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Giant Myxopapillary Ependymoma with Multi-Site Neural Axis Metastases: A Rare Case with Suboptimal Outcome

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Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
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Patient: Male, 44-year-old
Final Diagnosis: Myxopapillary ependymoma
Symptoms: Low back pain
Clinical Procedure: Laminecromy • laminoplasty • radiotherapy
Specialty: Neurology • Neurosurgery • Oncology • Pathology • Radiology

Objective: Rare disease

Background: Myxopapillary ependymoma is a rare type of slow-growing tumor that mainly occurs in the spinal cord, particularly in the region of the conus medullaris and the cauda equina. It originates from the ependymal glial cells found in the filum terminale.

Case Report: We present a clinical case of a 44-year-old male patient who presented with symptoms of non-specific pain in the lower back persisting for the past 2 years. He did not report any specific neurological deficits or radicular symptoms. Unenhanced MRI of the lumbar spine showed a giant intradural, extramedullary, heterogenous, expansive tumor at the level L1-S4 with erosion of the sacral bone and invasion of presacral tissue.
Based on its characteristic localization and growth pattern, suspicion arose for myxopapillary ependymoma. Biopsy confirmed the initial diagnosis. Partial resection of the tumor with laminectomy and laminoplasty was deemed necessary. Preoperative neural axis MRI showed contrast-enhancing lesions in the cerebellum and the cervical and thoracic spine; therefore, adjuvant radiation therapy was administered.
Following the surgery, the patient experienced intermittent episodes of neurological deficits and required physiotherapy. Control MRI a year after the operation showed tumor growth and more metastases along the neural axis.

Conclusions: Complete surgical excision of the tumor is the preferred treatment approach, but there is a risk of recurrence even after total excision, so radiotherapy is recommended to minimize the risk of recurrence. Prior to surgery, it is essential to conduct MRI/PET/CT of the head and spine to assess the possibility of metastases.

Keywords: Brain Metastases • Spine Metastases • Central Nervous System • Ependymoma • Magnetic Resonance Imaging • Medical Oncology • Myxopapillary Ependymoma

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Background

Myxopapillary ependymoma belongs to a group of glial tumors, primarily developing from the ependymal cells of the filum terminale and are commonly found in an intradural extramedullary location [1,2]. The exact cause of myxopapillary ependymoma remains unclear. It does not seem to have any strong genetic or environmental associations [3].

In contrast to previous WHO classifications, myxopapillary ependymoma is now considered as WHO Grade 2, as its potential for recurrence is now recognized to be comparable to that of conventional spinal ependymoma [4].

The primary locations where myxopapillary ependymoma (MPE) is predominantly found are the conus medullaris and the filum terminale. However, there have been occasional reports of rare cases where tumors have occurred in the brain, upper spinal cord, or even outside the central nervous system (CNS) [5-7].

The tumor's compression on the spinal nerve roots results in distinctive symptoms, including radicular back pain, bowel and bladder dysfunction, sensory loss, and dysfunction of lower extremities. If the lumbar spine is affected, patients may also experience paresthesia, a tingling or prickling sensation in the lower extremities [8,9]. Moreover, the symptoms associated with myxopapillary ependymoma can mimic other conditions, making it even more challenging to diagnose correctly.

Myxopapillary ependymoma can dilate the spinal canal and cause deformation of the vertebral bodies, and its growth can be seen on conventional X-ray [10,11]. Typical MRI findings are well-circumscribed intradural tumors, round or elongated in shape, lobulated in outline, usually occupying more than 1 vertebral body height level. Smaller tumors dislocate nerve roots and the cauda equina. Larger-sized myxopapillary ependymomas often compress or envelop them [12].

The tumor's location near critical structures, such as nerves and blood vessels, can make surgical removal quite complicated [13,14]. While surgery is often the primary treatment, achieving complete resection without causing neurological deficits is a major challenge for neurosurgeons [15].

Case Report

The 44-year-old male patient presented with symptoms of nonspecific pain in the lower back persisting for the past 2 years.

