PRIMARY MALIGNANT MELANOMA OF THE SPINAL CORD
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PRIMARY melanoma of the spinal cord is a rare cause of compression of the cord. It is the purpose of this paper to present such a case, with an analytical review of 25 cases in the literature, 16 verified by autopsy and 9 surgically verified.

CASE REPORT

G.S. (MH #70748, Autopsy #13900). A 42-year-old white male had sudden onset of constant pain in the left flank about 4 months before admission. The pain lasted for 2 days and was relieved by the passage of a renal calculus. With subsidence of this pain, he experienced low-back pain which radiated anteriorly to the abdomen, and which was relieved somewhat by a corset. During the next few months pain in the back became more severe. A "pins-and-needles" sensation developed and then numbness of both legs and of the pelvic area. There was occasional buckling of his knees, subsequently followed by frank weakness, more marked on the right side. In the month prior to admission, there was some difficulty in initiating urination, as well as some dribbling. He was admitted on Jan. 23, 1955.

Examination. General physical findings were within normal limits. Neurological examination revealed marked weakness of both lower extremities and a wide-based gait. There was a complete sensory level at T12. The myotatic reflexes in the legs were hyperactive, with bilateral Babinski’s sign and right ankle clonus.

Laboratory Data. Hemoglobin, count of red and white blood cells, and urine were within normal limits. Roentgenograms of skull, chest and thoracic and lumbosacral spine were normal, as was the intravenous pyelogram. Lumbar puncture: initial pressure 90 mm. water, with evidence of partial manometric block; total protein 120 mg. per cent; sugar 85 mg. per cent; 9 fresh red blood cells.

Four days after admission, lumbar puncture revealed a partial block on manometry. Myelography immediately thereafter showed complete obstruction to the passage of the contrast material at the level of T10, with the characteristics of an intramedullary tumor (Fig. 1).

Operation. Laminectomy of T7 to T11 vertebrae was performed (by Dr. Sidney W. Gross), with disclosure of an infiltrating intramedullary tumor between T8 and T10. The normal anatomy of the cord was completely obscured, the area being grayish-white with many large veins. On the right lateral surface of the tumor was a black area which, when opened, proved to be a cystic cavity containing approxi-

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Fig. 1. Myelogram, with obstruction to flow of contrast medium in cranial direction at level of T10. (Left) Anteroposterior view: cord appears deviated toward the left. (Right) Lateral view: curved border of column is suggestive of intradural, possibly intramedullary, lesion (note that anteroposterior view has some characteristics of intradural, extramedullary localization).

mately 5 cc. of black material. Following evacuation of this material, pulsations of the cord were noted for the first time.

Pathologic Report. Microscopic sections of the surgical specimen consisted of many fragments of neoplasm. The cells were poorly defined but showed pale, generally ovoid nuclei varying considerably in size. The cytoplasm was pale and eosinophilic, with many fine dark-brown granules of pigment. Diagnosis: malignant melanoma.

Course. There was gradual improvement following surgery, so that the patient could walk with aid. In the 3rd postoperative month, radiotherapy (Cobalt) was started, with a total of 6,000 r given to the T7–L1 area in a period of 2 months. Despite this, his condition gradually deteriorated from the 4th postoperative month, with increase of spasticity in the lower extremities, development of decubiti, urinary-tract infection, and marked hypochromic anemia. Repeated examinations of urine for melanin gave negative results. He died on Aug. 17, 1955, 6½ months after admission.

Autopsy. General anatomical findings. A laminectomy scar was present. Decubiti of buttocks, hips, and legs were noted, with necrosis and fracture of the neck of the right femur. General pathological examination was completely negative for melanotic lesions. This included examination of both eyes (there was a localized chorioretinopathy in one) and microscopic sections of the following organs and structures:
heart, aorta, lung, liver, spleen, gallbladder, cecum, adrenal, kidney, prostate, testis, bladder, lymph nodes, thyroid, pituitary, skeletal muscles, nerves, multiple skin areas, and vertebrae.

Gross neuropathology. The brain and associated structures were all within normal limits.

Spinal cord. The specimen measured 44 cm. in length. The dura mater was normal. The meninges of the cord were discolored dark red-black along the entire thoracic and lumbar cord. In the lower thoracic portion of the spinal cord at T10 and T11, at the site of maximum discoloration, there was a fusiform swelling with a maximum diameter of 1.4 cm. On sections through the swollen area, a firm hemorrhagic black neoplasm was noted, almost completely replacing the substance of the cord, which was visible as a thin band around the periphery of the lesion to one side, and measured about 0.1 to 0.2 mm. in thickness (Fig. 2). Sections through the thoracic cord above the lesion revealed complete ascending and descending demyelination.

