CASE REPORT

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Myxopapillary Ependymoma–a Case Report of Rare Multicentric Subtype and Literature Review

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ABSTRACT

Background: Myxopapillary ependymoma is a rare type of primary spinal tumor, it is distinctly a slow-growing tumor that originates in the conus medullaris, cauda equina, or film terminals and is rarely identified as a multicentric type. Myxopapillary ependymoma has a unique histological characteristic and is associated with a generally better prognosis. **Objective:** We present a case of a rare multicentric myxopapillary ependymoma. **Case presentation:** A 28-year-old male with 1-year history of low back pain and 3 months of radiating pain to left lower limb with perianal anesthesia. Magnetic resonance imaging (MRI) exhibited a large intradural intramedullary lesion from the level of the conus medullaris extending to the filum terminals at the level of T12 to L3 with smaller multiple enhancing lesions seen opposite to L4 and L5 level as well as within the exiting nerve roots, at the left side of L1/L2 and L2/L3 and right side of L3/L4 and L5/S1 level. The patient underwent surgical resection with significant improvement in symptoms and no tumor progression on follow up MRI scan. **Conclusion:** We hereby present a case of multicentric myxopapillary ependymoma with a literature review of the previous reported cases. We believe that our study will make a significant contribution to the literature and will be of interest to the readership regarding of the rarity of multicentric Myxopapillary ependymoma and it will help in decision making for the proper surgical Intervention on these kinds of cases.

Keywords: Multicentric ependymoma, Multifocal ependymoma, Myxopapillary ependymoma, Ependymal tumor.

1. BACKGROUND

Spinal myxopapillary ependymomas are a distinct variant of ependymomas that originates from the conus medullaris, cauda equina, and film terminals. They are primarily benign slow-growing tumor and arises from ependymal glial cells that line the central spinal canal. According to the updated World Health Organization (WHO) classification, myxopapillary ependymoma is a grade 2 tumor which is morphologically and histologically distinct from other ependymoma subtypes (1, 2).

Spinal myxopapillary ependymoma accounts for 1 to 5% of all spinal tumors, with an estimated incidence of 1 per one million people. Males are more commonly diagnosed than females, affecting individuals between 20 and 40 years old. However, spinal myxopapillary ependymoma can be seen in the pediatric population with a ratio of 8 to 20% of all cases. The clinical presentation depends on the size, location of the tumor and its extension; however, the symptoms usually precede the diagnosis by a few months to years due to the tumor indolent growth features (3-5).

The ideal management for spinal myxopapillary ependymoma is gross total resection (GTR); however, if gross total resection (GTR) is unachievable, a subtotal resection (STR) combined with adjuvant radiotherapy is an option. Despite the risk of recurrence and metastasis, spinal myxopapillary ependymomas generally harbors a favorable prognosis (3).

Multifocal or multicentric spinal myxopapillary ependymomas are uncommon, and the factors associated with multifocal lesions are debatable.

2. OBJECTIVE

There have been 15 adult cases reported with multicentric myxopapillary spinal ependymoma, and we present a case of surgically resected multi-

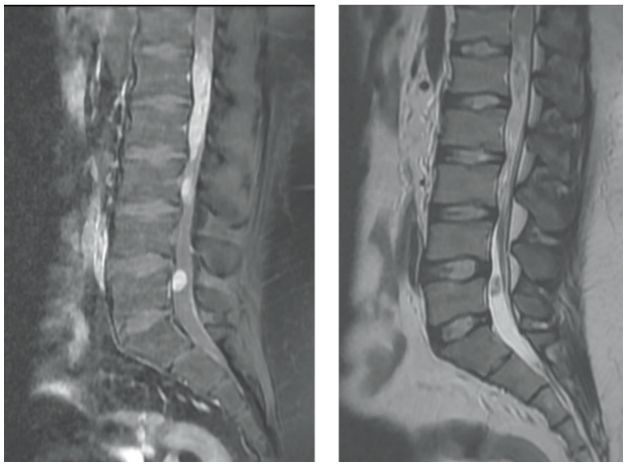

Figure 1. Intense enhancing intradural intra-medullary lesion from the level of the conus medullaris extending to the filum terminalis from the level of T 12 to L3

medullaris extending to the filum terminalis from the level of T12 to L3, and it displays low signal intensity in T1WI and heterogeneous predominant high signal intensity in T2WI. Furthermore, multiple enhancing lesions are seen opposite to L4 and L5 levels and within the exiting nerve roots, at the left side of L1/L2 and L2/L3 and right side of L3/L4 and L5/S1 levels (Figures 1 and 2).

Management

Decision was made to surgically resect most likely symptomatic lesion which was the largest conus medullaris lesion. Patient underwent T12- L3 laminectomy and resection of targeted largest lesion. Patient tolerated surgery with marked improvement on his initial complain.