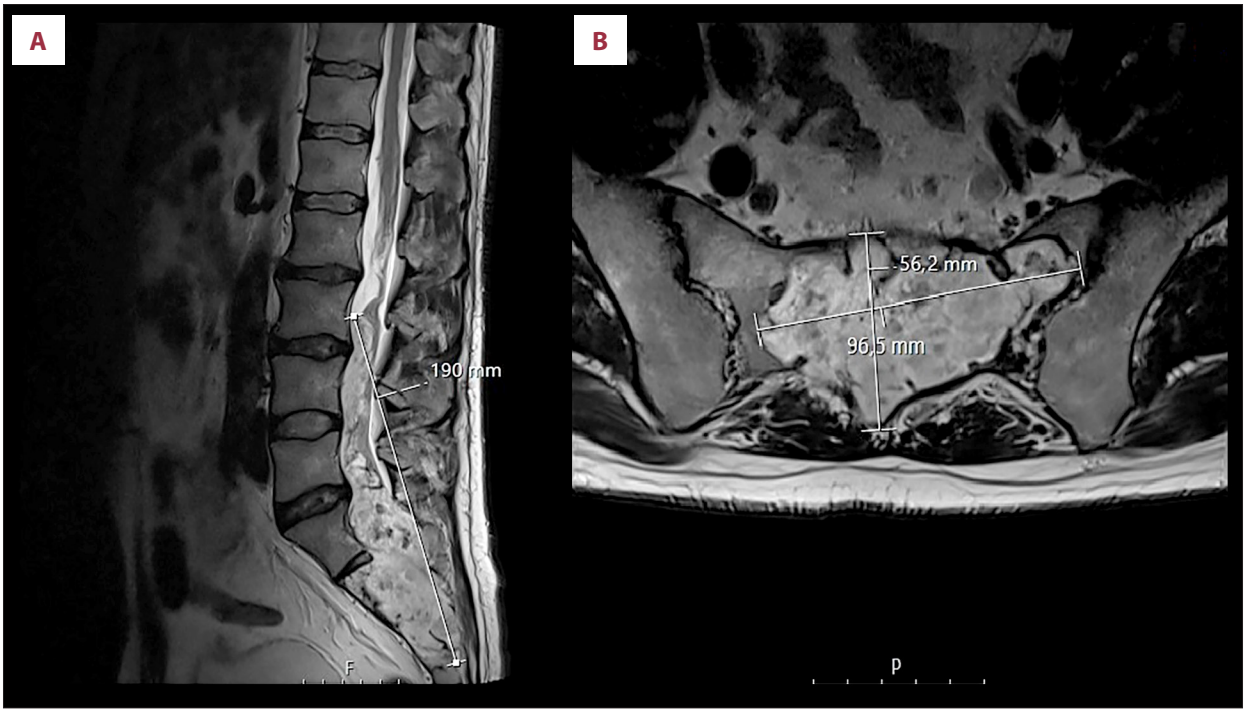

Figure 1. T2-weighted sagittal and axial MRI of the lumbar spine without intravenous contrast. A giant, intradural, extramedullary, heterogeneous, lobulated, expanding tumor was found in the lumbar spine and sacrum canal. Tumor dimensions were 56.2 mm (AP)×96.5 mm (LL)×19.0 mm (craniocaudal). Starting from the L5-S1 level, there was significant erosion of the sacrum, lateral masses more on the left side, expanding the channel, as well as openings with the spread of the presacral and in the back muscles with the incoming nerve root and plexus displacement.

