Primary Malignant Melanoma of the Cervical Spinal Cord

—Case Report—

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Primary malignant melanom of the spinal cord is extremely rare. The best treatment appears to be total surgical excision, when possible and postoperative irradiation, but the value of radiotherapy is unknown because the number of cases that have been irradiated postoperatively is small.

The 2-year survival rate for primary malignant melanoma of the spinal cord is usually less than 16% and most patients died within 1 year.

The prognosis of this tumor is poor.

Key Words: Primary malignant melanoma of the spinal cord, Radiotherapy.

INTRODUCTION

Malignant melanoma of the central nervous system is rare. Especially, primary malignant melanoma of the spinal cord is extremely rare and only few cases has been reported[1~5], one of which involved the thoracic and cervical spinal cord.

It can be argued that the primary central nervous system lesion could actually be a metastasis from an occult primary site in the skin, retina, esophagus, rectum, or other site[5].

Malignant melanoma is the thrid most common cause of central nervous system metastases next to carcinoma of the breast and bronchus[6]. The incidence ranged from 1.4% to 15.7% among all patients with malignant diseases[7~9].

Survivals following resections of primary melanoma of the spinal canal has been reported in a few cases[4,10]. Unfortunately, most patients with melanoma of the central nervous system have many of diffuse lesions[11,12] and surgical treatment is not feasible.

Various approaches have been tried in managing patients with malignant melanoma of the central nervous system; surgery, radiotherapy, and corticosteroids, or/and immune therapy. A combination of corticosteroids and radiation can provide substantial palliation in some patients with melanoma of the central nervous system[13].

The prognosis of this tumor is poor.

CASE REPORT

A 20-year-old man was admitted to Korea University Hospital in January 1986, because of fever, headache, and urinary retention. 1 month prior to admission, he noted the insidious onset of multiple myalgia on the upper extremities, the neck stiffness, and the weakness of the left leg.

The general physical examination was normal. The neurological examination showed the mydriasis of the left eye, the numbness of the left arm, and the urinary retention. Hypalgesia below T-3 dermatome was demonstrated on the left side of the body. The patellar and Achilles reflexes were intact.

Lumbar puncture revealed bloody CSF.

A complete blood cell count, urinalysis, LFT, chest X-ray, skull X-ray, and bone scan were all normal. X-rays of cervical spine showed the stiffness of normal curve of cervical spine but the disc spaces were well preserved. Brain CT scan was negative.

Metrizamide myelogram was performed and showed that there was complete obstruction of metrizamide dye column at the level of upper border of T-1 vertebral body and intradural extramedullary mass lesion on the right (Fig. 1). Cervical spinal CT scan was performed and revealed an Dumb-ball shaped mass on right intervertebral foramen between C-7 and T-1 vertebral

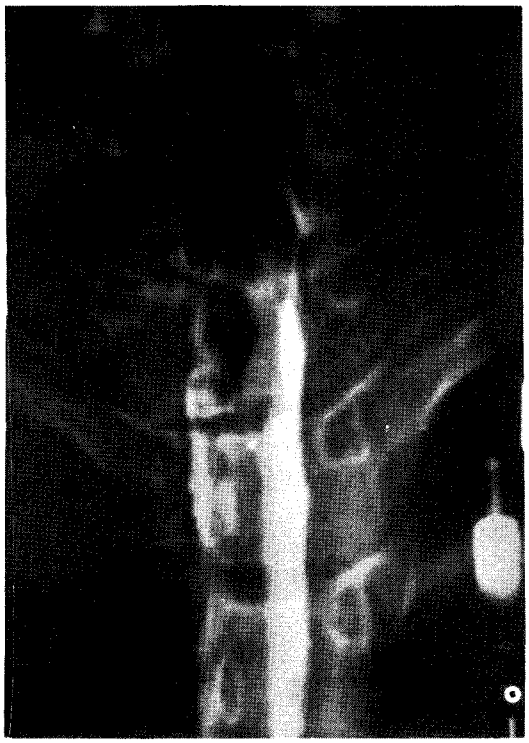

Fig. 1. Metrizamide myelogram shows a complete block at the level of T-1 which suggestive of intradural and extramedullary space occupying lesion.

bodies and the cord was deviated to the left side (Fig. 2).

Spinal angiogram shows hypervascular tumor staining in the right site of paraspinal area at the level of C-7/T-1 with tortuous enlarged venous drainage into the upper thoracic vein.

He underwent the laminectomy of C-6 through T-1 on January, 24, 1986 and the spinal cord was found to be displaced to the posteriorly and leftward by the tumor mass. The mass was dumb-

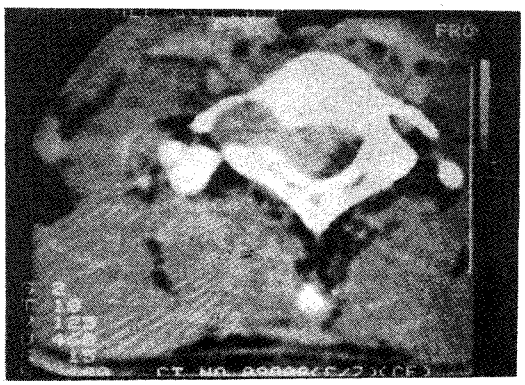

Fig. 2. Cervical spinal CT scan prior to operation : there is evidence of a dumb-bell shaped mass on the right intervertebral foramen through the level of C-7 and T-1, and the spinal cord was deviated to the left side due to the mass.

