

Magnetic Resonance Imaging in a case of myxopapillary ependymoma in a young female.

Dr.Sanjana Shetty,Dr.Rajesh Venunath,Dr.Raghuraj U,Dr.Lathika Shetty.

* Department of Radiodiagnosis, K.S Hegde Medical Academy, Nitte University, Deralakatte Mangalore, Karnataka – India

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Abstract- In children, tumours of the spine are much rarer than intracranial tumours. They are classified into intramedullary, intradural and extradural tumours. Magnetic resonance imaging provides crucial information regarding the extent, location and internal structure of the mass thus critically narrowing the differential diagnosis. Our department reports a case of a 16 year old female who presented with history of proximal muscle weakness and MR features of the above disease. We discuss the typical MR features of myxopapillary ependymoma and determine the role of MR imaging in identifying the tumour, establishing its extent and defining its relationship with adjacent intraspinal structures.

Index Terms- Myxopapillary ependymoma,,MRI ,Intramedullary,Syringomyelia.

I. INTRODUCTION

Myxopapillary ependymomas are highly vascular tumours arising almost exclusively in the dorso lumbar region and produce symptoms that can mimic discogenic pathology. Ependymomas constitute approximately 8% of all intracranial gliomas in children and 63% of primary intraspinal gliomas.³Myxopapillary ependymoma is a distinct pathological sub type that occurs almost exclusively in the conus medullaris

and filum terminale; it accounts for a majority of ependymomas in this region.²

Patients clinically present with back pain, radiculopathy, paraparesis, bowel and bladder disturbances with a peak age incidence of 30-40 years however has been reported at all ages with a predilection in males.

Case history:

A 16 year old female presented to the department of paediatrics with history of dysphagia ,pneumonia and proximal muscle weakness.

MRI:

MRI brain with gadolinium was performed in a Siemens Magnetom Avanto 1.5 Tesla scanner.

The study reveals a well defined intra medullary altered signal intensity lesion in the region of the conus measuring around 6.9cmx1.3cm with widening of the spinal canal. The lesion shows intermediate signal intensity on T1 and T2 and appears hyperintense on STIR sequences with a hypointense rim noted on T2 weighted sequences in the lower margin of the lesion. Post contrast study shows intense enhancement. An extensive intra medullary CSF signal intensity cavity with septations noted proximal to the lesion extending upto C2 level. These features are consistent with myxopapillary ependymoma with syringomyelia.



Figure 1: Shows an intensely enhancing intra medullary lesion on post contrast.

Figure 2 shows an intra medullary signal intensity cavity extending proximal to the lesion upto the level of C2.



Figure 3 shows the lesion with altered signal on T2 sequences with a hypointense rim.

Figure 4 shows a hyperintense signal intensity lesion on STIR sequence.

Histopathological findings:

The patient underwent a partial excision biopsy of the lesion and the histopathological evaluation revealed tissue composed of tumor cells arranged in papillary pattern. The cells were cuboidal to elongated radially arranged around vascularised

and myxomatous stroma cores. The cells are also seen in clusters in a loose myxoid matrix. The nucleus is mildly enlarged, with fine chromatin. Areas of hemorrhage seen. Features suggestive of myxopapillary ependymoma –conus medullaris WHO Grade I.

