Case report of a huge primary malignant melanoma of the spine

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Abstract: Primary spinal melanoma (PSM) is very rare and the diagnosis of PSM is difficult because of its unusual location and variations in its appearance on MRI output. A 42-year-old female patient was admitted to our hospital with her chief complaint being back pain, especially at night, over an 8 month period. The physical examination of the patient showed hypoesthesia in the lateral area of her left lower leg and myodynamia of the right hip flexor muscles and knee extensor muscles weakened to Grade 4. Lumbar CT scan and MRI confirmed the bone destruction of the L5 vertebrae and posterior elements. A biopsy pathological examination supported a melanoma-rich tumour. A surgery of posterior partial tumour resection, spinal canal decompression, and instrumentation with a pedicle screw system was performed. The tumour was black, invasive to the L5 vertebra and posterior elements, surrounding the anterior part of the L5 vertebra, and involving muscles, tissues, and dura. HE staining showed a melanoma-rich tumour with significant fibrous, and granulation, tissue present. Immunohistochemical staining results showed S-100 (+), HMB45 (+), A103 (+). Systemic examinations excluded the possibility of a metastatic tumour. The patient was finally diagnosed as primary spinal malignant melanoma. After surgery the patient was sent to the oncology department for further treatment which included chemotherapy and radiotherapy. No deterioration of neurological function was detected with a follow-up after three months. In conclusion, the diagnosis of malignant melanoma is based on the discovery of melanin granules in the tumour cells upon histological examination, and a positive test for HMB-45, melanoma-specific antigen (Melan-A), and S-100 upon immunohistochemical examination. Systemic examinations including full-body skin examinations, head-MRI, chest and abdomen CT scan, abdominal and pelvic ultrasound examination, and PET/CT were also recommended to exclude the possibility of a metastatic tumour.

Keywords: Primary malignant melanoma, spine, vertebrae, diagnosis

Introduction

Since Hirschberg first reported a case of primary spinal melanoma (PSM) in 1906, approximately 30 cases of PSM have been reported to date [1]. PSM can occur throughout the cranial and spinal regions, however, it occurs most frequently in the middle or lower thoracic spine [2]. PSM was observed more frequently in Caucasians and PSM in Asians is rare. Most PSMs were reported to be intraspinal without irritation of the vertebrae and peri-vertebral space: the diagnosis of PSM is difficult because of its unusual location and the possible variations in its appearance on magnetic resonance imaging (MRI) output. In some cases, even where post-operative histopathological and immunohistochemical examinations were performed, the diagnosis can still be difficult as was the case with this patient. Considering the paucity of knowledge of this area we present this special case of huge primary malignant melanoma involved in the L5 vertebrae and peri-vertebral space with unusual radiographic features in a non-Caucasian patient to share our experience.

Case report

The patient provided informed consent for the publication of her clinical, pathological, and radiological data. This case report was approved by the Medical Ethics Committee of West China Hospital, Sichuan University.

A 42-year-old female patient was admitted to our hospital with her primary complaint being back pain, especially at night, over an eight...
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Figure 1. The pre-operative lumbar computed tomography (CT) scan three-dimensional reconstruction images of this patient. The pre-operative lumbar computed tomography (CT) scan three-dimensional reconstruction images (A, B: Coronal view reconstruction images; C, D: Axial view images) showed the bone destruction of the L5 vertebrae and soft tissue image around L5: this supported the diagnosis of malignancy.

Figure 2. The pre-operative lumbar magnetic resonance imaging (MRI) of this patient. The pre-operative lumbar magnetic resonance imaging (A, B: Sagittal view; C, D: Axial view) showed the bone destruction of L5 vertebrae and posterior elements, the vague boundary supported the diagnosis of malignancy.

