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Olgu Sunumu

Cerebellopontine Angle Meningioma as a Long-term Complication of Cranial Radiotherapy Used for the Treatment of Posterior Fossa Medulloblastoma: Case Report and Review of the Literature

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Medulloblastoma is one of the most common primary malignant brain tumors of central nervous system in children. The recommended treatment of this malignant neoplasm is aggressive surgery, followed by craniospinal radiotherapy (RTP) with/without chemotherapy. Radiation-induced meningioma is the most common brain neoplasm known to be caused by ionizing radiation. This report presents a patient who had undergonde an operation 25 years ago because of a cerebral mass lesion. The patient had experienced unsteady gait associated with nausea and vomiting 25 years ago. Cranial CT demonstrated a large, solid and contrast- enhanced midline vermian mass. Through midline suboccipital craniectomy mass was removed totally which was histopathologically diagnosed as desmoplastic medulloblastoma. The patient was given postoperative adjuvant RTP. The patient tolerated RTP well and she had been called for yearly controls for 25.5 years. The patient who had not any complaint at routine controls for 25.5 years had been brought to our hospital with headache, unsteady gait, nausea and vomiting. MRI demonstrated an isointense mass in the right cerebellopontine angle that consisted of two compartments; the medial compartment did not show contrast enhancement and the lateral compartment that was associated with dura showed moderate, and homogenous contrast enhancement The patient had been managed surgically using right suboccipital craniectomy followed by total resection of the lateral compartment and biopsy of the medial compartment was obtained which was thought to be radiation-induced fibrotic tissue. The mass was diagnosed histopathologically as fibroblastic meningioma WHO Grade I and the biopsy of medial compartment showed to be arachnoidal fibrosis. The patient was well and had no recurrence öat postoperative 30 months.

Keywords: Radiation-induced meningioma, medulloblastoma in children, late effect of radiation therapy

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Posterior Fossa Medülloblastomun Tedavisinde Kullanılan Kraniyal Radyasyon Geç Komplikasyonu Olan Serebellopontin Köşenin Menenjiomu: Olgu Sunumu ve Literatür Taraması

Medülloblastom çocukluk çağında primer merkezi sinir sistem tümörlerinin en sık rastlanan malign beyin tümörlerinden biridir. Bu malign tümörün tedavisinde agresif cerrahi sonrasında kraniyospinal radyoterapi (RTP) ve/veya kemoterapi önerilmektedir. Radyasyonla ilişkili menenjiom, iyonize radyasyonun sonucu olarak en sık rastlanan beyin tümörüdür. Bu makalede, beyin kitlesi nedeniyle 25 yıl önce ameliyat geçiren hasta sunulmuştur. Yirmi beş yıl önce dengesiz yürüyüşle birlikte bulantı ve kusması vardı. Kraniyal BT'sinde büyük, sert, kontrast tutan orta hat vermiyan kitle izlenmiştir. Orta suboksipital kraniyektomi yapılarak histopatolojik olarak desmoplastik medülloblastom tanısı konulan kitle total olarak çıkartılmıştır. Hastaya ek olarak postoperatif RTP verildi. Hasta RTP'yi iyi tolere edilip 25,5 yıldır yıllık kontrollere çağırıldı. Bu rutin kontrollerde yakınması olmayan hasta 25,5 yılın sonunda baş ağrısı, dengesiz yürüyüş, bulantı ve kusma ile hastanemize getirildi. Çekilen MRG'inde sağ serebellopontin köşesinde iki komponentli izointens kitle görüldü. Kitlenin mediyal komportmanı kontrast tutulumu göstermemişken dura ile ilişkili lateral komportmanı ise homojen olarak kontrast tutulum göstermiştir. Hastaya sağ suboksipital kraniyektomi yapılarak lateral kompartmanın total rezeksiyonu ve radyoterapiye bağlı gelişen fibroz dokudan şüphe edilen mediyal kompartmandan biyopsi alındı. Kitlenin lateral kompartmanı histopatolojik olarak fibroblastik menenjiom (WHO grade I) ve mediyal kompartmanın biyopsi sonucu araknoidal fibroz olarak tanı konulmuştur. Hasta 30 aylık takibinde iyidir nüks saptanmamıştır.

