Ependymoma presenting as a paravertebral mass

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Citation

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Abstract

We present a case report of a 30 year old female patient who presented with progressively increasing pain lower back and right lower limb. On radiological investigations she was found to have a well-defined dumbbell shaped mass lesion with both intracanalicular and a large extracanalicular component with expansion of the associated neural foramen at L4-5 level on right side. It revealed a well defined hypointense rim on all pulse sequences. Debulking of the tumor was done followed by radiotherapy. On histopathology it was diagnosed as Myxopapillary ependymoma.

INTRODUCTION

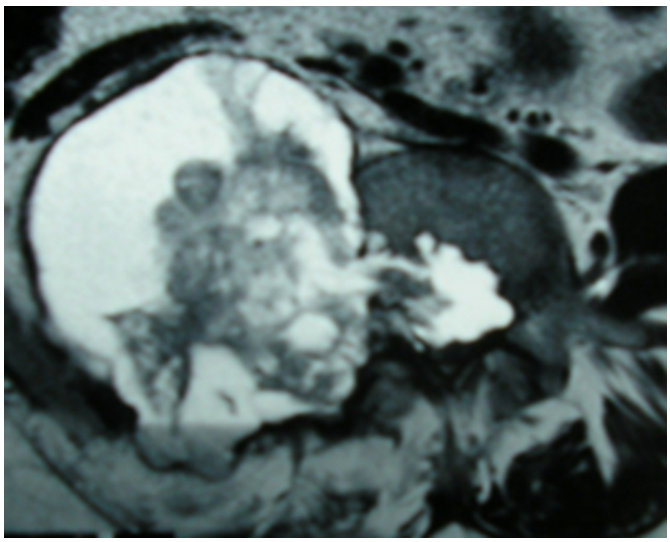
Ependymomas are relatively uncommon neoplasms, comprising about 5% of all neurogenic tumours, [1] 60% of glial spinal cord tumors overall and 90% of primary tumors in the filum terminale and cauda equine. [2] Ependymomas arising outside the central nervous systems are uncommon and when they do occur are usually seen in the sacrococcygeal region. Other sites including the pelvis, spinal nerve roots, lung, etc have also been reported. [3]

CASE REPORT

A 30 year female patient presented with progressively increasing pain lower back on right side with radiation to right lower limb of 06 month duration and parasthesia of 02 month duration. She took analgesics off and on for the pain. On presentation there was no significant weakness or involvement of bladder and bowel. All hematological parameters were normal. X-ray showed an expansile osseous lesion in the region of L5 pedicle abutting the vertebral body

Figure 1

Figure 1: Axial T2 weighted MRI image revealing a dumbbell tumor with intracanalicular as well as a large extracanalicular component expanding the ipsilateral neural foramen.

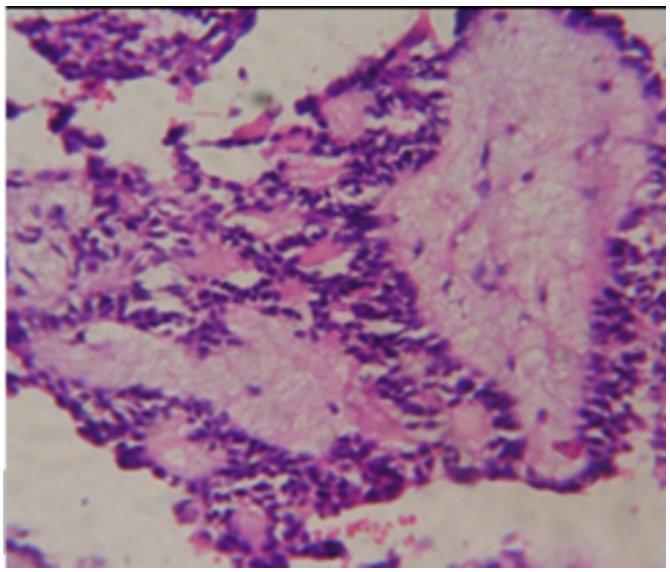


Magnetic resonance imaging (MRI) revealed a large well-defined dumbbell shaped tumor with both intracanalicular and a large extracanalicular component at L4-5 level on right side. There was expansion of the associated neural foramen. It was hypointense on T1 weighted images and predominantly hyperintense on T2 weighted images with diffuse enhancement on contrast enhanced images. It revealed a well-defined hypointense rim on all pulse sequences. In addition hypointense lesions were seen on all pulse sequences within the tumor extending from the vertebral canal. (Fig 1) Patient was operated through right retroperitoneal approach. Debulking of the mass lesion was

