was unrecognized for three years despite the classic presentation. We discuss the immunohistochemical features in support of this diagnosis and bring awareness to this tumor that although uncommon, should be considered among the differential diagnoses of recurrent sciatic pain.

Methods

A 38-year-old male with a three-year history of intermittent but progressive and severe lower back pain radiating to the legs with associated bilateral numbness. He has no other significant medical history and has been treated in the past with analgesic and anti-inflammatory medications with temporary resolution of symptoms. He presented to our hospital due to bladder incontinence two days prior in addition to back pain not responding to his usual treatment. He denied any numbness in the groin or bowel incontinence. Admission evaluation included a lumbar MRI with contrast showing an oval shaped intradural extramedullary mass at the conus medullaris (Image 1), which was subsequently resected and sent to pathology for evaluation.

Results

Microscopic examination of hematoxylin and eosin (H&E) stained slides at low 4X (Figure 1) and high-power 40X (Figure 2) magnification show cuboidal to elongated basophilic tumor cells arranged in a papillary fashion around myxoid material radiating from blood vessels. The myxoid material is highlighted with PAS (Figure 3) and Alcian blue positive stains (Figure 4). Mucicarmine stain was negative. The tumor is positive for GFAP (Figure 5), vimentin and focal S-100 immunohistochemical (IHC) stains; while negative for EMA (Figure. 6), CK-cam5.2, CK-7, CK-20, and pankeratin. The immunohistochemistry in addition to the morphology, clinical history and location, supported the diagnosis of myxopapillary ependymoma, WHO Grade 2 (CNS WHO 5th edition).

Images

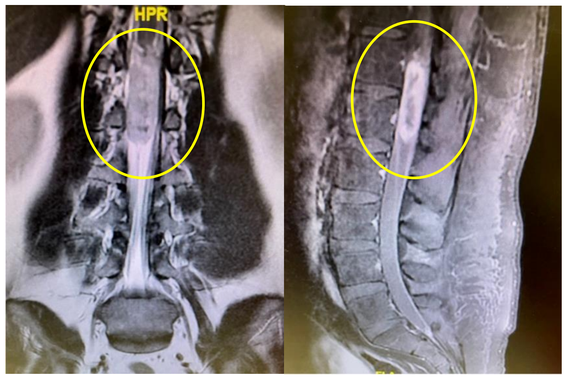

Image 1. coronal (left) and sagittal (right) views

Results (Continued)

Figures

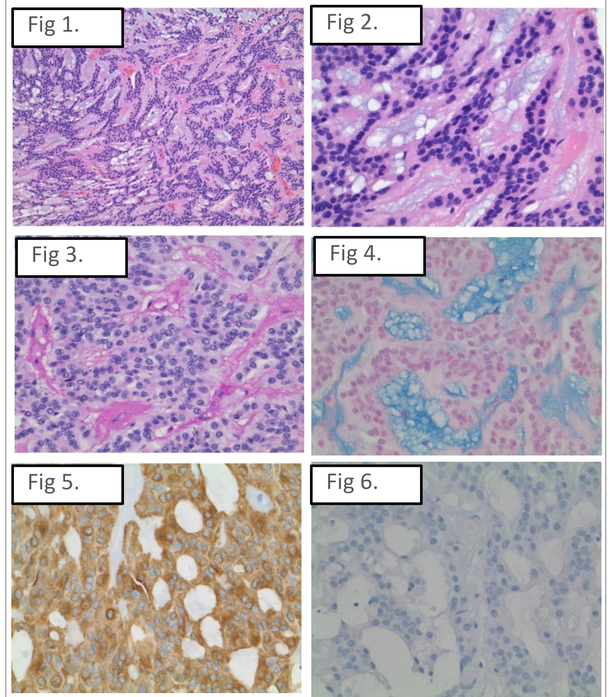

Table 1.

Diagnostic criteria for myxopapillary (CNS WHO 5th ed.)
Essential: Glioma with papillary structures and perivascular myxoid change or at least focal myxoid microcysts
AND: Immunoreactivity for GFAP
[AND (for unresolved lesions): DNA methylation profile aligned with myxopapillary ependymoma]
Desirable: - Papillary arrangements of tumor cells around vascularized fibromyxoid cores - Location in the filum terminale or conus medullaris

Conclusions

Although a rare entity, myxopapillary ependymomas should be considered in the diagnostic work-up of persistent sciatic pain. MRI is the best imaging technique, showing hyperintensity in T1 weighted images. Grossly, MPE's are well defined, soft and pink to tan-grey sausage shaped lesions, with a smooth surface. Microscopic features include many small papillary structures surrounded by cuboidal to elongated cells, round nuclei, and abundant basophilic myxoid material around blood vessels. On IHC, they are GFAP and Vimentin positive, S100 focally positive, and cytokeratin negative, helpful in excluding other tumors in this location including paragangliomas, choroid plexus tumors, chordomas, chondromas, chondrosarcomas, and schwannomas.

MPE's are distinctly designated as WHO Grade 2 tumors. A summary of the diagnostic criteria is provided in Table 1. Recognizing MPE's compared to other forms of ependymomas or tumors in the same location is of significant clinical importance due to their good prognosis after radical surgical resection with a 10-years overall survival rate of over 90%.

References & Acknowledgements

Reference A:
Limaiem F, M Das J. Myxopapillary Ependymoma. PubMed. Published 2022. Accessed September 13, 2022. https://www.ncbi.nlm.nih.gov/books/NBK559172/

Reference B:
Rosenblum M. Myxopapillary Ependymoma. WHO Classification of Tumors online. Central Nervous System Tumors (5th edition). https://tumourclassification.iarc.who.int/chaptercontent/45/52. Accessed September 13, 2022