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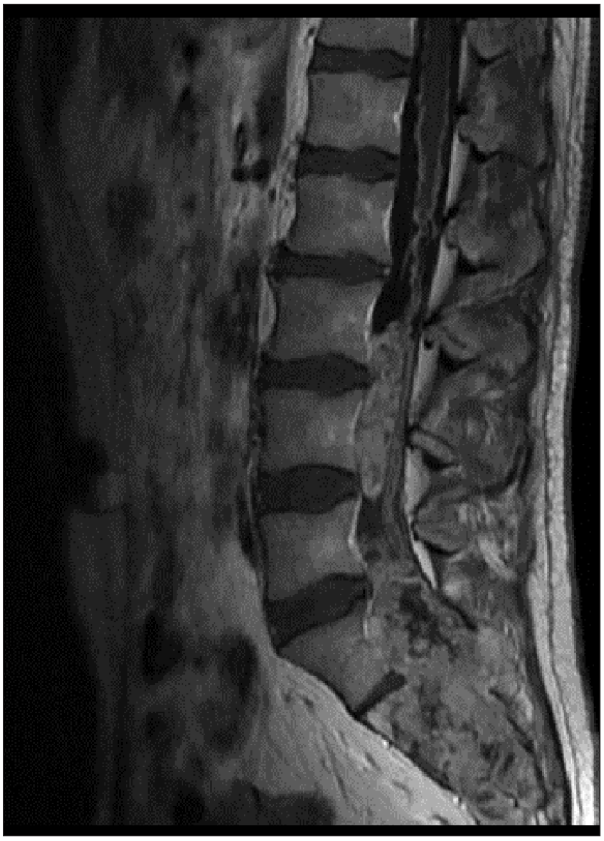

Figure 2. T1-weighted sagittal MRI of the lumbar spine with intravenous contrast administration showing a large, intradural, extramedullary tumor of the lumbar sacrum canal of the spine. The tumor started from the L1 to S3 level. Pathological tissue can also be observed in the end sheaths of the conus medullaris.

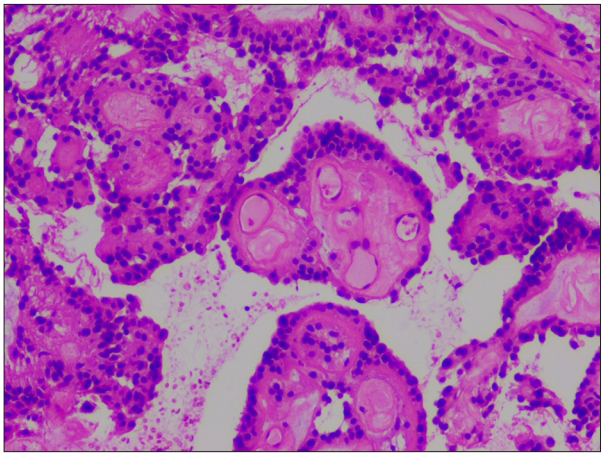

Figure 3. Hematoxylin-eosin staining. The papillary formation exhibits flat-cubic cell coverage on its papillary structures; microcysts with mucinous degeneration are present in the formation.

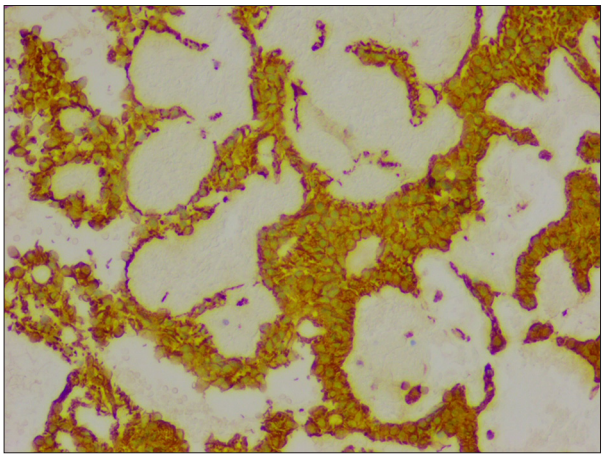

Figure 4. Glial fibrillary acidic protein staining. GFAP expression in tumor cells (×100).

He did not report any specific neurological deficits or radicular symptoms, and denied having any other chronic diseases.

On 16/07/2021 a conventional X-ray of the lumbar spine was conducted. X-ray showed mild degenerative changes – deformative spondylosis.

In the dynamic context on 08/09/2021 native MRI was performed, showing a giant intradural, extramedullary, heterogeneous, lobulated expanding tumor in the lumbar spine and sacrum canal. This tumor can be considered as giant due to its substantial dimensions, measuring 56.2 mm in the anterior-posterior projection, 96.5 mm in the lateral projection, and 19.0 mm in the craniocaudal direction. The tumor tissue in the L1-L2 level of the spinal cord was small, elongated, and centrally located between the roots of the cauda equina. However, starting from the lower edge of L3, the tumor tissue became larger and occupied the entire intradural space, causing deformation and peripheral and dorsal displacement of the roots of the cauda equina. At the level of L5-S1, the sacrum showed significant erosion, affecting the S1 vertebral body and arch, as well as practically the entire S2-S3 vertebral bodies. The lateral masses were more prominent on the left side, resulting in expansion of the spinal canal and spread of the tumor into the presacral area. Additionally, the tumor affected the back muscles and caused displacement of the nerve roots and plexus (**Figure 1**).

