Case Report
A unique case of upper cervical spinal dural fusocellular sarcoma in a 25-year-old male: a case report and comprehensive treatment analysis

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Abstract: Fusocellular sarcoma is a rare malignant tumor of mesenchymal cells, and a case of fusocellular sarcoma originating from the dura mater spinalis has never been reported before. The high degree of malignancy, undefined histological characteristics and poor clinical prognosis indicate the need for more clinical research of this disease. We hereby present a case of upper cervical spinal dural fusocellular sarcoma. The patient initially appeared to have a meningioma but was eventually diagnosed with fusocellular sarcoma originating from the dura mater spinalis. Despite receiving surgical management and chemotherapy promptly after the diagnosis and undergoing close observation following treatment, the patient still developed metastases to multiple sites of the lung and brain. Taking into account the similar presentation of this rare disease to other entities, the early and accurate diagnosis of cervical spinal dural fusocellular sarcoma is vital. The condition should be considered a life-threatening condition with the possibility of metastasis to other sites of the body.

Keywords: Fusocellular sarcoma, dura mater spinalis, diagnosis, therapy

Introduction

Fusocellular sarcoma is a rare type of malignant tumor [1], and a case of fusocellular sarcoma originating from the dura mater spinalis has never been reported before. The surprisingly high degree of malignancy regarding its clinical manifestation indicates the need for more clinical research of this disease [2, 3]. We hereby present a case of upper cervical spinal dural fusocellular sarcoma. The patient initially appeared to have a meningioma but was eventually diagnosed with fusocellular sarcoma originating from the dura mater spinalis. Despite receiving surgical management and chemotherapy promptly after the diagnosis and undergoing close observation following treatment, the patient still developed metastases to multiple sites of the lung and brain. The diagnostic and therapeutic challenges strongly emphasize the importance of this topic for future investigations.

Case presentation

A 25-year-old man was admitted for a five-month history of discomfort in the neck, a 15-day history of limb weakness, and sudden paralysis for one day. Five months ago, cervical discomfort appeared with no obvious cause; symptoms manifested mainly as cervical stiffness and paroxysmal symptoms with each attack lasting for a different length of time. Then, the patient underwent a large cervical vascular color Doppler ultrasound examination at a local hospital. The results indicated occlusion of the left internal carotid artery. Four months ago, he was again admitted to the hospital with a diagnosis of arteritis. At the hospital, he received hormone therapy and treatment to improve circulation as well as various other symptomatic treatments. However, there was still no significant improvement in his symptoms. Pulmonary computed tomography (CT), cranial CT and magnetic resonance imaging
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(MRI) examination were not obviously abnormal at that time. However, fifteen days ago, limb weakness appeared with a gradual deterioration of symptoms, at which point the patient came to our hospital. We learned that the patient had experienced dysuria the night before, and his limb weakness subsequently became gradually more severe. He exhibited poor appetite and weight loss of greater than 20 kg in the past 20 days. Medical examination revealed the following: neck turning and shrugging was abnormal; muscle strength testing revealed 2/5 in the bilateral upper extremity; and the bilateral lower extremities were 3-/5 for strength. The only other myelopathic sign was a positive bilateral Babinski sign. An MRI of the spine revealed an obvious lesion of the cervical spinal cord with obvious compression against the cervical medulla spinalis. An upper cervical segment of the spinal cord was blocked, forming a cupped mouth shape (Figure 1A, 1B). An abnormal signal shadow with a shuttle shape appeared outside the medulla spinalis but inside the canalis spinalis at C4-C5. The mass crossed over the intervertebral foramen, and compression and displacement of the corresponding spinal cord was apparent. The size of the occupying lesion was approximately 2.5×1.6×0.6 cm. The initial diagnosis was occupying lesions of the cervical spinal cord, with a more detailed diagnosis indicating meningio-mas. Then, we performed surgical resection of the cervical spinal cord occupying lesions using a posterior median approach. When the dura mater spinalis was open, we found that the subdural space was filled with pale red granulation tissue. A granulation tissue mass, tightly adhered to the medulla spinalis, was apparent on the backside of the corresponding spinal segment. Even light stimulation could cause great pain for the patient. Partial resection was performed for a postoperative biopsy. Two indwelling drainage tubes were left for postoperative draining after adequate hemostasis. Part of the resected tissue was taken for biopsy (Figure 2).