Month period. Physical examination of the patient showed hypoesthesia in the lateral area of her left lower leg, normal sensation in the saddle area, myodynamia of the right hip flexor muscles, and knee extensor muscles weakened to Grade 4 (the myodynamia of other muscles was of Grade 5), limitation of lumbar movement, Abdominal reflex (+), Hoffmann sign (-), Babinski sign (-), and Ankle clonus sign (-). Laboratory findings were as follows: Alpha foetal protein (AFP) 2.66 ng/ml, Cancer embryo antigen (CEA) 0.59 ng/ml, CA15-3 12.34 U/ml, CA19-9 7.00 U/ml, CA-125 11.46 U/ml, CA72-4 10.32 U/ml, CYFRA21-1 1.29 ng/ml, NSE 12.75 ng/ml, ALP 130 IU/L, N-MID 8.1 ng/ml, CRP 55.00 mg/L, ESR 73.0 mm/h, RBC 3.82 × 10¹²/L, HGB 116 g/L, PLT 255 × 10⁹/L, WBC 6.84 × 10⁹/L, ALB 38.7 g/L, and β-HBA 0.31 mmol/L. Lumbar anterior-posterior, lateral, extension and flexion X-rays showed bone destruction of L5 and limitation of lumbar movement. A lumbar computed tomography (CT) scan confirmed the bone destruction of L5 vertebrae and soft tissue around L5 (Figure 1). Lumbar MRI confirmed the bone destruction of the L5 vertebrae and posterior elements (Figure 2). Positron Emission Tomography/Computed Tomography (PET/CT) showed a tumour lesion at L5 and no other tumour-like findings elsewhere. Biopsy pathological examination supported the diagnosis of a melanoma-rich tumour (Figure 3). Chest X-ray, abdominal ultrasonography, and other examinations showed no significant abnormality. Considering the lack of a past history of tumours, no evidence of other tumours...
from the findings of systemic examinations, the patient was diagnosed as having a primary lumbar tumour.

A surgery plan of posterior approach tumour resection, spinal canal decompression, and instrumentation with a pedicle screw system was reached after discussion among several spinal surgeons in our department and effective communication with the patient and her family. The tumour was black, invasive of the L5 vertebra and posterior elements, and it surrounded the anterior part of the L5 vertebra, and involved muscles, tissues, and dura. The patient suffered a massive haemorrhage upon completion of spinal canal decompression and their blood pressure decreased to 50/80 mmHg thereupon. Considering the difficulty of complete resection and the massive haemorrhage, the surgery was changed to partial tumour resection, spinal canal decompression, and instrumentation with a pedicle screw system (Figure 4). The tumour was surgically resected and then the tumour was sent for pathological examination. Pathological haematoxylin-eosin (HE) staining and immunohistochemical staining were performed to determine the nature of the tumour. HE staining showed a melanoma-rich tumour with a large amount of fibrous and granulation tissue present (Figure 5). Immunohistochemical staining results were as follows: S-100 (+), HMB45 (+), A103 (+), PCK (-), EMA (-), CR (-), CK8 (sporadic +), PAX-8 (-), RCC (-), TFE-3 (-), CD 10 (-), CgA (-), Inhibin-α (-), SMA (-), TFEB (-), Ki-67 positive rate 5% (Figure 6).

As it was difficult to make a definite diagnosis, pathologists, radiologists, oncologists, and spinal surgeons in our hospital took a part in a group consultation. At first, difficulties in diagnosis were centred around the differential diagnosis between three kinds of tumour: 1) malignant melanoma; 2) TFE-3/TFEB Wilms tumour; 3) malignant PE-coma. The results of immunohistochemical examination tended to favour the diagnosis of malignant melanoma but pathologists still recommended that we focus on a renal examination to exclude certain rare renal tumours. At the same time pathologists also recommended systemic examinations to confirm a primary malignant melanoma or a metastatic malignant melanoma. According to their suggestions, systemic examinations, including full-body skin examinations, head-MRI, a chest and abdomen
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CT scan, and abdominal and pelvic ultrasound examination, were performed.

Considering no evidence of other tumours from the findings of repeated systemic examinations (including PET/CT), the patient was finally diagnosed as having a primary malignant melanoma. Surgery involving partial tumour resection, spinal canal decompression, and instrumentation with a pedicle screw system was performed and the patient was sent to the oncology department for further treatment including chemotherapy and radiotherapy. No deterioration in neurological function was detected with a follow-up assessment after three months.

