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MYXOPAPILLARY EPENDYMOMA: CASE REPORT, LITERATURE REVIEW AND RECENT UPDATES

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Abstract

Spinal myxopapillary ependymoma (MPE) arises from the conus medullaris, cauda equina, and filum terminale. MPEs originate from ependymal glial cells that border the central spinal canal and are mostly benign. We present a case report of MPE of filum terminale in a 27 year male patient diagnosed based on histopathology. Brief overview of the clinical features of the lesion with treatment options and prognosis is also presented. MPE occurs primarily in young people, with a slightly increased male preponderance. Pediatric patients with spinal MPE report a greater risk of recurrence following resection and a more aggressive expression. Gross complete resection is the primary treatment for spinal MPE because it yields the best results and reduces the risk of tumor spread and recurrence. Factors, including large tumor size, incomplete tumor resection, multifocality, sacral spine involvement, dural perforation by the tumor, intraoperative blood loss of more than 500CC, and longer operation duration more than three hours, are associated with the recurrence and prognosis of MPE. The general prognosis for the tumor is favorable.

INTRODUCTION

A distinct variant of ependymoma that arises from the conus medullaris, cauda equina, and filum terminale is called a spinal myxopapillary ependymoma (MPE) [1]. They originate from ependymal glial cells that border the central spinal canal and are mostly benign, slowly developing tumors [2]. MPE is classified as a grade 2 tumor in the latest World Health Organisation (WHO) classification system [3].

It differs from other ependymoma subtypes in both macro and microscopic morphology [4]. With an estimated frequency of one per million persons, spinal MPE makes up 1 to 5% of all spine tumors [5]. Men are diagnosed with the condition more frequently than women, usually in the age range of 20 to 40. Nonetheless, 8–20% of all instances of spinal MPE occur in the pediatric population [6].

Due to the tumor's slow growing nature, the symptoms typically appear months or years before the diagnosis, depending on the size, location, and extent of the tumor [7]. Gross complete resection is the best course of treatment for spinal MPE; however, in cases where this is not possible, subtotal resection plus adjuvant radiation therapy is an alternative. Although there is a chance of metastasis and recurrence, spinal MPE often has a good prognosis [7, 8].

CASE REPORT

A 27 year male patient presented with chronic lower backache and sciatica for the past one year, worsening over the past 3 months. Radiological evaluation showed Well circumscribed, sharply demarcated, ovoid, contrast enhancing mass in the filum terminale region. The lesion was Hyperintense on T1 and T2 weighted MRI with intense contrast enhancement.
Surgical excision of the lesion was done. Gross examination revealed an encapsulated tan gray lesion which was soft in consistency. Microscopic examination revealed

radial arrangement of cuboidal to epithelioid elongated glial tumor cells around hyalinized fibrovascular cores in a papillary configuration [Figure 1]. Accumulation of basophilic myxoid material around blood vessels (myxoid stroma) and in microcysts was noted [Figure 2].

DISCUSSION

Primary central nervous system tumors known as ependymomas are divided into four groups according to their pathological characteristics: anaplastic ependymoma, conventional ependymoma, myxopapillary ependymoma, and subependymoma [9]. The World Health Organisation (WHO) previously categorized MPE as a grade 1 tumor; however, in the revised WHO 2021 classification, MPE is regarded as a grade 2 tumor [10].

It frequently appears in the cauda equina and filum terminale. More research is required to determine the etiology or pathophysiology behind the myxopapillary architecture [11, 12].

The histology of the MPEs is distinct from that of the other subtypes. The distinctive ependymal rosettes and perivascular pseudorosettes on microscopy, that are indicative of ependymomas, are often absent from the myxopapillary group [13].

However, vascular parietal hyalinization is frequently seen in MPEs, and the mucoid stroma is a characteristic feature of the myxopapillary subtype [14, 15]. It is not common to see calcification, necrosis, cyst development, or mitotic figures [16].

Clinically, MPE occurs primarily in young people, with a slightly increased male preponderance [17]. Symptoms of spinal MPE can include discomfort, weakness, and altered sensation. Depending on the location of the injury and the extent of compression of the spinal cord or nerve roots, symptoms of sexual, bowel, or urine dysfunction may be evident [18].

Spinal MPE is more common in adults than in children, and it usually affects the lumbar spine (52.9%), with the lumbosacral area being involved in about (25%) of cases [19]. However, cerebral ventricles or the brain are seldom the site of MPE, and the majority of cases occur in children [20].

The median age at diagnosis for females was 45 years, which was substantially higher than for males. In 94% of instances, the lumbar region was the most common site of the tumor, and in 88% of cases, the tumor was typically located at the level of L1, L2, or L3 [21].

Pediatric patients with spinal MPE report a greater risk of recurrence following resection and a more aggressive expression. As opposed to adults, several research studies have been done on MPE in children, including the impact adjuvant radiation treatment plays in tumor growth or recurrence [22].

Multifocal spinal MPE is not common, and the theory behind these variations is not fully understood [23]. Numerous perspectives, however, indicate that lumbosacral spine involvement is more likely in pediatric MPE, which puts it at risk for lymphatic metastases and dissemination [24].

Furthermore, some have proposed that pediatric MPE is histologically diverse, which accounts for the differences and is unrelated to the tumor's anatomical location [25].

Another reason for the delay in presentation and diagnosis is the challenging nature of pediatrics as a population to interact with.