Anahtar kelimeler: Radyasyonla ilişkili menenjiom, çocuklarda medülloblastom, radyoterapinin geç etkisi

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INTRODUCTION

Medulloblastoma is one of the most common malignant brain tumors in children, accounting for 20-30% of all childhood brain tumors [6]. The recommended treatment of these entities is aggressive surgery that has to be followed by adjuvant treatments as craniospinal radiotherapy (RTP) with/without chemotherapy. More than half of the children with medulloblastoma can be expected to be free of disease 5 years later [6].

Some of the delayed side effects of irradiation on neural tissue include visual deterioration, hearing loss, hormonal disturbances, fatty replacement of bone marrow, and the induction of new CNS neoplasms [9]. A lot of neoplasms may develop after cranial curative RTP. Indeed, cavernomas, meningiomas, sarcomas, astrocytomas, and schwannomas have been described. Children with medulloblastoma and treated using RTP after surgery, appear to be particularly sensitive to develop RIMs.

The marked rarity of pediatric post-radiation meningiomas compared with their adult counterparts could be due to the fact that meningioma is a slow-growing tumor with late manifestations, and an average latency period of 18.1 years [12].

This report describes a rare case of 38-year-old female patient diagnosed as a RIM, 25 years after curative radiotherapy for desmoplastic (nodular) medulloblastoma.

CASE REPORT

A 38 year-old female patient had undergone craniectomy to remove the midline vermian mass (Figure 1) that caused unsteady gait, headache, nausea and vomiting at age of 13 in 1988. After histopathological examinations revealed that the mass was desmoplastic (nodular) medulloblastoma (Figure 2), she subsequently received high-dose curative craniospinal RTP at a dose of 50 Gy directed at CNS, and the posterior fossa as a focal boost dose of 15 Gy. The patient tolerated RTP well and she had been controlled at yearly