Histopathological examination

A fresh specimen was sent to the pathology for frozen

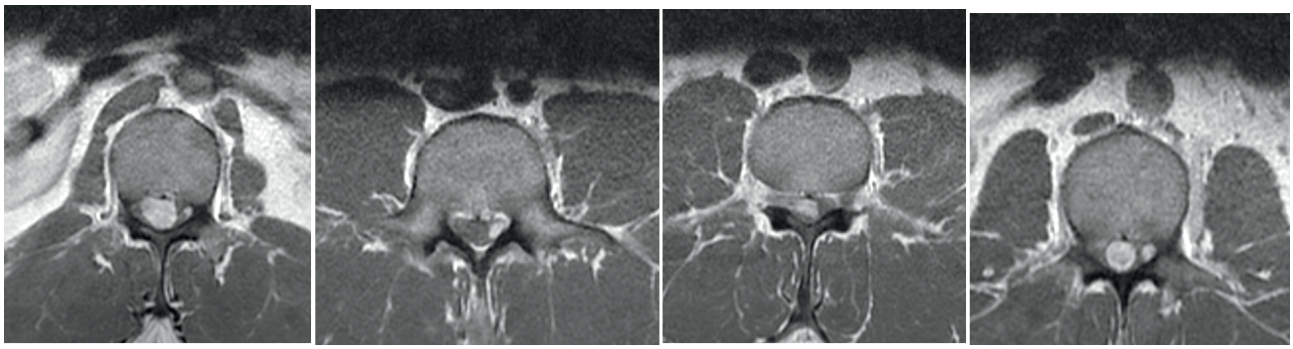

Figure 2. Multiple enhancing lesions are seen opposite to L4 and L5 levels and within the exiting nerve roots, at the left side of LI/L2 and L2/L3 and right side of L3/L4 and L5/Sl levels.

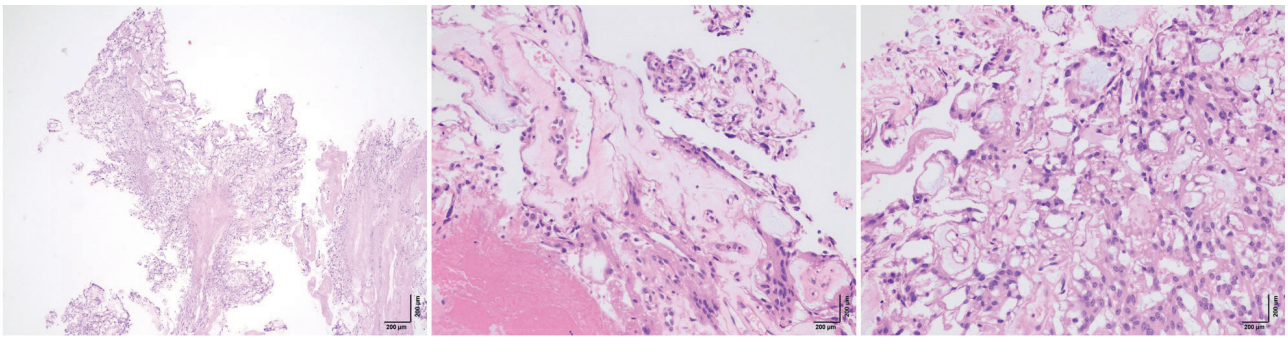

Figure 3. A. The low power view shows lesional tissue exhibiting papillary architecture.(Hematoxylin & Eosin, 4x magnification). B & C. The papillae are lined by slightly elongated cells surrounding vascularized myxoid cores.(llematoxylin & Eosin, 20x magnification).

centric spinal myxopapillary ependymoma, as well as a literature review of previously reported cases.

3. CASE PRESENTATION

Preoperative History and Examination

A 28-year-old man, medically free, complained of axial low back pain over the past year that has progressed during the last three months, and he also started to have radiating pain to the left lower limb. The pain is associated with perianal anesthesia without any sphincter disturbance. Neurological examination revealed an inability to walk on toes and heels with preserved sensory function. Straight leg raise test was positive at a level of 60 degrees.

Initial spinal MRI showed intense enhancing intradural intra-medullary lesion from the level of the conus

section diagnosis, which revealed an Ependymal tumor consistent with Myxopapillary Ependymoma. An additional tissue has been fixed in 10% Neutral buffered formalin and sent to the pathology lab for permanent section diagnosis, totally submitted in six cassettes. The microscopic examination reveals a lesioned tissue displaying papillary architecture (Figure 3A). Elongated cells are radially oriented around hyalinized fibrovascular cores with mucoid degeneration with no significant cytological atypia or mitotic figures (Figures 3B and 3C). Based on the histopathological evaluation, the final diagnosis is Myxopapillary Ependymoma, WHO grade 2. A Ki-67 proliferation index is about 1%.