To address and comprehend the distinction between MPE in adults and pediatrics, as well as the causes of primary spreading of MPEs, more investigation and study are needed [26].

Gross complete resection is the primary treatment for spinal MPE because it yields the best results and reduces the risk of tumor spread and recurrence [27]. Subtotal resection carries a 30% chance of tumor recurrence rate within an average of 3.5 years [28]. Tumor capsular disruption has also been linked to recurrence; the risk might increase to 45% after subtotal resection and 15% after gross complete resection [29].

Adjuvant radiation is advised in cases of capsular breach, irrespective of the amount of resection, to lower the risk of metastasis or recurrence. Overall, there is ongoing debate regarding the benefits of adjuvant radiation therapy for spinal MPE.

While some studies indicate that the adjuvant radiation may reduce the risk of recurrence, particularly in cases involving subtotal resection, a retrospective study found no such benefit following gross total or subtotal resection.

Adjuvant radiation therapy is not advised, although, in situations when gross complete resection was accomplished without capsular breach, primarily if postoperative CSF examination revealed no signs of dissemination [30].

The recurrence rate of adult MPEs is around 32% after subtotal resection and 10–20% with complete excision [31]. In children, the tumor is linked to a more aggressive nature and a greater frequency of being multifocal; as a result, there is a higher and potentially 40% chance of recurrence [32].

Numerous factors, including large tumor size, incomplete tumor resection, multifocality, sacral spine involvement, dural perforation by the tumor, intraoperative blood loss of more than 500CC, and longer operation duration more than three hours, are associated with the recurrence and prognosis of MPE [1].

The general prognosis is favorable, with 90% of patients experiencing a satisfactory outcome without a severe neurological disability, despite the possibility of tumor spread and recurrence. But the long-term prognosis has not been well researched, necessitating adequate and prolonged follow-up [33].

CONCLUSION

Myxopapillary ependymoma is an ependymal cell tumor that develops predominantly in the conus medullaris, cauda equina, or film terminals. Patients with this slow-growing tumor are often in their third decade of life, with a slightly increased preference for men.

The size and location of the tumor influence the clinical presentation. In adults, MPE seldom manifests as a multifocal lesion and is linked to an increased likelihood of spread and recurrence. The mainstay of therapy is gross complete resection. Adjuvant radiation and subtotal resection are available alternatives. The general prognosis for the tumor is favorable.

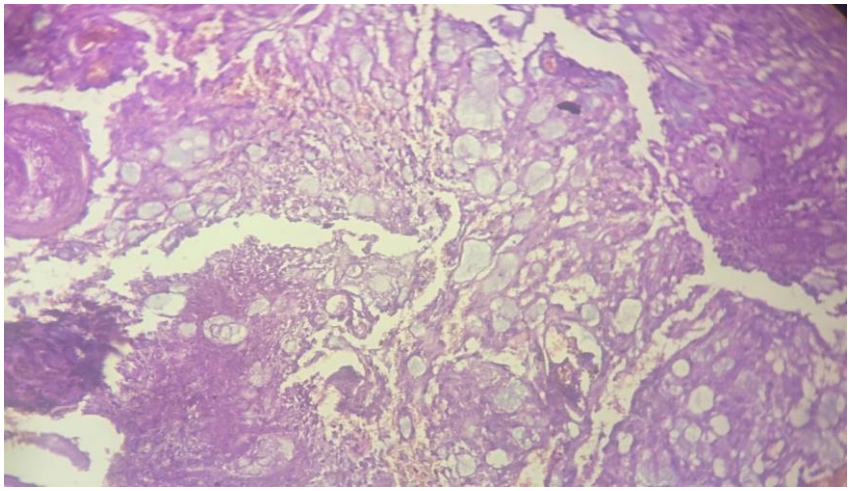

Figure 1: glial tumor cells around hyalinized fibrovascular cores in a papillary configuration

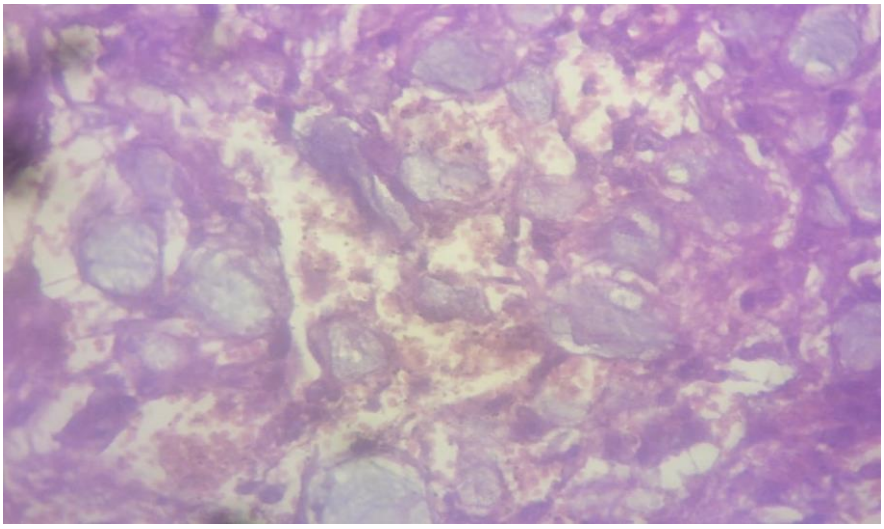

Figure 2: Accumulation of basophilic myxoid material around blood vessels (myxoid stroma)

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