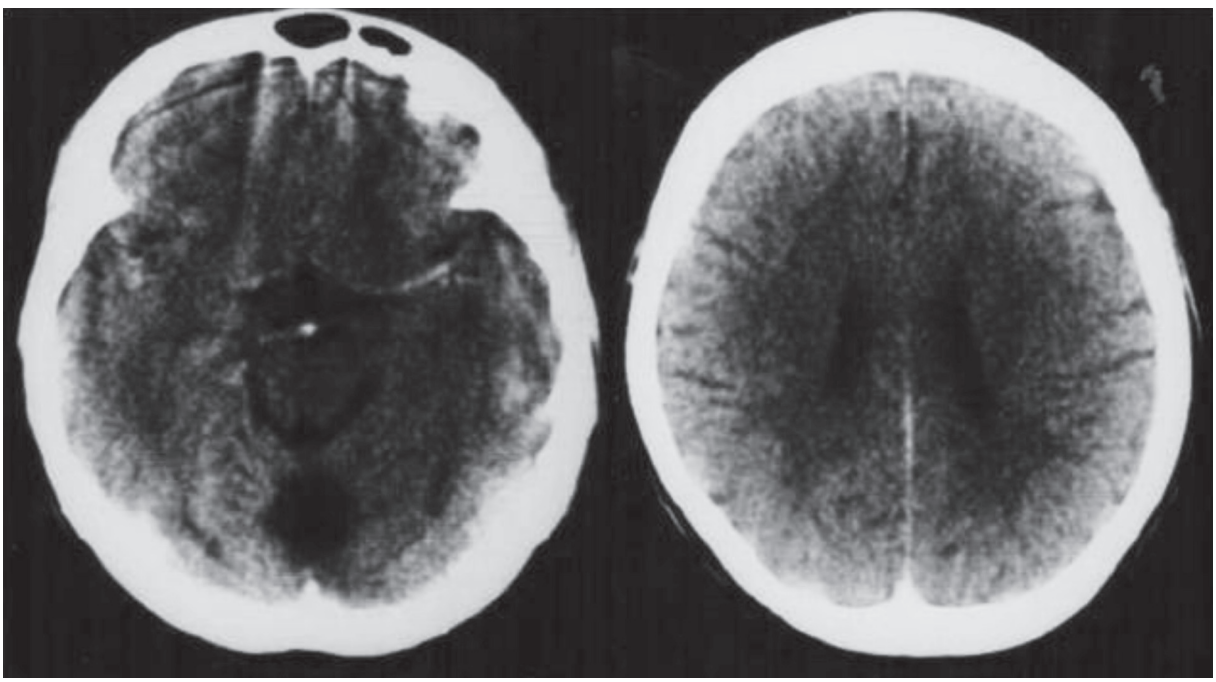

Figure 1. Postoperative computed tomography (CT) which performed after the first surgery in 1988. Note postoperative changes such as the area of encephalomalacia at the middle line of vermian.

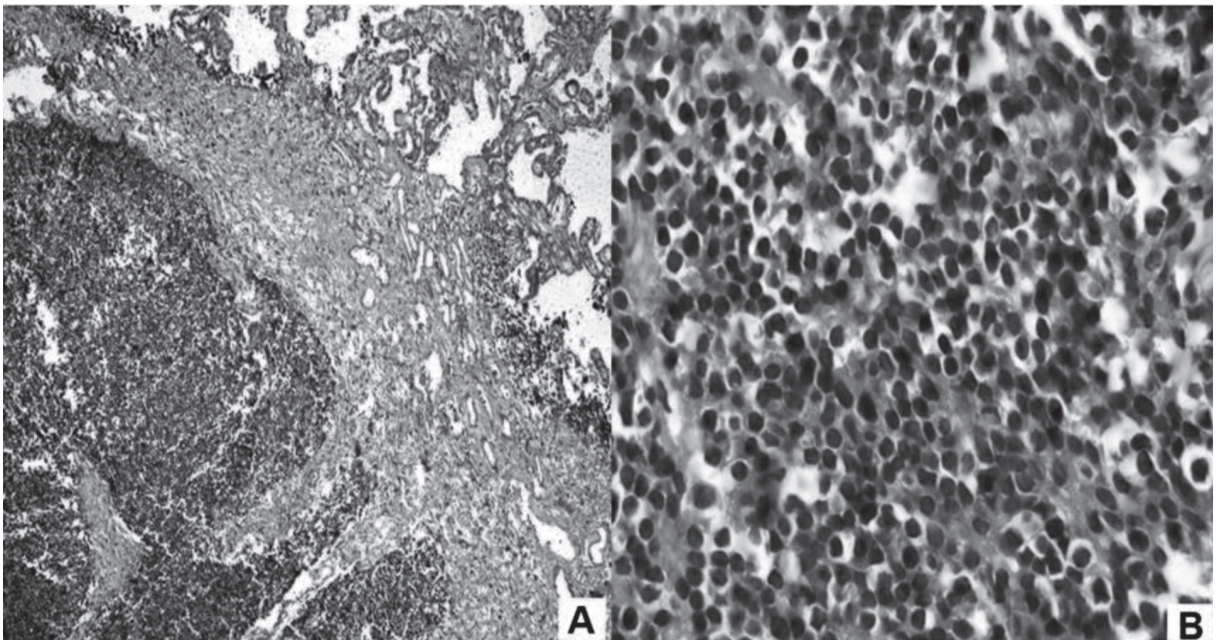

Figure 2. Histolopathological findings of the middle line vermian mass tissue that obtained from patient in 1988, these findings led to diagnose the tumor as desmoplastic (nodular) mdulloblastoma (WHO Grade IV). (A): At the right top in this photo there is choroid plexus and at the left bottom an infiltration of the round tumor cells is seen. (B): Malignant round cell tumor at the high power (HE x200).