Under a light microscope, tumor cells appeared spindle-shaped and bunchy, indicating an infiltrative tendency. Endochylema was slightly dyed. The nuclei were spindle-shaped and of various sizes, and nuclear division was apparent. In addition, remnants of skeletal muscle were present. Thus, the primary diagnosis could be a malignant spindle cell tumor (epidural mass). Histopathological examination revealed the following: CD117 (-), CD34 (-), CK (-), Desmin (-), EMA (-), Ki-67 (+10~20%) (-), PR (-), S-100 (+), SMA (+), Vimentin (+), 34βE12 (-), ALK (-), CK5/6 (-), CK7 (-), HBM45 (-), and Melan-A (-). If clinical metastasis could be excluded, meningo-associated fibrosarcoma could be considered a more accurate diagnosis. However, surprisingly, postoperative re-examination revealed that multiple lesions with nodulous cumulus high-density shadows with obscure boundaries were observed in the pulmonary CT image (Figure 3A, 3B). The larger lesion may have a diameter of approximately

Figure 1. Radiological findings: Sagittal T1-WI (A) and axial T2-WI (B) of MRI revealed an intraspinal mass with a protrusion into the medullary cavity (arrow). MRI: magnetic resonance imaging.

Figure 2. Histopathological examination of the resected tissue taken for biopsy after the operation. Immunohistochemical analysis further indicated that the tumor was immunopositive for S-100 (+), SMA (+) and Vimentin (+).
2.3 cm. Bilateral pleural effusion and pulmonary swelling of the bilateral lower lobe was apparent. In addition, the anterior inferior mediastinal lymph nodes were slightly enlarged, and this might indicate tumor metastasis. Cranial CT (Figure 4) indicated an occupying lesion of the right pars frontalis under the cranial plate, together with a soft tissue mass of the left pars parietalis under the scalp. Thus, tumor metastasis was found to occur in both the lung and the brain, and the final diagnosis was spindle dural fusocellular sarcoma. Furthermore, a postoperative cervical CT of the upper cervical segment further indicated soft tissue swelling at C4-C5, with a patchy high-density shadow inside and an unclear boundary. In addition, spinal canal stenosis and a slight release of compression of the mass of the dura mater spinalis against the spinal marrow formed by the cervical spinal dural fusocellular sarcoma was observed. A discontinuous fever occurred after the operation, but no clear focus of infection was found. Dexamethasone has been continuously administered since the operation. In addition, rehydration, hemostasis and anti-infection treatments as well as nutrition replenishment for the brain and various other symptomatic treatments were used. The patient’s general condition was good in the days following the operation; the patient exhibited stable vital signs, and his muscle strength improved slightly. Then, the patient was transferred to the oncology department for further chemotherapy. Postoperative follow-up revealed that this patient died one month later with systemic deterioration of the whole body, which was likely due to rapid tumor metastasis.

Discussion

Sarcomas are a heterogeneous group of solid tumors arising from either soft tissues or bone and accounting for approximately 1% of all cancers in adults [4]. Fusocellular tumors could be of complex morphology and are generally accompanied by an interstitial component. When fusocellular tumors appear in mesenchymal tissues, fusocellular sarcoma is formed. Since large variations in morphology and structure can be observed in fusocellular sarcoma, understanding the patients’ information regarding clinical symptoms and anamnesis of related diseases could be significantly important for making accurate diagnoses. Histopathological features and immunohistochemical characteristics were indispensably significant for the final histopathological diagnosis. Intriguingly, fusocellular sarcoma is one of the most common types of soft tissue sarcomas in adults, with an occurrence rate of 50%. It can occur in people of any age, and the ratio of affected males to affected females is approximately 1/3. The tumors are usually located in the dermis and subcutaneous tissues of the extremities, presenting as a nodule or rash externally. However, fusocellular sarcoma originating from...
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the dura mater spinalis is quite rare, and there have been no other reports of this type in recent years. Additionally, since the histological characteristics were not obvious, further classification of them was difficult. When fusocellular sarcoma originating from the dura mater spinalis occurred, it mostly presented with a high degree of malignancy and poor prognosis.