Myxopapillary ependymoma was suspected. The tumor should usually be differentiated from a schwannoma but does not correspond to root displacement. For clarification, it is necessary to perform an additional MRI with intravenous injection of a contrast.

MR of the lumbar spine with iv contrast was conducted to establish the diagnosis on 11/10/2021. It showed a giant intradural, extramedullary tumor of the lumbar sacrum canal of the

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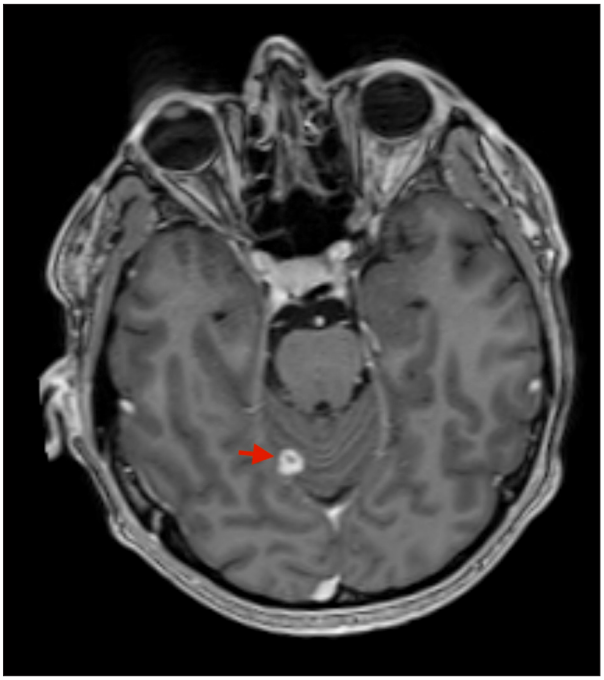

Figure 5. T1-weighted coronal MRI after iv contrast injection before surgery shows a small, 5-6 mm pathological mass, most likely a metastasis, in the right cerebellum in the posterior fossa (arrow).

spine starting from the end of the spinal cord. At the S2-S3 level, tissue was more on the left side, expanding the channel in conjunction with the back muscles and the presacral zone. Pathological tissue was also seen in the end sheaths of the conus medullaris, where the accumulation of contrast material with a markedly uneven contour was observed (**Figure 2**).

The patient was hospitalized in scheduled for an arranged biopsy on 12/11/2021. Visually, the sacrum was bluish, remodeled, and thin. The maximum thickness of the bone was 1-1.5 mm. Access was made to the tumor, which visually resembled an ependymoma. Tissue fragments were taken for histological examination.

The tumor was stained with hematoxylin-eosin, revealing papillary formation, with papillary structures covered with flat-cubic cells, in some places microcysts, in the formation, with mucinous degeneration (**Figure 3**). The morphological and immunohistochemical picture was consistent with myxopapillary ependymoma. After undergoing the WHO grading system, the tumor was graded as Grade 2 (**Figure 4**).

The patient was prescribed surgical therapy. Before surgery, MRCI of the head with iv contrast and MRI of the thoracic spine with intravenous contrast T1 dix sag C+ were ordered. Metastases were visualized along the CSF pathways in all parts of the spinal cord. Compared to the previous MR examination,