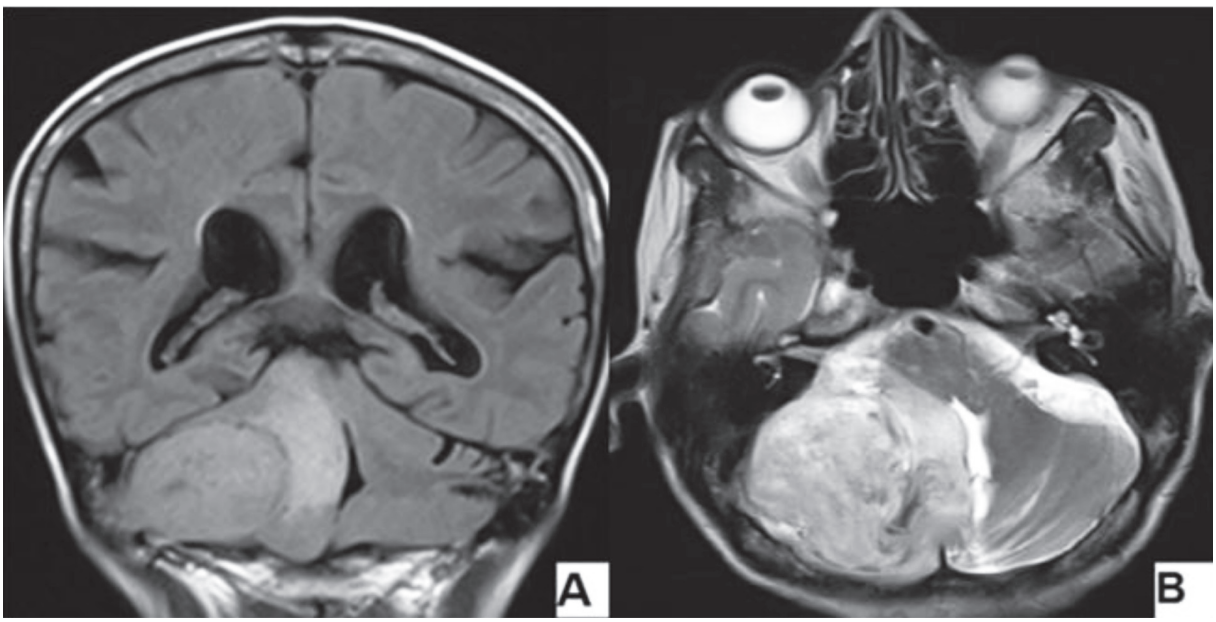

Figure 3. Preoperative magnetic resonance imaging (MRI) performed in last year control before surgery of RIM. (A): Preoperative coronal T2-weighted fluid-attenuated inversion recovery (FLAIR) MRI showing a tumor of right CPA that measures 5.5x7x3 cm. (B): Preoperative axial T2-weighted MRI. Note that there are two compartments showed different contrast enhancing.

intervals. After 25.5 years, the patient had brought to our hospital with headache, unsteady gait, nausea and vomiting. Excluding abnormal right cerebellar test results, the patient was healthy. MRI revealed isointense tumor in the right cerebellopontine angle that consists of two compartments (Figure 3); the medial compartment did not show contrast enhancement, whereas the lateral compartment that belong to dura showed moderate, and homogenous contrast enhancement.