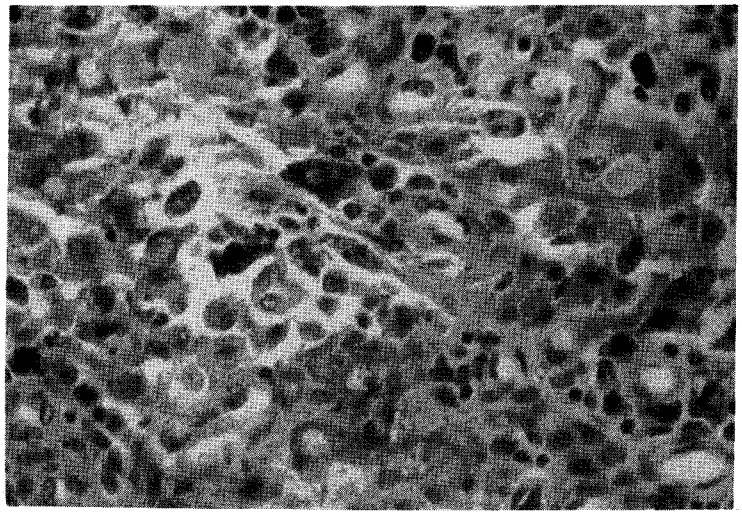

Fig. 3. High-power view of the photomicrograph of tumor mass reveals many pleomorphic cells with melanin pigments, compatible with diagnosis of melanoma.

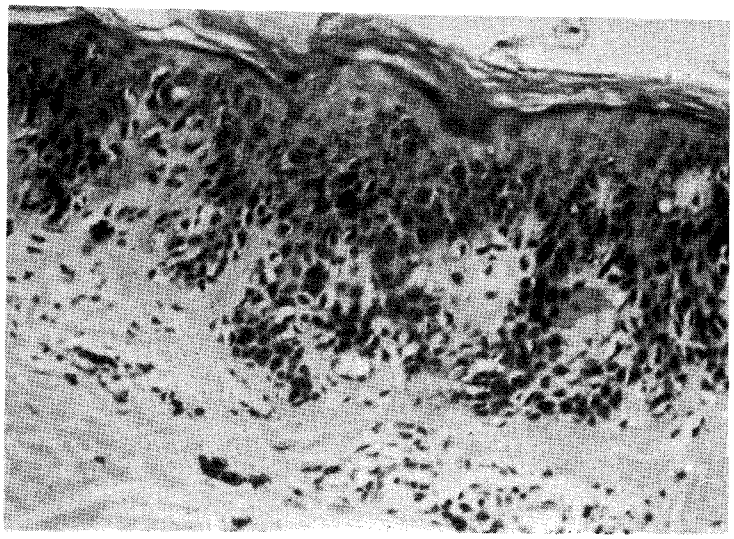

Fig. 4. Photomicrograph of the trunk showed deposition of atypical melanocytes in the basal layer with pigmentation which indicates in sity lesion.

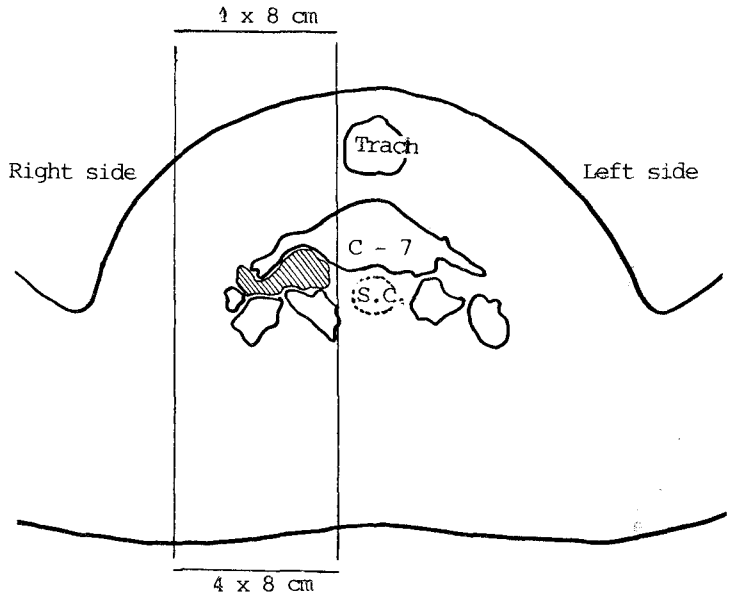

Fig. 5. At the dose of 3,600 cGy, cone-down boost field to the residual tumor area which was treated AP and PA parallel opposing portals by 4 x 8 cm.

bell shaped, and measuring about 2 x 1 cm in size, located half between inside and outside of the spinal canal. Tumor attached to the lateral surface of the dura and tumor mass extended to the extradural portion through the neural foramen. Intradural portion of the tumor was removed totally, but extradural portion was excised partially. After laminectomy, the urinary symptom and the numbness of the left arm improved.