MRI is useful in the diagnosis of cervical spinal dural fusocellular sarcoma, and should be considered the first choice for diagnosing this condition. It is important to have an accurate histological diagnosis to determine the treatment plan. Thus, a timely preoperative biopsy is considerably important since the diffusion and cancer metastasis of fusocellular sarcoma could occur rapidly and uncontrollably. In this case, we observed that the tumor cell membrane was complete and that the tumor edge was clear and smooth; this could easily be misdiagnosed as meningioma or neurofibroma [5, 6]. However, the pathological diagnosis after operation indicated that this was fusocellular sarcoma, with an unexpected and extremely high degree of malignancy. We concluded that, with only the patient history and imaging features, a differential diagnosis between cervical spinal dural fusocellular sarcoma and various other spinal tumors, such as meningioma, could be a challenge without the current histological diagnosis. Furthermore, the intraspinal lesions near the intervertebral foramen indicated signs of malignancy. Additionally, various other cervical vertebrae and even the thoracic vertebrae exhibited abnormal signal shadows further confirming the severe metastasis of this malignant tumor.

Regarding the treatments, it might be roughly separated into preoperative chemotherapy (neoadjuvant chemotherapy), surgical operation and postoperative chemotherapy together with radiation therapy. Chemotherapy could be an important treatment for cervical spinal dural fusocellular sarcoma. Chemotherapy can improve survival rates by inhibiting lung metastasis. Furthermore, preoperative chemotherapy leads to the necrosis of tumor cells, promoting decreases in tumor size and in the edema region of the reaction zone and advancing the disappearance of newly formed tumor vessels. This could also reduce the “unplanned excision”, which was introduced to describe operations performed for any excision of soft tissue sarcoma without regard for preoperative imaging or the need to remove a margin of normal tissue covering the cancer [7-9]. In addition, pain relief or disappearance could further support the use of preoperative chemotherapy. High-dose chemotherapy could inhibit lung metastasis and systemic micrometastasis of the whole body. Intriguingly, the effect of preoperative chemotherapy could also partly predict cure rate, which warrants additional research in the future. These treatments should be performed as early as possible, since small lesions are more sensitive to chemotherapy than large tumors and larger ones could be more resistant. In addition, it is known that the key for prolonging the survival of patients with pulmonary metastasis is the complete resection of metastasis, and neoadjuvant chemotherapy can reduce and delay the appearance of pulmonary metastasis. Thus, the appropriate chemotherapy could increase the cure rate of this disease.

Taking into account the similar presentation of this rare disease to other entities, the early and accurate diagnosis of cervical spinal dural fusocellular sarcoma is vital, and the condition should be considered a life-threatening condition with the possibility of metastasis to other sites of the body. The possible forthcoming standard treatment could involve a multidisciplinary approach including surgery, chemotherapy, and even radiation therapy with the particular preferential order determined on a case-by-case basis. Survivorship is approximately 45% at 5 years after treatment [10]. Although the clinical data in this report need to be more concrete, we believe that they could provide critical information for the design of larger and more focused clinical studies that will be necessary to systematically constitute more standard treatments. Given the low incidence of this pathology presenting in tumors of the spinal cord and the lack of treatment guidelines, each patient’s plan should be considered on a case-by-case basis until further studies are performed to determine optimal evidence-based treatments.

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Disclosure of conflict of interest

None.

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