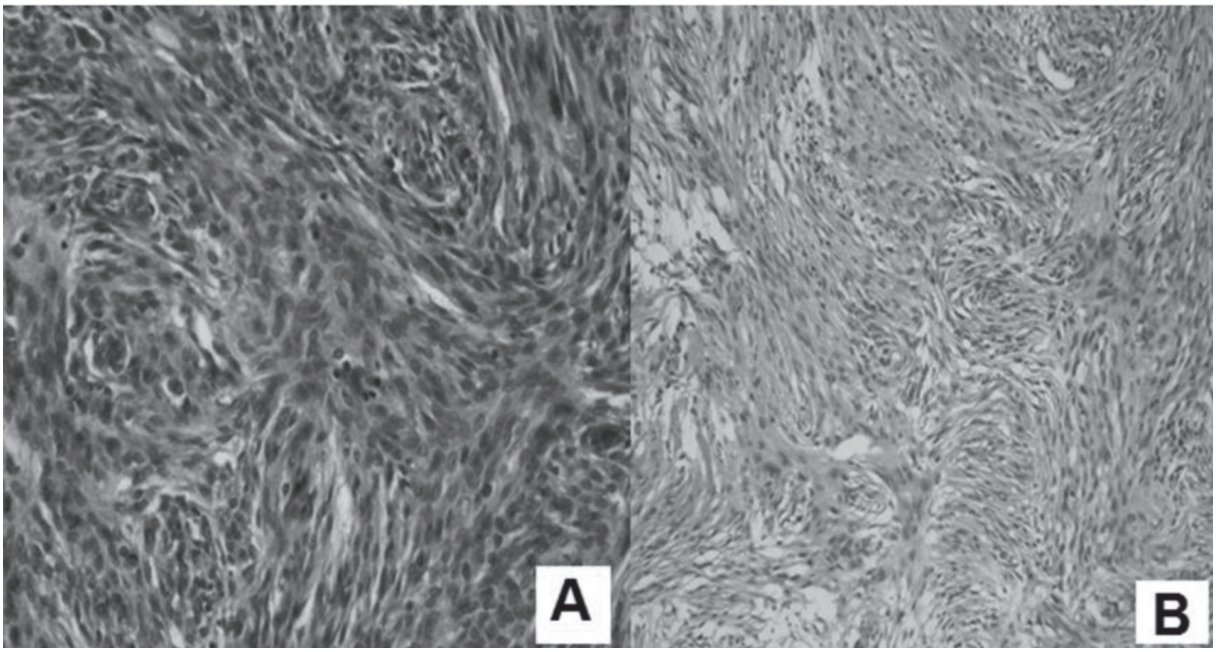

Figure 4. (A) and (B) Histolopathological findings of the right CPA meningioma tissue lead to diagnose the tumor as fibroblastic meningioma (WHO Grade I).

The patient had been managed surgically using right suboccipital craniectomy followed by total resection of the lateral compartment and biopsy material was obtained from the medial compartment that thought to be radiation-induced fibrotic tissue. The excised mass was diagnosed histopathologically as fibroblastic meningioma (WHO Grade I) (Figure 4). There was no necrosis. Only one mitosis was detected per 10 HPF (1 mitosis/10 HPF). Pleomorphism was not observed. The biopsy of the medial compartment demonstrated the presence of arachnoidal fibrosis. The patient was cured, and discharged on the third postoperative day. After thirty months she had not any disease recurrence. Yearly controls were recommended.

DISCUSSION

In 1953 Mann et al. firstly reported a a case with radiation-induced meningioma (RIM). The patient was 6-year- old girl who received 6500 rad after resection of an optic nerve glioma [(1)]. A meningioma was diagnosed 4 years later within the radiotherapy field [(1)]. There is no doubt that radiation injury is a factor in the development of meningiomas and other neoplasms.

