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Case Report

Posterior fossa metastasis of retinoblastoma accompanying a supratentorial second focus in a child: a case report

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ABSTRACT

The central nervous system, as a metastatic focus, has an extreme predisposition to retinoblastoma and has been indicated as the most common site for this disease. Herein, we report a 4-year-old girl presenting with multiple cranial metastases of retinoblastoma. To our knowledge, this presented case is valuable and significant in terms of being reported as the first cerebellar metastasis of retinoblastoma in the literature and removing this lesion together with supratentorial focus in a single operation with a single skin incision.

Keywords: Metastasis, Posterior fossa, Retinoblastoma

INTRODUCTION

Posterior fossa metastatic tumors are common in adults, but it is a rare condition in childhood.[1] Neuroblastoma, osteosarcoma, Ewing sarcoma and alveolar rhabdomyosarcoma can metastasize to the posterior fossa in pediatric group patients.[2] Retinoblastoma (Rb) constitutes 2-4% of childhood tumors and 83.9% of Rb metastasis is to the central nervous system (CNS).[3]

Retinoblastoma has a tendency for cranial hemorrhagic metastases.[4] Yet, no case has been seen in literature as cerebellar metastasis of Rb and subspecified to infratentorial localization. Eighty percent of all metastasis in the brain parenchyma is in the hemispheres, 15% is in the cerebellum and 5% is in the brainstem.[5] This paper presents a 5-year-old girl with cerebellar and cerebral metastasic masses histopathologically diagnosed as Rb metastasis, who was enucleated at the age of 4-years-old owing to Rb. In terms of surgical and clinical features, this case firstly demonstrates Rb metastasis in the cerebellum and accompanying second lesion in the supratentorial space, both operated on in a single operation. The importance of this case report lies in the fact that it is not only the first posterior fossa metastasis of Rb presented in the literature, but also the first time supratentorial and infratentorial lesions are operated on in a single session, to our knowledge.

CASE REPORT

A 4-year-old Turkish girl was admitted to our hospital with complaints of imbalance and difficulty in walking. On neurological examination, she had a tendency to sleep and a left hemiparesis with 2/5 muscle strength grading scale (oxford scale). On physical examination, it was reported that her right eye was enucleated, her blood pressure was normal, and she had anemia. She had a history of enucleation of her right eye a year ago after complaining about sudden vision loss and exophthalmos determination of an intraorbital tumor reported as Rb grade II.

Also, she underwent cranial radiotherapy and chemotherapy postoperatively and cranial magnetic resonance imaging (MRI) was performed, which found

strongly enhanced lesions in the supratentorial and infratentorial regions, which are considered as metastasis (Figure 1).

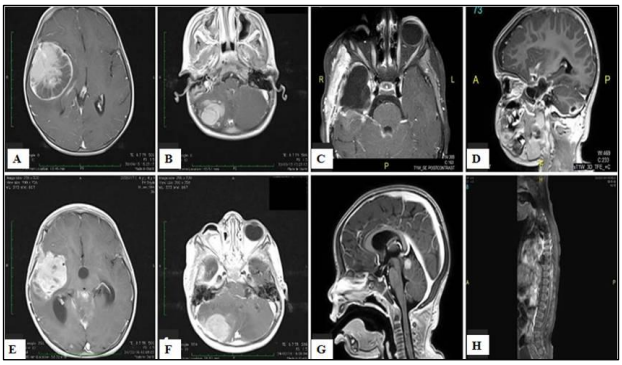

Figure 1a, 1b, 1c, 1d, 1e, 1f, 1g and 1h: (a) contrast enhanced t1-weighted MR images. A mass at the frontotemporal region with strongly enhanced. (b) Axial section at the level of cerebellar hemispheres shows a strongly enhanced mass located at the right cerebellar hemisphere. (c and d) no lesion was detected on the first month postoperative contrast enhanced t1- weighted MR sequences. (e, f, g and h) tumor recurrence at operation field and new strongly enhanced focuses, one is at the superior part of the vermis, one is at the gyrus rectus and the other is in the spinal cord at the thoracal 12th level.

Following the stabilization of the patient hemodynamically and after optimizing the pre-surgical vital conditions, emergency surgery was decided for the purpose of decreasing the intracranial pressure and preventing probable uncal herniation, downward herniation and compression of the brainstem. In the lateral decubitis position, right frontotemporoparietal craniotomy was performed for the supratentorial lesion because of perilesional wide brain edema and large lesion and classic right paramedian suboccipital craniotomy was performed for the infratentorial lesion with a single skin incision in a curvilinear fashion (Figure 2).