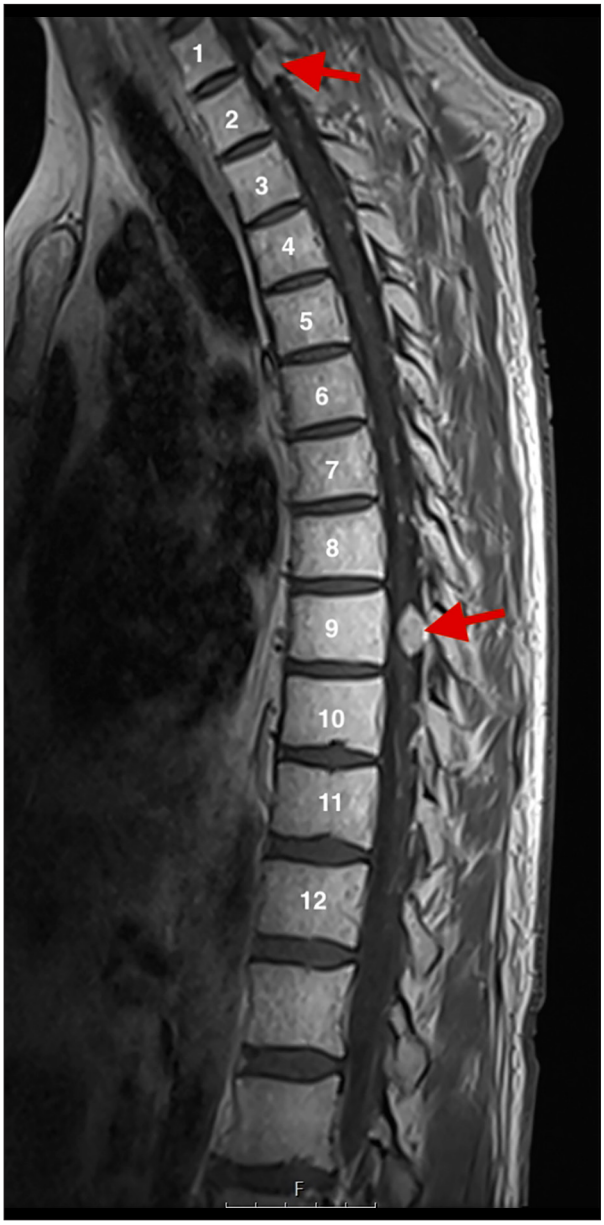

Figure 6. T1-weighted sagittal MRI of thoracic spine with intravenous contrast injection before surgery T1 dix sag C+. Pathological masses, most likely metastases, are seen in the spinal canal in the chest area at C7-Th1, Th1-Th2, and Th10 levels (arrows).

the pathological formation in the sacral canal was without significant increase in size. There was dissemination in the spinal canal in the thoracic region at C7-Th1, Th1-Th2, and Th10 levels and in the cerebellum (**Figures 5, 6**).

The patient was hospitalized in for arranged surgery. The procedure involved a L3-S4 laminecromy, partial resection of spinal intradural tumor, and L3-S4 laminoplasty (**Figures 7, 8**).

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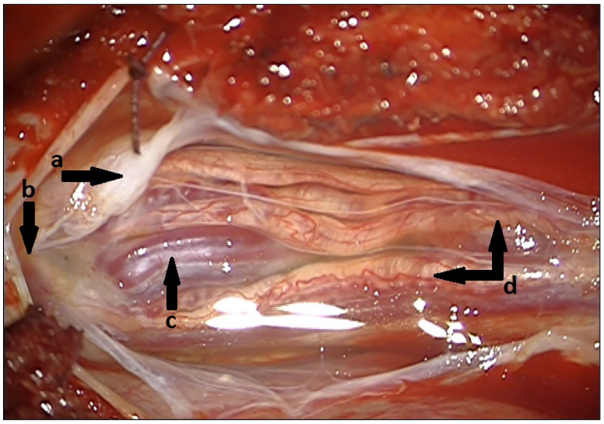

Figure 7. The affected area of the lumbar spine before surgery. (**a**) dura mater, (**b**) conus medullaris, (**c**) ependymoma, (**d**) nerve roots.