performed and histopathology revealed papillae containing large amount of myxoid material and lined by small cuboidal ependymal cells suggestive of myxopapillary ependymoma (Fig 2). After surgery, the tumor site was radiated with 30 cycles radiation with total dose 50 Gray. At follow up 01 year later patient was asymptomatic except for mild foot drop on right side.

Figure 2

Figure 2: Photomicrograph shows papillae containing large amount of myxoid material and lined by small cuboidal ependymal cells. (H&E X 100)



DISCUSSION

Myxopapillary ependymoma is a distinctive subtype of glioma that occurs almost exclusively in the filum terminale and conus medullaris accounting for nearly 90% of primary tumors in the cauda equine region. The peak age incidence is the fourth decade. They arise from ependymal cells lining the central canal or its remnants, and from cells of the ventriculus terminalis in the filum terminale. [2] Less commonly, myxopapillary ependymoma may occur outside the central nervous system from direct metastatic extension of an intrathecal tumor, as a primary tumor outside the thecal sac or as extraneural metastasis. Ependymomas as a primary tumor outside the central nervous system have been reported to occur as primary sacral, pelvic, abdominal, primary tumor of the skin and subcutaneous tissue of the sacrococcygeal area, [4] posterior medistinum [5] and primary intracranial tumor. [6] Moser FG et al [7] and Wilson RW et al [8] postulated the presence of ectopic ependymal tissue to explain the origin of these neoplasms. On reviewing the literature, they found that 20 to 30% of ependymomas in ectopic locations were associated with spina bifida occulta, indicating developmental failure that resulted additionally in ectopic ependymal rests. However they found no associated anomaly in their patient. [7]

Myxopapillary ependymoma is a low grade glioma. They present as highly vascular soft tissue tumors consisting of cellular elements in a papillary pattern, distinctively abundant supporting fibrous connective stroma, with foci of mucinous degeneration and mucin secretion by tumour cells. [2] The appellation “myxopapillary” is based on the tendency of this ependymoma to produce mucin and form papillae by virtue of its arborizing vasculature, replacing the architecture of the normal filum terminale. [9] The biologic behaviour of myxopapillary ependymomas is relatively benign compared with ependymomas which occurs in the spinal cord itself or in the brain, but the local recurrence is significant if the lesion is incompletely excised. [10] Extraneural metastases in the lungs, the pleura, the liver, and the thoracal and abdominal lymph nodes have been reported. [11]

Typically, they are grossly well circumscribed and on T1-weighted MR images they are homogenous and isointense, and on T2-weighted images they are homogenous and hyperintense. [12] Intense enhancement after gadolinium injection is characteristic. The presence of focal necrosis, cystic degeneration, or hemorrhage may result in an inhomogenous lesion with mixed signal intensity. [10]

Spinal nerve root ependymoma is a rare lesion; however they have been described in the literature. We believe that this is the first reported case of Spinal nerve root ependymoma presenting as a dumb-bell tumor with a large paravertebral component. Differentiation of this mass from neurofibroma may be difficult delaying diagnosis.

Surgery aims at complete removal of the mass; this is usually possible with neurofibroma, but in about half of the patients with ependymoma it may not be feasible, and radiotherapy will then be given. The prognosis is usually good, with symptomatic recurrence in only one-third of ependymomas incompletely removed. [14]

CONCLUSION

Extraneural ependymoma is rare. Various sites such as pelvis, mediastinum, sacrococcygeal region, spinal nerve roots have been described. However, its presentation as a dumbbell shaped lesion with a large paravertebral component has not been described in the literature to the best of our knowledge.

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