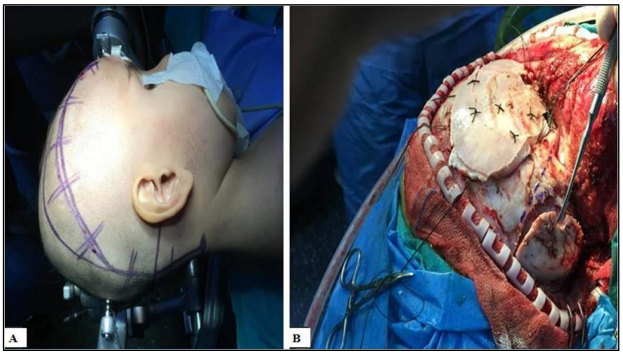

Figure 2a and 2b: (a) Long and single curvilinear skin incision including two cranial bone flaps extending from the midpupillary line behind the hairline anteriorly and ending as a linear paramedian line posteriorly; (b) Two distanced craniotomies.

Subsequent to dural opening under a microscope to protect the cortical and vascular integrity and to avoid parenchymal damage, cortical cohesiveness to the dura and discoloration over the tumor were observed in each tumor location (Figure 3a, d).

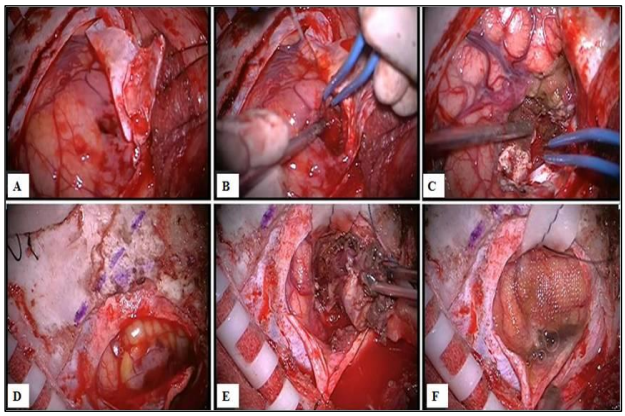

Figure 3a, 3b, 3c, 3d, 3e and 3f: (a) Dural opening, discoloration of cerebral cortex and gyri above the tumor and dural attachment to cortex. (b) During tumor debulking haemorrhage due to the hypervascularization of the tumor is seen. (c) Large operative field after gross total resection of tumor by adding temporal lobectomy. (d) Enlargement and discoloration of cerebellar cortex, yellowish surface due to hemorrhage. (e) Total removal of cerebellar solitary mass with tumor free zone, circumferentially. (f) Postoperative view after resection of cerebellar lesion.

The second surgical step was concentrated on tumor debulking to lower the intralesional and intracranial pressure as fast as possible with safe surgical manipulations. The hemorrhagic tumors in each location were hyper vascularized and infiltrative (Figure 3b, 3e). For each lesion, gross total removal was performed to control the bleeding, prevent possible recurrence and create empty space for the enlarging brain due to postoperative edema (Figure 3c, 3f). The patient's neurological conditions and muscle strength improved postoperatively. She was discharged from the hospital at the 10th day postoperative. The follow up radiological examination a month later showed no recurrence or residual tumor (Figure 1c, 1d).

Histopathologic examination showed highly cellular tumor associated with wide zones of necrosis and hemorrhage. High power microscopy demonstrated that the tumor was comprised of primitive small undifferentiated pleomorphic cells with minimal cytoplasm (Figure 4a). Mitosis and individual cell necrosis was frequent. The enucleated intraorbital tumor was reevaluated and it was noted that both of the tumors were similar, morphologically (Figure 4b). The intracranial tumor mass was diagnosed as a small blue round cell tumor, concordant with Rb metastasis.

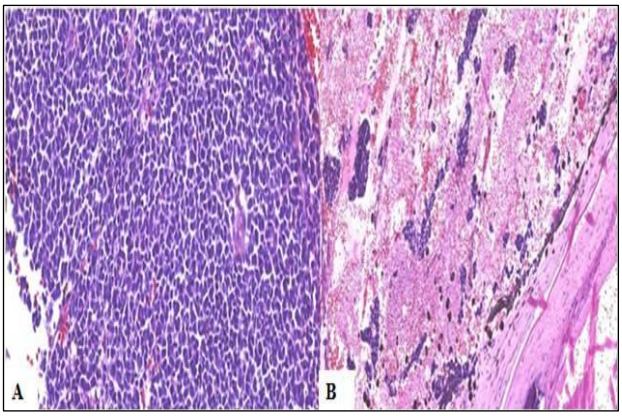

Figure 4a and 4b: (a) Hematoxylin-eosin stained sections of intracranial tumor mass showing diffuse sheath of primitive undifferentiated cells with high nucleocytoplastic ratio and finely dispersed chromatin; H&Ex280. (b) Intraocular tumor showing massive necrosis. Small groups of intact primitive undifferentiated cells adjacent to degenerated and calcified lens capsule, H&Ex190.

