Case Report

Rare case of trilateral retinoblastoma with spinal canal drop metastasis detected with fluorine-18 fluorodeoxyglucose positronemission tomography/ computed tomography imaging

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ABSTRACT

Trilateral retinoblastoma (TRb) is a rare syndrome associating hereditary bilateral or unilateral retinoblastoma (Rb) with an intracranial neuroblastic tumor. The latter arises in the midline, most often in the pineal gland, less frequently in the suprasellar or parasellar region. The outcome is usually fatal because of secondary spinal dissemination. We report 10-year-old boy presented with a right eye proptosis and leukocoria, and the magnetic resonance imaging (MRI) showed right orbital mass lesion infiltrating optic nerve and diagnosis of retinoblastoma was made. He was referred for fluorodeoxyglucose-positron emission tomography/computed tomography (FDG PET/CT) to find out the extent of the disease. PET/CT showed abnormal FDG-uptake within right orbital mass lesion, suprasellar enhancing lesion and drop metastasis in the cervical spinal canal level. He was diagnosed as a case of TRb with spinal canal drop metastasis. He underwent chemotherapy and craniospinal irradiation.

Keywords: Drop metastasis, fluorodeoxyglucose-positron emission tomography/computed tomography intracranial neuroblastic tumor, trilateral retinoblastoma

INTRODUCTION

Trilateral retinoblastoma (TRb) is a rare disease associating intraocular retinoblastoma with intracranial primitive neuroectodermal tumor.[1] Treatment is difficult, and prognosis is poor. TRb can occur with both familial and sporadic forms of retinoblastoma.[2] To the best of our knowledge, this is the first case report of positron emission tomography/computed tomography (PET/CT) imaging features in TRb with suprasellar mass and drop metastasis.

CASE REPORT

A 10-year-old boy presented with a right eye proptosis and leukocoria. He underwent magnetic resonance imaging (MRI) which showed right orbital mass lesion infiltrating optic nerve, and diagnosis of retinoblastoma was made. He was referred for fluorodeoxyglucose (FDG) PET/CT to find out the extent of the disease. Whole body contrast enhanced PET/CT [Figure 1a] showed abnormal FDG-uptake within right orbital mass lesion [Figure 1b] and suprasellar region [Figure 1c], sagittal fused-PET/CT showing enhancing lesion in the cervical spinal canal level [Figure 1d, arrows]. He was diagnosed as a case of TRb with spinal drop metastasis. He underwent chemotherapy and craniospinal irradiation.

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DISCUSSION

TRb is a disease associating unilateral or bilateral Rb with an intracranial midline primitive neuroectodermal tumor, which usually arises in the pineal gland (77%).[1] The risk of developing TRb in Rb patients is less than 0.5% for sporadic unilateral disease, 5–13% in sporadic bilateral disease, and 5–15% in familial bilateral Rb.[2] Patients with TRb were frequently present with signs of intracranial hypertension.[3] The unusual presence of leptomeningeal dissemination at diagnosis raises the question whether the suprasellar

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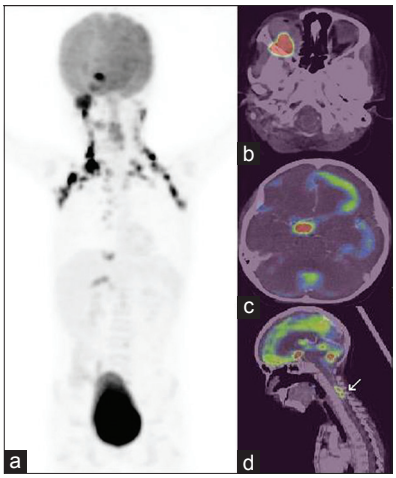

Figure 1: Whole body contrast enhanced fluorodeoxyglucose-positron emission tomography/computed tomography (FDG PET/CT) maximum intensity projection image (a) Axial fused-PET/CT showed intense uptake in the soft tissue lesion in the right orbit (b) suprasellar mass lesion and (c) sagittal fused-PET/CT showing drop metastasis in cervical spinal canal level (d, arrows). Also, physiological brown fat uptake noted in bilateral cervical, supraclavicular, and paravetrbral locations

tumor could be a metastasis. Its strict midline location, the absence of continuity and intraorbital extension on MRI, the absence of diffuse intracerebral nodules, are strong factors against metastasis as well as the close relationship to Rb.[4] Marcus *et al.,*[5] who described in their pathologic review on 80 cases of TRb, 9 suprasellar and parasellar masses with hypointense images on MRI and significant enhancement of gadolinium. In 36 cases, intracranial tumor spread was documented at autopsy and comprised seeding along the spinal cord and canal, diffuse meningeal, ependymal and subependymal involvement, invasion into brain parenchyma, and optic nerve invasion. CT or MRI of the orbit and brain are the standard imaging modalities used for diagnosing and evaluating disease extent in retinoblastoma.[6] To the best of our knowledge, the role of 18F-FDG PET/CT in TRb has not been reported in the published literature. There has been previous report on the use of 18F-FDG PET alone without CT in 4 patients with retinoblastoma[7] and Radhakrishnan *et al.,*[8] described the role of PET/CT in staging and evaluation of treatment response after three cycles of chemotherapy in locally advanced retinoblastoma. This is the first case of identifying TRb with drop metastasis in PET/CT. Recognizing and understanding the clinical findings may determine the overall management of the patients. Treatment of these patients is very difficult and prognosis is poor despite a multimodality approach, as most of them die of leptomeningeal dissemination.[9]

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