Microscopic Examination. Sections of the neoplasm revealed a malignant melanoma. The tumor was disposed in sheets of polygonal cells with poorly defined outlines. The cytoplasm was finely vacuolated and was very faintly basophilic. The nuclei were ovoid and in some areas were elongated to the point of resembling the nuclei of a sarcoma. Nucleoli were prominent and a moderate variation in nuclear size was seen. Mitoses were rare. Abundant melanin was demonstrated with the Fontana stain and bleached in the hydrogen-peroxide preparation. A stain for iron revealed only a few inconspicuous granules of iron salts. Spielmeyer and fat stains of the intact portions of the cord above and below the level of almost complete cord transection by tumor revealed a classical demonstration of ascending and descending demyelination. Diagnosis: Primary malignant melanoma of low thoracic spinal cord and leptomeninges.

ANALYSIS OF THE LITERATURE

This analysis of the literature is based on 25 individual case reports of so-called primary malignant melanoma of the spinal cord (Table 1). The
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Year</th>
<th>Author</th>
<th>Age</th>
<th>Sex</th>
<th>Back Pain Before Cord Compression</th>
<th>Duration of Compression before Admission</th>
<th>Cerebrospinal Fluid Findings</th>
<th>Myelography</th>
<th>Laminctomy</th>
<th>Autopsy</th>
<th>Diagnosed during Life</th>
<th>Remarks</th>
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<td>1</td>
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<td>Hirschberg</td>
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<td>F</td>
<td>+</td>
<td>3 mos.</td>
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<td>2</td>
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<td>Boit</td>
<td>51</td>
<td>M</td>
<td>+</td>
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<td></td>
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<td></td>
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<td>Esser</td>
<td>32</td>
<td>M</td>
<td>+</td>
<td>½ mo.</td>
<td></td>
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<td></td>
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<td>1910</td>
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<td>F</td>
<td>+</td>
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<td></td>
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<td>+</td>
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<td>6</td>
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<td>+</td>
<td>1½ yrs.</td>
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<td>M</td>
<td>+</td>
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<td>61</td>
<td>F</td>
<td>+</td>
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<td>9</td>
<td>1929</td>
<td>Bau-Prussak &amp; Mackiewicz</td>
<td>29</td>
<td>M</td>
<td>+</td>
<td>8 wks.</td>
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<td></td>
<td></td>
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<td>10</td>
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<td>Bell</td>
<td>48</td>
<td>F</td>
<td>+</td>
<td>3 mos.</td>
<td></td>
<td></td>
<td></td>
<td>Yes</td>
<td>Alive</td>
<td>+</td>
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<td>11</td>
<td>1930</td>
<td>De Blasi</td>
<td>71</td>
<td>F</td>
<td>+</td>
<td>6 mos.</td>
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<td></td>
<td></td>
<td>Defect</td>
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<td>12</td>
<td>1933</td>
<td>Van Bogaert &amp; Verbrugge</td>
<td>38</td>
<td>M</td>
<td>+</td>
<td>6 mos. Xantho.</td>
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<td>13</td>
<td>1938</td>
<td>Schnitker &amp; Ayer</td>
<td>49</td>
<td>F</td>
<td>+</td>
<td>Xantho. 85 mg. %</td>
<td></td>
<td></td>
<td></td>
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**Table 1**