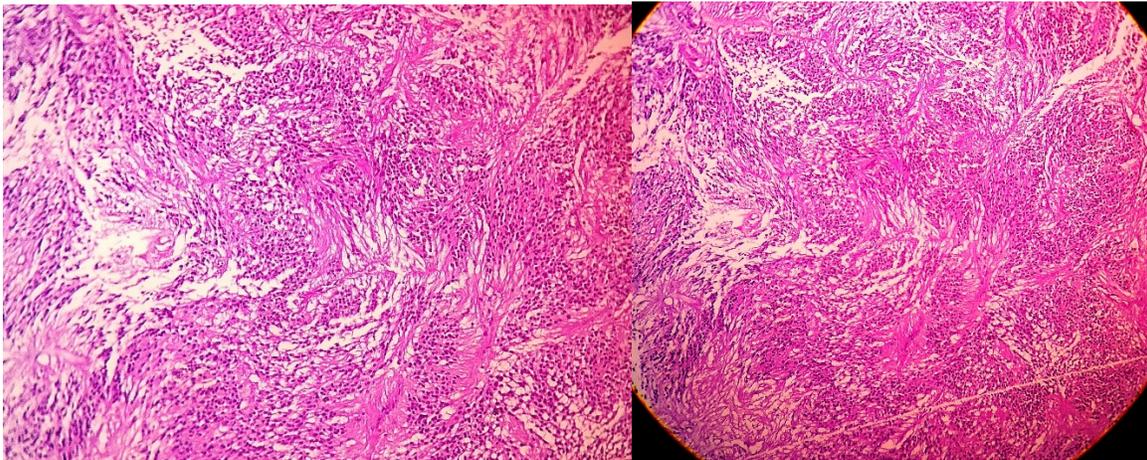


Figure 5 and 6: Slides were air dried and fixed in alcohol and stained by Giemsa and Papinicolaou stain respectively. Smears were highly cellular showing cuboidal and columnar cells with surrounding myxoid material.

II. DISCUSSION

Ependymomas are the most common intra medullary spinal cord tumours in adults. Even so, they are quite rare with only about 227 cases in the United States each year. The two most common ependymoma subtypes are cellular and myxopapillary ependymomas. Cellular ependymomas can arise anywhere however they usually occur in the cervical cord whereas myxopapillary ependymoma occurs almost exclusively in the conus medullaris and filum terminale⁵

Patients with these tumours typically present with low back pain which may or may not be associated with sciatica. Other symptoms, such as sensorimotor disturbances and bowel and bladder dysfunction are much less common⁵

At gross examination, myxopapillary ependymomas are soft, vascular, lobular or sausage-shaped masses which are often encapsulated and may show haemorrhagic or mucinous degeneration. The site of these tumours are usually within the

filum terminale but may also extend and incorporate the conus medullaris. Myxopapillary ependymomas may grow to a quite large size, filling and expanding the spinal canal. Tumours located around the sacrum can cause bone destruction, although such cases are rare⁵

Radiographs of myxopapillary ependymomas are usually normal but may show a widened spinal canal or bone destruction. Myelograms may demonstrate a well defined intradural mass at the conus medullaris⁵

On unenhanced CT scans, myxopapillary ependymomas are typically isoattenuating to the spinal cord. Small tumors produce nothing more than non-specific canal widening whereas larger tumors may produce scalloped vertebral bodies, neural foraminal enlargement. On contrast-enhanced CT scans, myxopapillary ependymomas typically demonstrate intense homogenous enhancement.⁵

The MRI features of myxopapillary ependymoma are non-specific. Tumors are usually isointense relative to the spinal cord on T1-weighted images and hyperintense on T2-weighted

images. At times they can be hyperintense relative to the spinal cord on un-enhanced T1-weighted images due to proteinaceous mucoid matrix. This is a feature which may be useful in distinguishing myxopapillary ependymoma from other ependymoma subtypes which are virtually always hypo- or isointense on T1-weighted images⁶. Myxopapillary ependymomas enhance intensely after administration of contrast material. Enhancement is homogenous but may be heterogenous when haemorrhage or necrosis is present. MRI may also show expansion of the spinal canal and neural foramina, bone destruction and surrounding soft tissue invasion.

The differential diagnosis for small myxopapillary ependymomas in the conus medullaris and filum terminale includes schwannoma and sub ependymoma, both of which can mimic features almost similar to myxopapillary ependymoma. The differential diagnosis also includes astrocytoma, hemangioblastoma, ganglioma paraganglioma and other ependymoma subtypes.⁵ For large ependymomas that cause sacral destruction the differential diagnosis should include sacral tumours such as aneurysmal bone cyst, chordoma, plasmacytoma, metastasis and giant cell tumour.⁵

Conclusion

Ependymoma is the most common intra medullary spinal tumour, however they are quite rare with only 227 intra dural spinal ependymomas diagnosed in the United states each year. MR imaging is valuable in identifying the extent of thoracolumbar myxopapillary ependymomas and in defining the relationship to intraspinal structures. The MR findings in myxopapillary ependymoma are nonspecific, however the diagnosis can be suggested by a large, intensely enhancing, intradural extramedullary thoracolumbar mass that extends for several vertebral levels. Myxopapillary ependymoma should also be included in a differential diagnosis of a post sacral mass.

Imaging protocols should examine the entire dorsolumbar region and include IV contrast material administration.²

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AUTHORS

- First Author** – Dr. Sanjana Shetty. Junior resident. K.S Hegde Medical Academy.
Second Author – Dr. Rajesh Venunath, Assistant professor. K.S Hegde Medical Academy.
Third Author – Dr. Raghuraj. Associate professor. K.S Hegde Medical Academy.
Fourth Author – Dr. Lathika Shetty. Professor .K.S Hegde Medical Academy.