Histopathological study identified the tumor as a

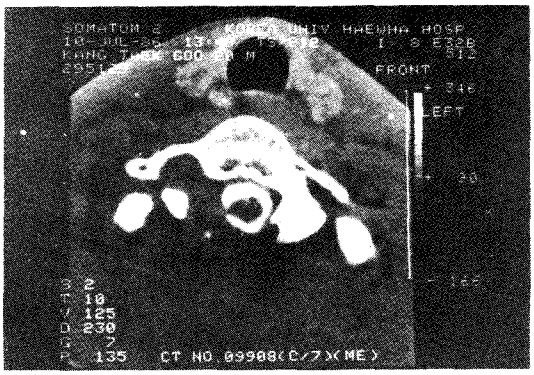

Fig. 6. 3 months follow-up spinal CT scan after completion of postoperative RT : there is no evidence of tumor growth and no evidence of cord deviation.

malignant melanoma which sections of the specimen showed many pleomorphic cells with melanin pigments (Fig. 3).

Brownish pigmented lesion was present on the posterior aspect of trunk, which was about 2 x 5 cm in size. On February, 1, 1986, he underwent the excisional biopsy of the pigmented lesion of the trunk. Microscopic sections of the specimen revealed the atypical melanocytes in basal layer with pigmentation and the lesion is limited to the basal layer (Fig. 4). The diagnosis was malignant melanoma in situ. Therefore, the previously excised spinal tumor may be considered as the primary.

He received the postoperative irradiation with Co-60 external beam therapy via the parallel opposing anterior and posterior portals. The upper border of treatment field was the 4th cervical spine and the lower border of treatment field was the 5th thoracic spine.

At the tumor dose of 3,600 cGy/18 f/3 1/2 weeks, the field size was reduced and boost to the tumor area a total tumor dose of 6,600 cGy/33 f/6 1/2 weeks (Fig. 5). The patient tolerated his treatment very well and had no specific neurological signs during radiotherapy.

3 months after the completion of radiotherapy, follow-up spinal CT scan was obtained and showed that there was no evidence of growing of the residual mass and no cord compression (Fig. 6). Now, he is alive well, and attends university as a student and has no signs of recurrence for 13 months after radiotherapy.

DISCUSSION

Virchow was the first to report primary malignant melanoma of the central nervous system[14]. Pigmented tumors of the central nervous system may be classified into three types: 1) primary melanoma, 2) metastatic melanomas, and 3) melanotic variants of various types of central nervous system tumors[15].

Melanin-containing cells are a normal constituent of many sites beside skin-iris, ciliary body, choroid and retina of the eye, chromaffin tissue (carotid body and adrenal gland), substantia nigra and the meninges. Gibson, et al, pointed out that the pial melanoblast may be the cell of origin for a primary melanoma in the central nervous system[16]. Masson showed that dermal melanomas originate from specialized cells in the sensory nerve endings[17,18].

In the spinal cord, pigmentation was mostly in the anterior median fissure and between the anterior and posterior roots.

Primary central nervous system melanoma can metastasize to the brain, spinal cord, liver, and others[5,19].

Analysis of ages reveals a peak incidence in the fourth decade, with a secondary peak noted in the first 10 years of life[16]. No particular predilection of sex was noted.

The differential diagnosis of pigmented lesion in this location is limited to three considerations: 1) pigmented meningiomas, 2) hematoma (hematocele), and 3) metastatic melanoma.

Treatment for this tumor is not very successful, for only 16 per cent of the patients have a course of longer than 2 years[4]. The best treatment appears to be total surgical excision, when possible. Because the number of cases that have been irradiated postoperatively is small, therefore the value of radiotherapy is unknown.

The usual progress of these melanomas is inexorable.

After originating in the lepto meninges, typically about a nerve root, they tend to invade the adjacent neural parenchyma and spread along the subarachnoid space as a tumor meningitis[20].

The prognosis of this tumor is poor; most patients succumb in less than 1 year.

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═ 국문초록 ═

경추 척수의 원발성 흑색종양

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김 철 용 · 최 명 선

척수의 원발성 흑색종양은 매우 희귀한 것으로 알려져 있다. 이러한 종양의 가장 유효한 치료로서는 가능한 종양을 완전히 제거하는 것이며, 수술 후에 방사선치료를 같이 병행하고 있으나 증례가 적어서 방사선치료의 효과는 아직 잘 알려져 있지 않다.

치료후 2년 생존율은 16% 이하로 극히 저조하며, 대부분이 1연 내에 사망하는 아주 예후가 나쁜 종양으로 보고되고 있다.