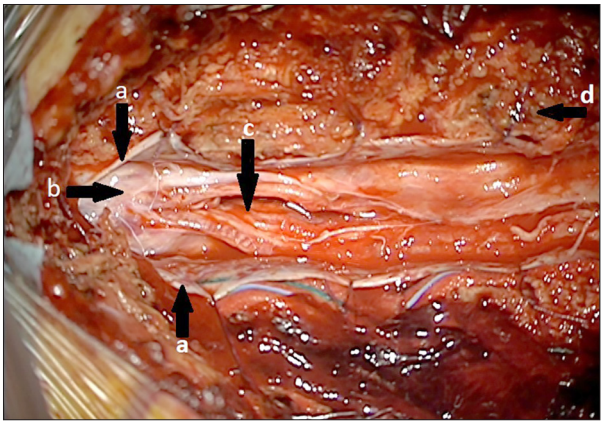

Figure 8. The affected area of the lumbar spine after surgery. (**a**) dura mater, (**b**) conus medullaris, (**c**) surgical site, (**d**) adipose tissue.

The identification of contrast-enhancing lesions in the cerebellum, cervical spine, and thoracic spine led to the conclusion that adjuvant radiation therapy should be initiated. This decision was made based on the presence of these lesions, which indicated the need for additional treatment to target and manage the affected areas, ensuring comprehensive care for the patient's condition.

MRI was performed 1 day post-operatively. After the applied surgical treatment, the tumor was largely removed. At the L3-L4 level, the tumor tissue is not differentiated against the background of edema, and the roots of the cauda equina are grouped at this level. A smaller amount of residual tumor tissue remained at the L5-S1 level (**Figure 9**).

PET/CT examination was performed after surgery and radiotherapy. Little and moderate metabolism was visualized in the spinal canal of the sacrum, according to the remodeled area of the bone, with metabolism that was suspected to be specific

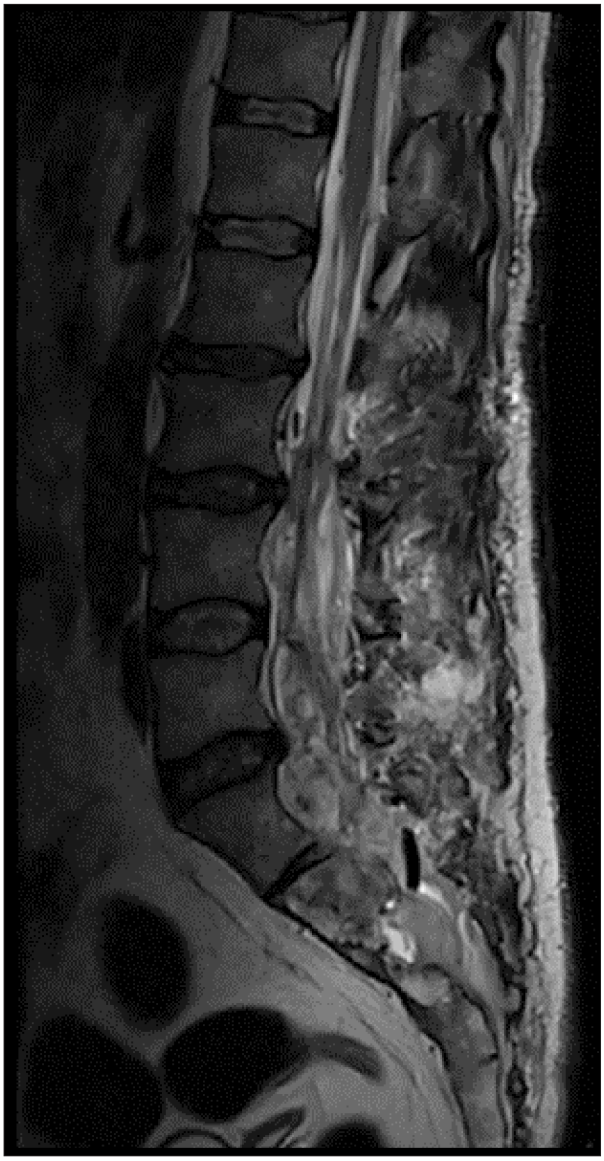

Figure 9. T2-weighted sagittal MRI of the lumbar spine one day after surgery. The tumor has been mostly excised; a small amount of remaining tumor tissue can be observed at the L5-S1 level. The retro-sacral tumor tissue has been completely removed. The surgical wound contains hemorrhagic contents with air pockets.