Discussion

Malignant melanoma has been reported to be an aggressive malignancy associated with a high mortality rate which often occurs in the skin [3, 4]. According to a report from the National Cancer Research Centre, the incidence of cutaneous malignant melanoma in the Caucasian American population increased from 7.5 per 100,000 in 1973 to 22.5 per 100,000 in 2011, and cutaneous malignant melanoma accounts for 75% of all skin cancer related mortalities in the United States [5, 6]; however malignant melanoma can also occur in other parts such as lung, liver, vagina, oesophagus, and spine (although these incidences were rare) [2, 7-15]. Considering the paucity of knowledge of this area, we present this special case of a huge primary malignant melanoma involved in the L5 vertebrae and peri-vertebral space with emphasis on the difficult diagnosis. At the same time, previously reported primary spinal malignant melanoma cases were also reviewed in the literature (Table 1) [16-21]. Pathological HE staining and HE depigmentation staining were the examination techniques which often showed the presence of melanin granules: immunohistochemical examination plays a crucial role in the diagnosis of malignant melanoma especially in differential diagnoses from other carcinomas such as Wilms tumour and malignant PEcoma. Upon immunohistochemical analysis, malignant melanoma was positive for HMB-45 and S-100; this was important with regard to accurate diagnosis. The diagnosis of malignant melanoma is based on the discovery of melanin granules in the tumour cells upon histological examination, with a positive result for HMB-45, melanoma-specific antigen (Melan-A), and S-100 in immunohistochemical examinations [22, 23]. If it is difficult to reach a differential diagnosis through pathological examination, clinical and radiological examinations can be helpful. Clinical and radiological examinations were also important in determining the nature of any primary, or metastatic, malignant melanoma. Systemic examinations including full-body skin examinations, head-MRI, chest and abdomen CT scan, abdominal and pelvic ultrasound examination, and PET/CT were recommended before making a definite diagnosis.

Primary spinal malignant melanoma is exceptionally rare and most cases are reported as being intraspinal. Diagnosis is difficult because of the unusual tumour location and the variations in its appearance on MRI output: in some
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Figure 6. Immunohistochemical examinations of HMB-45, A103 and S-100. Immunohistochemical examinations showed positive results for A103 (A), HMB-45 (B), and S-100 (C), the diagnosis of malignant melanoma was reached as positive for HMB-45, A103, and S-100 based on immunohistochemical examinations.

Table 1. Summary of the reported cases of primary spinal malignant melanoma

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Age</th>
<th>Gender</th>
<th>Location</th>
<th>Symptom</th>
<th>Diagnosis</th>
<th>Treatment method</th>
<th>Follow-up time</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kobayashi</td>
<td>2012</td>
<td>78</td>
<td>Female</td>
<td>T11 level</td>
<td>Paraparesis and disturbance of bowel movements</td>
<td>Pathological examination</td>
<td>Surgery, Radiation</td>
<td>3 years</td>
</tr>
<tr>
<td>Lee</td>
<td>2010</td>
<td>71</td>
<td>Female</td>
<td>C6-7 level</td>
<td>Left upper extremity tingling sensation</td>
<td>Pathological examination</td>
<td>Partial resection</td>
<td>Unclear</td>
</tr>
<tr>
<td>Kwon</td>
<td>2004</td>
<td>45</td>
<td>Female</td>
<td>C7 level</td>
<td>Neck pain</td>
<td>Histopathological investigation</td>
<td>Surgery</td>
<td>Unclear</td>
</tr>
<tr>
<td>Ryu</td>
<td>2010</td>
<td>55</td>
<td>Male</td>
<td>T4 level</td>
<td>Hypesthesia</td>
<td>Pathology report/ PET-CT</td>
<td>Surgery</td>
<td>Unclear</td>
</tr>
<tr>
<td>Kanatas</td>
<td>2007</td>
<td>76</td>
<td>Female</td>
<td>C6/7 level</td>
<td>Neck pain, paraesthesia, and weakness in her right arm</td>
<td>Pathological examination</td>
<td>Surgery, Radiation</td>
<td>6 months</td>
</tr>
<tr>
<td>Nishihara</td>
<td>2009</td>
<td>49</td>
<td>Male</td>
<td>T6 level</td>
<td>Headache</td>
<td>History + MRI</td>
<td>Radiation, interferon beta</td>
<td>38 months</td>
</tr>
<tr>
<td>This study</td>
<td>2016</td>
<td>42</td>
<td>Female</td>
<td>L5 level</td>
<td>Back pain</td>
<td>Pathology report/ PET-CT</td>
<td>Surgery, Radiation</td>
<td>12 months</td>
</tr>
</tbody>
</table>

cases, even where post-operative histopathological and immunohistochemical examinations were performed, the diagnosis remains difficult, as was the case with this patient. The diagnosis of malignant melanoma is based on the discovery of melanin granules in the tumour cells upon histological examination, with a positive result for HMB-45, Melan-A, and S-100 found after immunohistochemical examination. Systemic examinations including full-body skin examinations, head-MRI, chest and abdomen CT scan, abdominal and pelvic ultrasound examination, and PET/CT were recommended to determine the nature of the primary, or metastatic, malignant melanoma.

Disclosure of conflict of interest

None.

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References

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