Post operative MRI

Follow up post operative MRI reveal status post laminectomy at T12-L3 level with resection of the lesion

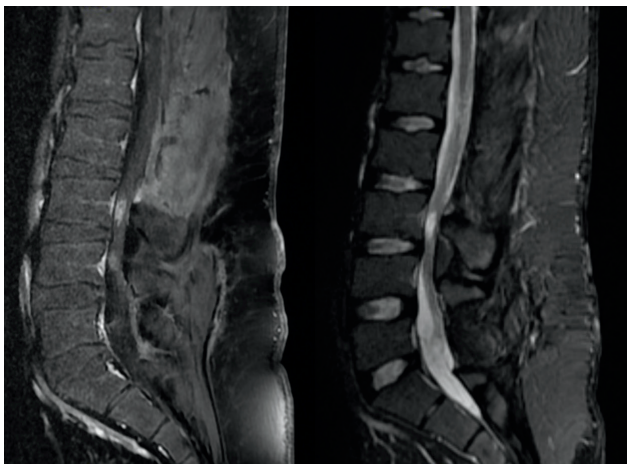

Figure 4. Status post laminectomy at Tl 2-L3 level with resection of the lesion extending from T12-L3 with underlying postoperative changes noted in form of subcutaneous surgical tract with enhancing of the thecal sac at the surgical bed as well as enhancing paraspinal soft tissue.

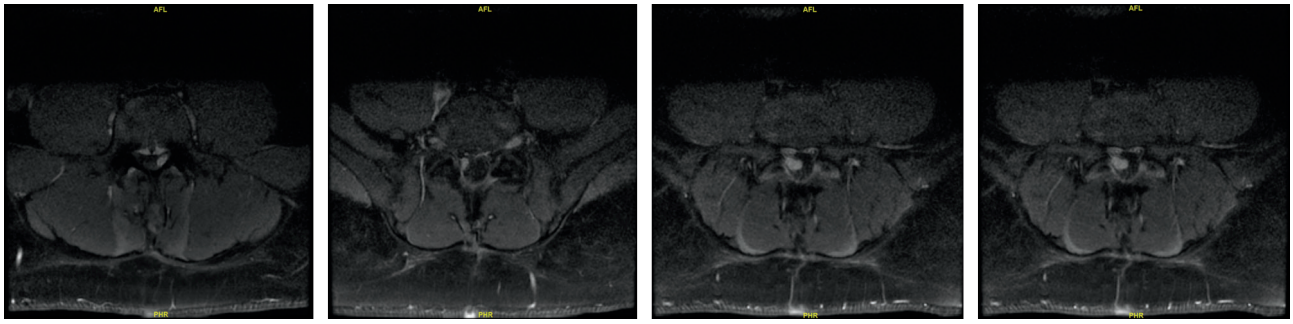

Figure 5. Multiple enhancing lesions are seen opposite to L4 and L5 levels and within the exiting nerve roots, at the left side ofLl/L2 and L2/L3 and right side ofL3/L4 and L5/Sl levels.

extending from T12-L3 with underlying postoperative changes noted in form of subcutaneous surgical tract with enhancing of the thecal sac at the surgical bed as well as enhancing paraspinal soft tissue (Figures 4 and 5).

4. DISCUSSION

Ependymomas are primary central nervous system tumors classified based on pathological features into four categories: subependymoma, myxopapillary ependymoma, conventional ependymomas, and anaplastic ependymoma. Formerly, myxopapillary ependymoma was classified by the world health organization (WHO) as a grade 1 tumor; however, in the new WHO 2021 classification, myxopapillary ependymoma is considered a grade 2 tumor. It's often arises in the filum terminalis and cauda equina. The reason or the pathophysiology behind the myxopapillary appearance needs to be better understood. However, myxopapillary category differs histologically from the other subtypes of ependymal tumors (1, 2, 6).

Histologically, myxopapillary group usually lacks the ependymal rosettes and perivascular pseudo rosettes, which are specific marks of ependymomas. On the other hand, the mucoid stroma is a distinctive feature of the myxopapillary subtype and vascular parietal hyalinization is commonly observed in myxopapillary ependymomas. Calcification, necrosis, cyst formation, and mitotic figures are not usually present (6).

Clinically, myxopapillary ependymoma (MPE) peaks in young adults with slight male predominance. Patients with spinal MPE commonly present with weakness, sensory changes, and pain. Symptoms of urinary, bowel or sexual dysfunction may be present depending on the site of the lesion and the degree of a nerve root or spinal cord compression (7).

Spinal myxopapillary ependymoma is seen in adults more than in pediatrics and is located in the lumbar spine in more than half of the cases (52.9%), followed by lumbosacral region involvement in almost (25%) of cases. On the other hand, myxopapillary ependymoma rarely arises in the brain or cerebral ventricles, and most of these are pediatric cases (3, 7).

