Spinal Metastasis of Medulloblastoma in Adults: A Case Report

Nenad Živković, Iva Berisavac, Marko Marković, Sanja Milenković
1Department of Neurosurgery, Clinical Hospital Center Zemun, Belgrade, Serbia; 2Department of Clinical Pathology, Clinical Hospital Center Zemun, Belgrade, Serbia

INTRODUCTION
Medulloblastoma is a primitive neuro-ectodermal malignant tumor most commonly seen in childhood and rarely and uncommonly in adult age, comprising only 0.4% of all adult tumors [1, 2]. It can metastasize extracranially through the cerebrospinal pathways rarely intra- or extradurally or hematogenously mostly involving bones. Although these metastatic tumors are uncommon in adult age, there is no universal accepted treatment strategy. The treatment consists of surgery, following radiotherapy and, in high risk patients, adjuvant chemotherapy. Regarding new strategies, there are three studies concerning the treatment of recurrent medulloblastoma with temozolomide [3].

We present a case of medulloblastoma metastasizing to the thoracic spine and describe the magnetic resonance and intraoperative finding.

CASE REPORT
A 45-year-old female patient was operated on in June 24, 2011 because of cerebellar medulloblastoma of the fourth ventricle. The tumor was totally removed and pathohystological examination confirmed medulloblastoma. During the postoperative period patient’s general and neurological status were intact. She received cranial irradiation a month later that was followed after one year. During that period control computer tomography (CT) scan did not show any sign of tumor recurrence and she was without any neurological deficit.

Seven days before she was readmitted to our hospital, on April 5, 2012, she complained of a sudden weakness in her legs which progressed into paraplegia of lower extremities and sphincter incontinence. Hypoesthesia of the level Th8 and no reflexes in the legs were also found during neurological examination. Magnetic resonance imaging (MRI) of the spine showed a total block in the thoracic spine with intradural extramedullar mass (Figure 1).

The patient was operated on in April 12, 2012 and total Th8-Th12 laminectomies were performed with no epidural lesions. Dura mater was opened with 5 cm linear cut and an intradural tumor solid mass was seen compressing the medulla (Figure 2). The tumor was extirpated (Figure 3) and histopathological examination showed primitive neuroectodermal tumor characterized by small, round and anaplastic cells compatible with medulloblastoma (Figure 4). Individual tumor cells were small with little cytoplasm and hyperchromatic nucleoli, frequently elongated or crescent shaped. After surgery neurological status did not change during the postoperative period. The patient died three weeks later.

DISCUSSION
Medulloblastoma, or primitive neuroectodermal tumor of the cerebellum, accounts for approximately 20% of all childhood brain tumors and is rarely seen in adults [1, 2, 3]. In 1936 Nelson [4] reported the first well-documented case of metastasis in a patient with cerebellar medulloblastoma. Although medulloblastomas spread along the CSF pathways other accepted modes of tumor spread to the spinal cord may include hematogenous routes [5, 6]. Usually they metastasize to the spinal cord on the
surface of the leptomeninges [6]. Sometimes spinal dural infiltration may occur in the absence of local recurrence or systemic metastases and can suggests the first focus of systemic metastasis [1]. Our patient had neither tumor recurrence in the posterior fossa nor systemic extra axial metastases. The patient had a diffuse involvement of thoracic spine canal which is rare according to the literature [2, 6], i.e. spinal infiltration was the first sign of metastasis. Although rare, seeding at presentation is a poor prognostic factor. Zumpano [7] and Barnwell and Edwards [8] suggested that metastatic spread of medulloblastoma can be explained on the basis of direct extension from a primary cerebellar medulloblastoma to an enlarged central canal.

The average time until the development of metastasis after the initial diagnosis of medulloblastoma is 18 months [1]. In some cases, 13 years had elapsed after initial diagnosis before metastasis developed [1, 9, 10]. In the case of our patient the tumor relapsed in the spinal axis 11 months later after the first presentation in the posterior fossa.

CT and MRI can be used in the diagnosis of metastatic intramedullary tumors.

Fluorodeoxyglucose positron-emission tomography/computer tomography (FDG-PET/CT) is a useful tool to evaluate metastasis in medulloblastoma. One FDG-PET/
CT study [11] revealed abnormal FDG from the cervical to the lumbosacral level suggesting the possibility of metastasis. MRI can be used in the diagnosis of metastatic intramedullary tumors and has more diagnostic value than other diagnostic procedures [12]. MRI gives detailed anatomic information and extent of lesions and it is important for further treatment planning such as radiation therapy or systemic chemotherapy.

In our patient MRT with T2 of sagittal thoracic spine showed an enhancing nodule and diffuse leptomeningeal enhancement consistent with metastatic medulloblastoma. The treatment consists of surgery, following radiotherapy and, in high risk patients, adjuvant chemotherapy. The first step is radical surgery; adjuvant radiotherapy must be 55 Gy on the posterior fossa and the remaining cranial-spinal axis; adjuvant chemotherapy may be useful in patients at high risk of recurrence. The median time to relapse was 23.5 months [13]. Late recurrences are also described in literature. In our case, the patient underwent second-protocol adjuvant radiotherapy for medulloblastoma treatment with good outcome for 11 months.

The survival results for medulloblastomas in adults compare favorably with those in children [14]. Spinal seeding is a poor prognostic factor for disease-free survival. A minimal dose of 54 Gy to the posterior fossa is essential for adequate tumor control. The interval between surgery and the start of RT is a significant prognostic factor and requires further study [13, 14, 15]. Although rare, the presence of metastasis is a poor prognostic factor. Treatment options for patients with metastases are limited and their prognosis continues to remain poor.

REFERENCES


Figure 3. Intraoperative images obtained after resection of thoracic extramedullary metastatic medulloblastoma

Figure 4. Microscopic image: small, round and anaplastic cells in a diffuse pattern (hematoxylin-eosin, original magnification ×100)
Спинална метастаза медулобластома код одраслих особа: приказ болесника

Ненад Живковић1, Ива Берисавац1, Марко Марковић1, Саня Миленковић2
1Неурохируршка служба, Клиничко-болнички центар „Земун”, Београд, Србија;
2Служба клиничке патологије, Клиничко-болнички центар „Земун”, Београд, Србија

КРАТАК САДРЖАЈ
Увод Медулобластом је примитивни неуроглокодермални малигни тумор који се најчешће јавља у детинству, а рекола је уз одраслих особа. Пациент је хируршко уз зрачну терапију. Уколико дође до рецидива, нема општеприхваћеног начина лечења. Метастазе тумора могу се видети у мрежама чворова, меким ткима, костима и удаљеним органима.

Приказ болесника У овом раду приказан је 45-годишња жена с торако-спиналним, екстрамедуларним, метастатским медулобластомом. Описани су прогресивно неуроглокодермално погоршење уобичајено 11 месеци након прве операције, налаз магнетне резонанције и интраоперациони налаз болеснице.

Закључак Премда ретка, метастаза је слаб прогностички фактор. Опције за лечење болесника с метастазама су ограничена, а њихова прогноза је и даље поша. Кључне речи: медулобластом; спиналне метастазе; хируршко лечење

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