(PET-positive). There was a weakly expressed, hypermetabolic focus in the spinal cord, at the Th9 level, and slightly uneven metabolism in the cervical spine, which, considering the MRI picture, suggested specific changes (PET/CT weakly positive). Reactive post-radiation changes were also visualized in the chest, waist, and sacrum, and in the soft tissues of the back, which were likely post-radiation changes. Elsewhere in the body examination, there was no indication of the spread of the oncoprocess, but it should be noted that primary PET

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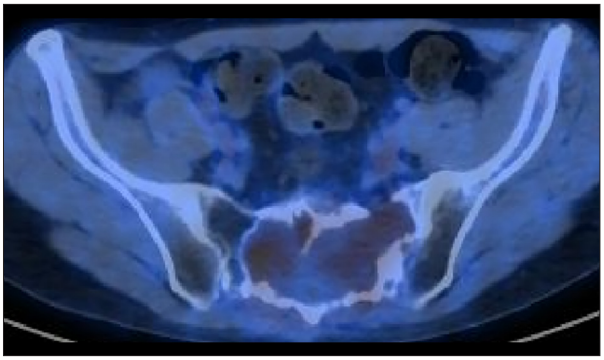

Figure 10. PET/CT axial slices showing pathologic changes in sacrum with slight and moderate uneven hypermetabolism similar to the metabolic background of liver. Standard uptake value in liver was 2.7, but in the lesion in sacrum SUVmax was 2.5.

data before therapy were not available, which may have limited the sensitivity (**Figure 10**).

Following the surgery, the patient experienced intermittent episodes of neurological deficits, and subsequently required physiotherapy and long-term follow-up.

Discussion

Comparing our clinical case with previously published articles, we observe a limited number of published cases of myxopapillary ependymoma [13,14], and even fewer clinical cases with such giant tumors.

The primary treatment for myxopapillary ependymomas is surgical resection to remove the tumor and relieve symptoms. Due to the large size of the tumor, the sacrum was remodeled, resulting in an exceptionally thin bone plate. Without appropriate intervention, this thin bone plate posed a severe threat to the patient's life, necessitating immediate medical attention to reduce the potentially fatal consequences. However, surgery in the spinal cord region carries risks such as nerve damage, spinal cord injury, and infections. These complications can impact the patient's mobility, sensory functions, and quality of life, affecting the overall prognosis.

Such large tumors can cause chronic pain due to nerve compression or irritation. Surgery and other treatments can alleviate the pain from the nerve compression, but there can be residual discomfort or chronic pain. These deficits can impact the patient's daily life and prognosis.

While most publications on myxopapillary ependymomas highlight tumor local growth without metastases, in our clinical case multiple metastases were observed. Our patient received radiation therapy that led to complications such as tissue damage and neurologic deficits. These complications can affect the patient's functional abilities and long-term prognosis. It is advisable to conduct screening of the complete central nervous system axis, both at the point of diagnosis and during subsequent follow-up examinations.

With our clinical case, we want to show that the outcome was suboptimal due to previously mentioned reasons. Myxopapillary ependymoma presents a challenging situation for patients and healthcare professionals. Its location, potential for recurrence, and limited treatment options make it a complex tumor to manage. Therefore, our work adds additional relevant information to the already published articles.

Clinical trials play a vital role in understanding of myxopapillary ependymoma and finding better treatment approaches. Lack of data makes it difficult for clinicians to determine the best course of action for each patient, leading to individualized treatment plans based on the tumor's size, location, and the patient's overall health. With ongoing research and a multidisciplinary approach, there's hope that we can improve outcomes and quality of life for patients affected by tumor masses that outgrow the vertebrae.

Conclusions

Although the survival rates frequently cited in the literature might indicate a favorable prognosis for individuals with such tumors, it is crucial to acknowledge the more comprehensive perspective. Complete surgical excision of the tumor is the preferred treatment approach, with the potential for a curative outcome. However, there is a risk of recurrence even after total excision; therefore, radiotherapy is recommended. Prior to surgery, it is essential to conduct a comprehensive MRI/PET/CT of the head and spine with iv contrast material to assess the possibility of metastases.

Having a comprehensive understanding of the characteristics, diagnostic techniques, and treatment alternatives for myxopapillary ependymoma is essential for effective management of this particular conus medullaris tumor.

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