*Malignant melanoma of the spinal cord (26 cases)*
<table>
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<tr>
<th>Patient</th>
<th>Year</th>
<th>Age</th>
<th>Duration</th>
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<th>Treatment</th>
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<td>14</td>
<td>1939</td>
<td>55</td>
<td>2 yrs.</td>
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<td>Yes</td>
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<td>15</td>
<td>1941</td>
<td>52</td>
<td>3 wks.</td>
<td>Xantho.</td>
<td>Yes</td>
<td>No block</td>
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<td>16</td>
<td>1942</td>
<td>32</td>
<td>10 mos.</td>
<td>Xantho.</td>
<td>66 mg. %</td>
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<td>190 mg. %</td>
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<tr>
<td>17</td>
<td>1950</td>
<td>52</td>
<td>1 yr.</td>
<td>Xantho.</td>
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<tr>
<td>18</td>
<td>1950</td>
<td>25</td>
<td>2 mos.</td>
<td>Xantho.</td>
<td>Yes</td>
<td>Block</td>
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<td>19</td>
<td>1950</td>
<td>57</td>
<td>?</td>
<td>Xantho.</td>
<td>4,500 mg. %</td>
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<tr>
<td>20</td>
<td>1951</td>
<td>47</td>
<td>2 mos.</td>
<td>Black</td>
<td>2,400 mg. %</td>
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<tr>
<td>21</td>
<td>1951</td>
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<td>Yes</td>
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<td>22</td>
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<td>53</td>
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<td>23</td>
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<td>1954</td>
<td>50</td>
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<td>51</td>
<td>?-1 day</td>
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<td>42</td>
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Xantho. = Xanthochromic.
age of the patients ranged from 25 to 71, the median age being 49. There were 12 males and 13 females.

1. **Preoperative Findings.** Admission to the hospital in all cases was prompted by spinal-cord symptomatology, with all but 4 patients referring their complaints to the lower extremities. Pain was frequent and severe in 20 cases. The duration of symptoms of cord compression prior to admission varied from 3 weeks to 2 years with the mean at 6 months. General physical findings were within normal limits. Neurologic examination revealed a level of the lesion in 21 cases. This consisted of severe sensory, motor, and sphincteric involvement, with typical paraparesis or paraplegia. Tetraparesis or tetraplegia was present in 4 patients. Roentgenograms of the spine were normal with the exception of 1 patient16 who displayed erosion of the vertebral pedicle at the level of the tumor (subsequent laminectomy showed extradural extension of tumor and invasion of bone). Lumbar puncture was performed in 17 patients and was abnormal in all but 2 cases. Xanthochromia and elevated protein were common, while 5 patients had pleocytosis, varying between 27 and 300 lymphocytes. Manometric block was noted in 7 instances. Myelography was performed in 10 patients and was abnormal in 9; a complete block was found in 7 patients and defects were outlined in the other 2.

In only 1 case, that of King and Propst,17 was a preoperative diagnosis of malignant melanoma made on the basis of "coal-black" cerebrospinal fluid obtained during myelography, which demonstrated a complete block at the L1 vertebral level.

2. **Surgery.** Of the 25 patients, 19 underwent surgical exploration (Fig. 3). In 14 cases tumor was found and removal was attempted. Histological examination of the surgical specimen established the diagnosis in all cases, except that of Schnitker and Ayer,28 which was thought to have been a case of atypical meningioma. Complete extirpation was accomplished in 3 cases of extramedullary tumor and in 1 case of extradural neoplasm. There were 2 negative explorations: in one, the laminectomy was performed at the wrong level and the lesion subsequently was verified at autopsy;29 in another,19 tumor was not found but the cord appeared large and hyperemic and did not pulsate; exploration was felt to be negative for tumor, but an intramedullary tumor was found at autopsy 5 months later.

At operation, when such a black mass was detected, the usual confusion was with a vascular lesion. Malignant melanoma of the central nervous system is often hemorrhagic and grossly it may be difficult to distinguish between hematomat and melanoma, requiring histological examination. In De Blasi’s patient,9 a suspected intramedullary hematoma was removed from the cord at T8, with no histological findings described for the surgical specimen; subsequently, an intramedullary malignant melanoma was disclosed at autopsy. A vascular malformation was felt to be present in the patient of Forbes and Maloney,11 although no attempt was made to remove the lesion; two separate intramedullary lesions were found at necropsy with diffuse leptomeningeal dissemination of tumor.
3. **Prognosis.** In 6 nonsurgical cases and 5 surgical cases without removal of tumor there was a progressive course with survival periods varying from 1 to 15 months with a mean at 2 months; death was usually caused by bronchopneumonia or infection of the urinary tract. Seven patients who had partial removal of tumor survived from 1 to 10 months after operation.

In 4 cases complete removal of tumor was performed. Death occurred in the operating room immediately after surgery in 1, and a second patient expired a few days following operation. In these 2 instances, the operative procedure itself was reported to be uneventful and the cause of death remained unsolved. In a third case, the original surgical specimen was called an atypical meningioma. However, the patient was rehospitalized 6 months postoperatively with severe cerebral involvement and died 2 weeks later from bronchopneumonia. Autopsy showed diffuse leptomeningeal dissemination (a minimal amount of melanin was found in the original surgical specimen). A fourth patient was alive at the time of the report and showed a rapid and practically complete recovery; the follow-up period after operation was not given.