In a retrospective study conducted on 101 patients diagnosed with myxopapillary ependymoma, the results revealed slightly male predominance (53.5%), and the median age of diagnosed females was significantly higher than males, with a median age at diagnosis of 45 years old. The most common tumor location was lumbar region in 94% of cases and mostly at the level of L1, L2, and L3 in 88% of cases (8).

Spinal myxopapillary ependymoma in the pediatric age group appears more aggressive and is associated with a higher recurrence rate after resection. In contrast to adults, there are multiple studies and reports of primary seeding of myxopapillary ependymoma at the time of diagnosis in the pediatric population and the role of adjuvant radiotherapy in tumor recurrence or progression. In adults, the diagnosis of multifocal spinal myxopapillary ependymoma is uncommon, and the concept of these differences is not well studied. However, many theories suggest that myxopapillary ependymoma in pediatrics more commonly affects the lumbosacral spine, which predisposes it to dissemination and lymphatic metastasis. Furthermore, some suggested that pediatric myxopapillary ependymoma is histologically varied and accounts for the difference, and it is not related to the anatomical site of the tumor. Another explanation is that the pediatric age group are not easy to communicate with, resulting in delayed presentation and diagnosis. More research and studies are required to address and understand the difference between myxopapillary ependymoma in adults and pediatrics and the reasons behind primary seeding of dissemination (3, 9).

The presence of spinal myxopapillary ependymoma with multifocal lesions at the time of diagnosis is rare, and 15 previous cases have been reported in adult patients and only two cases were for female patients. The most common presentation was low back pain that worsened over few months. Most cases were 26 years and older except for female patients who presented at 18 and 20 years of age, and this could be a characteristic of multifocal lesions hence the studies done on focal spinal myxopapillary ependymoma revealed older age of presentation among female patients. From the 15 patients, gross total resection was achieved in only six cases, and none of these six patients received adjuvant therapy. Nine patients were managed by subtotal resection, and five received adjuvant radiotherapy. Despite receiving radiation, one case had local recurrence after subtotal resection and three years of follow-up; however, most patients reported symptomatic improvement with no tumor progression (6, 9-18).

The mainstay management of spinal myxopapillary ependymoma is gross total resection (GTR) which provides the most remarkable result and minimizes the chance of tumor recurrence and metastasis. On the other hand, subtotal resection (STR) is associated with a higher risk of tumor recurrence with a 30% recurrence rate probability within an average of 3.5 years. Furthermore, there is a link between tumor recurrence and tumor capsular disruption; the risk may extend up to 45% following STR and 15% after GTR. In situations of capsular violation, adjuvant radiotherapy is recommended to reduce the risk of recurrence or metastasis regardless of the resection extent (7, 19).

Overall, the benefit of adjuvant radiotherapy in spinal myxopapillary ependymoma remains controversial; some studies suggest that it is associated with recurrence reduction, especially in cases managed by STR, and on the other hand, a retrospective study revealed that adjuvant radiotherapy is not associated with recurrence reduction after GTR or STR (19). However, adjuvant radiotherapy is not recommended in cases where GTR without capsular violation was achieved, mainly if post-operative CSF analysis did not show any indications of dissemination (20, 21).

Myxopapillary ependymoma tumors in adults could recure with a likelihood of around 32-32% following subtotal resection (STR) and 10-20% after grossly gross total resection (GTR). In pediatrics, the tumor is associated with more aggressive features and a higher incidence of multifocal behavior; consequently, the risk of recurrence is higher and may reach up to 40% (3).

The recurrence and the prognosis of myxopapillary ependymoma are related to multiple factors such as large tumor size, incomplete tumor resection, multifocal lesion, sacral spine involvement, dural perforation by the tumor, intraoperative need for blood transfusion, blood loss of more than 500CC during surgery, and longer operation time longer than 3 hours. Despite the risk of tumor recurrence and metastasis, the overall prognosis is good, and 90% of patients experienced a good result with no significant neurological deficit. However, the long-term prognosis is not well studied and requires sufficient and extended enough follow-up (8).

5. CONCLUSION

Myxopapillary ependymoma is an ependymal cell tumor that primarily arises in the conus medullaris, cauda equina, or film terminals. It is a slow-growing tumor that mainly affects patients in their third decade and is slightly predominant in males. Clinical presentation varies depending on the tumor site and size. It is rarely present as a multifocal lesion in adults and its associated with risk of recurrence and dissemination. Gross total resection is the mainstay of treatment. However, subtotal resection and adjuvant radiotherapy are alternatives. The overall prognosis of the tumor is good, with a lack of evidence of long-term prognosis and morbidity.

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