DISCUSSION

Retinoblastoma is the most common tumor of the eyeball, ranging from one in 15,000 to 20,000 live births.[6] Tumor occurs by the inactivation of the tumor suppressor Rb 1 gene, which localizes on chromosome 13q14.[7] While the metastatic disease of Rb is usually observed within two years after diagnosis, it is rarely seen after this period.[8] Because of the highly aggressive and rapidly progressive growing feature of Rb, like in our case, CNS metastasis occurred one year after the initial diagnosis of primary orbital tumor. Similarly, our patient was diagnosed a year ago.

The most common primary childhood posterior fossa tumors are reported as astrocytomas, medulloblastomas and ependymomas in the literature.[9] Metastatic tumors in childhood are rare in this region and the most common metastatic retinoblastoma is usually found in the frontal area.[1] Rb is capable of spreading to periorbital tissue via the optic nerve or trabecular structure and anterior chamber, to the central nervous system via CSF (cerebrospinal fluid) and distant metastasis hematogenously.[10] Some authors have published the regions of metastatic lesions of Rb as CNS masses, but the localizations of these lesions in the metastatic regions of Rb did not point out any specific region of CNS.[3,11] Thus, due to insufficient detailing and specifying of these regions in the literature, our case presented here can be encountered as a first case of retinoblastoma having a metastatic lesion in the cerebellum. In addition, the coexistence of two separate metastatic lesions of Rb located in the supra and infratentorial fossas also made this case important.

Bouffet et al, reported a retinoblastoma patient with supratentorial metastasis.[12] Atlas et al, reported two cases of supratentorial hemorrhagic Rb metastasis.[13] De Martino et al reported a case with suprasellar metastasis.[14] Leal-Leal et al presented 38 Rb patients with a mass in the CNS, but they did not clearly express the localization of the lesions.[13] Katayama et al reported a case with ectopic Rb within the third ventricle.[15] Mackay et al reported 20 Rb patients with a mass in the central nervous system, but they did not clearly express the localization of the lesions.[11] Wien et al reported 26 pediatric aged patients with solid-nonhematopoietic metastatic CNS lesion. 15% (4/26) of the metastatic lesions were infratentorial: neuroblastoma, osteosarcoma, Ewing sarcoma and alveolar rhabdomyosarcoma.[2]

We recommend emergency surgery as soon as possible after the diagnosis. Due to the rapid growth pattern of these lesions, especially for the ones in the posterior fossa, brainstem compression and downward herniation may occur in a very short time after patients' admission to the hospital. For this reason, even when the patient is seemingly in good clinical condition and neurological status, before the occurrence or suspicion of a newly beginning neurological deterioration, emergency surgery should be performed.

For the cases we report here, multiple large metastases may be seen in critical regions that may lead to herniation syndrome faster than in other locations. Regardless of the tumor location, the initial surgical aim and strategy should be focused on internal decompression of the tumor and decreasing the intracranial pressure as fast as possible.

One month after the surgery, three new metastatic focuses on the frontal lobe, cerebellum and spinal cord, which were absent on the previous MRI, were detected. Meanwhile, despite the lapse of one month after the operation, in addition to the rapid growth of previous lesions, having observed another two new focuses verifies how biologically aggressive and rapidly growing it is. Based on this clinical outcome, radiotherapy and chemotherapy should be scheduled as soon as possible after the surgery. Additionally, these patients should be followed up at short intervals and control MRI is suggested a month after surgery, at the latest, considering this case.

Although retinoblastoma has the tendency to develop cranial hemorrhagic metastases, and the location of Rb's metastatic lesions are termed in general meaning as the central nervous system, there has been no metastasis of retinoblastoma specified for the posterior fossa in the literature. Present report is remarkable since our case is the first demonstrated multiple posterior fossa metastasis of Rb in which supratentorial and infratentorial metastases were resected in the same session with a single skin incision. Metastasis especially associated with hemorrhage should be kept in mind in patients with a history of Rb who have supratentorial or infratentorial

hemorrhagic lesion. In our opinion, total resection of these metastatic lesions is possible.

CONCLUSION

Total surgical removal CNS metastasis of Rb improves patient outcome and follow up periods should be closer in these patients due to the rapid growth of residual tumor and the aggressive behavior of the primary tumor. This case is clinically important in terms of demonstrating the natural history of Rb and all its possible CNS metastatic targets, describing a surgical approach that may be defined as "maximally invasive" with an excellent patient outcome postoperatively.

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