Radiation was ineffective in all patients treated, although there were reports of 5 patients alive and improved following treatment; the follow-up periods were short, however. The longest follow-up was that of Kissel *et al.*, the patient showing excellent recovery 6 months after operation and radiotherapy; recurrence of tumor in situ was evident 5 months later. Furthermore, the patient in Case 14, reported later by Moersch *et al.*, was in critical
condition 4 months later. In the remaining 3 cases of patients who were alive at the time of report, the operative notes described encapsulated or apparently discrete meningeal tumors without invasion of cord substance. The histological descriptions were too brief for an accurate diagnosis; however, they did not seem to be malignant and a melanin-pigmented meningioma could not be ruled out completely. If the outcome of the case of Schnit-ker and Ayer\textsuperscript{28} is considered, originally thought to be one of pigmented meningioma but in which death occurred from leptomeningeal dissemination, it is obvious that a correct diagnosis cannot be made without further follow-up reports. Two cases of melanin-pigmented meningioma in the literature are not included in this series, since the postoperative course and survival were characteristic of a benign neoplasm and the histologic appearance was in all ways compatible with that of meningioma. Likewise, it should be noted that these patients all had excellent results following removal and were followed for periods of 5 years\textsuperscript{24} and 8 years.\textsuperscript{1}

4. Autopsy. Postmortem examination was performed in 16 cases. The level and position of the tumors in the cord at autopsy are indicated in Fig. 4. A diagnosis of melanoma of the spinal cord was not made until autopsy in 6 nonsurgical and 5 surgical cases.\textsuperscript{9,11,13,19,25} All patients in whom melanoma was discovered at operation had an extensive survey without detection of a primary lesion during life. Five of such patients came to autopsy, including 2 who had had “total removal”;\textsuperscript{28,29} tumor was found at the operative site without further extension.

Two patients,\textsuperscript{13,28} who had development of cerebral symptomatology
PRIMARY MALIGNANT MELANOMA OF SPINAL CORD

terminally showed diffuse spinal leptomeningeal dissemination extending into the basilar cerebral leptomeninges, without actual invasion of the cerebral parenchyma; both patients became blind. In 3 additional cases there were similar postmortem findings without noticeable clinical evidence of cerebral involvement. Although all 5 cases presented diffuse leptomeningeal dissemination, the level of cord involvement did not change. The remaining 10 cases in which the brain was examined did not demonstrate lesions.

General autopsy was performed in 15 cases. Extraneural metastases were described in 2: to lung, liver and uterus, and to liver and spleen. Two patients had benign pigmented nevi on the skin which the respective authors considered of no significance. The 11 remaining autopsies failed to show any lesion outside of the central nervous system.

DISCUSSION

Starting with the initial report of Virchow many authors have discussed primary malignant melanoma of the central nervous system, and a little over 60 cases have been reported up to the present time. These authors affirmed the concept of primary malignant melanoma of the central nervous system as a definite pathological entity with origin most likely in the meninges of the brain and cord. It is uncommon to find a patient presenting solely with symptoms of cord compression caused by melanoma. Indeed, malignant melanoma metastatic to the spinal cord and coverings occurred only once (epidurally) in a series of 127 metastatic lesions of the spinal cord verified by autopsy at this institution; in this case there were widespread metastases to all organs.

The 25 cases reviewed herein were considered to be primary melanomas of the spinal cord by each of the respective authors, although, as noted above, in only 10 cases was the brain normal at autopsy. In order to substantiate such a diagnosis, it is necessary to have a complete postmortem examination (including the eyes and the entire central nervous system and coverings), and thereby prove both the absence of a malignant extraneural pigmented lesion, and of a cerebral lesion. In only 9 instances in this series was such a complete postmortem examination performed, leaving some doubt concerning the primary nature of the remainder of the cases.

SUMMARY

1. A case of primary melanoma of the spinal cord is presented and the previously recorded 25 cases are analyzed with respect to symptomatology, neurological findings, operation, and autopsy findings.

2. The present case is the only recorded instance in which a diagnosis of a primary intramedullary melanoma of the spinal cord was made at operation and biopsy, and in which subsequent complete postmortem examination proved it to be the sole lesion in the central nervous system.

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REFERENCES