Criteria of the radiation-induced tumors are well reported in the literature. To be considered a radiation-induced lesion, the tumor must fulfill the following criteria: 1) it must occur in the field of radiation; 2) tumor should be absent prior to irradiation as ensured in the first MRI performed at the initial diagnosis, prior to surgery and radiotherapy; 3) there must be a delay of several years from the time of treatment to its appearance (after high-dose radiotherapy for medulloblastoma, RIMs have been described after latency periods ranging from 5 to 27 years, with an average latency period of 17.8 years); 4) the induced lesion must differ histologically from the original tumor that was irradiated; and 5) the patient does not have a genetic disease (such as neurofibromatosis type II or others) or a condition predisposing to secondary malignancy [(3)]. In our case, all requirements were met. However, no histological diagnosis was ever made for either lesion.

The review of the literature has shown that

high-dose RIMs occur after an average latency period of 19.5 years (range 3.5-63 years) [4,10]. RIMs typically display female preponderance, although the difference between sexes may be less distinct compared with spontaneous meningiomas (SMs) [2,5,7,11]. The female predominance of 1.29:1 in a survey of approximately 760 RIMs have been reported up to now (according to the best of our knowledge).

Banerjee et al confirmed that RIMs occurred only in their patient series treated with radiation doses of 21 Gy or more [2]. When the time since irradiation is taken into account, both patient groups (18-21 Gy vs. >21 Gy) seemed to be at equal risk for developing meningioma. Neglia et al. reported higher relative risks for all brain tumors at doses exceeding 30 Gy, and the risk for RIM was on the order of 50-100 [8]. The higher doses therefore elicit a more rapid loss of cellular control mechanisms and a failure of the DNA repair system, leading eventually to tumor formation [10]. Our patient received subsequently high-dose RTP as 50 Gy directed at whole CNS, and the posterior fossa as a focal boost dose of 15 Gy.

RIMs differ from SMs as for patient's age at onset and its multiplicity, aggressiveness, and rate of tumor recurrence. Mean age at presentation has been reported as 29.2-37.9 years in patients exposed to high-dose radiation and 45-58 years in those who received low-dose treatment, whereas SM generally arises in the fifth or sixth decade of life [11].

Meningotheliomatous, transitional, and fibroblastic histological subtypes are the most commonly seen RIMs [11]. Six distinctive histological features characteristic of RIMs have been reported as follows: a high degree of cellularity, cellular pleomorphism, numerous bizarre cells, necrotic changes, increased mitotic figures, and nuclei with pseudoinclusions [11]. Histopathological examination of our case revealed frequent mitoses, hypercellularity, and focal necrosis. Tumor in our patient was diagnosed as fibroblastic meningioma (WHO Grade I) (Figure 4).

Reported rates of recurrence vary between 18.7% and 25.6%, as compared with a 3%-11.4% rate of recurrence among SMs. RIM in our patient did not show recurrence till 30 months after the surgical treatment.

Given the aggressive nature and high recurrence rates of RIMs, leaving wide resection margins in the involved dura mater are vital. In cases in which the tumor involves under- or overlying bone, the osseous portion of the tumor should be radically removed, because osseous invasion has been linked to higher rates of tumor recurrence [11]. The authors recommend removal of the bone if there is suspicion of the osseous invasion and replace it with acrylic graft. Surgery may be followed by RTP in patients with RIMs if radical excision cannot be achieved. For our patient; she had been managed surgically using right suboccipital craniectomy followed by total resection of the lateral compartment and biopsy of the medial compartment that thought to be radiation-induced fibrotic tissue had been obtained. Because of the dural invasion, synthetic dura was used.

CONCLUSION

RIM is a late effect of curative RTP. In children with medulloblastoma, RTP is still the main part of curative treatment, therefore, the incidence of the RIM will increase in the future. In such patients yearly MRI controls had to be achieved to scan all RTP received field. To avoid recurrence, aggressive surgical removal is recommended.

Competing Interest

The authors declare that they have no competing interests. All authors certify that they have no affiliations with or involvement in any orga-

nization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.

Patient Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent form is available for review by the Editor-in-Chief of this journal.

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