Case Report

Primary intracranial malignant melanoma in an adolescent girl: A case report

Sajeeb Mondal, Rajashree Pradhan, Subrata Pal, Supratik Bhattacharya[1], Arindam Banerjee[2], Debosmita Bhattacharyya[3]

Departments of Pathology, [1]Physiology and [2]Pediatrics, College of Medicine and Sagore Dutta Hospital, [3]Department of Pathology, R. G. Kar Medical College and Hospital, Kolkata, West Bengal, India

ABSTRACT

Primary intracranial malignant melanoma is a very rare tumor, and most of the central nervous system melanomas are metastatic diseases. Diagnosis needs extensive dermatological, opthalmological, and radiological workup to exclude metastatic melanoma. Histologically, it should be differentiate from benign melanocytic lesions, pigmented choroid plexus carcinoma, and pigmented papillary medulloblastoma. Here, we are reporting a case of primary malignant melanoma of posterior fossa in an adolescent girl diagnosed in squash cytology as well as in histology and confirmed by immunohistochemistry and by excluding metastatic melanoma.

Key words: Central nervous system, histology, primary malignant melanoma, squash cytology

INTRODUCTION

Primary intracranial melanoma is a rare neoplasm accounting only 1% of all melanoma cases.[1,2] Melanocytic tumors of central nervous system (CNS) may range from benign melanocytoma to malignant melanoma.[2] Most of the CNS melanomas are metastatic diseases.[1] After lung and breast carcinomas, melanoma is the third most common tumor which produces brain secondary.[1] Primary malignant melanoma is indistinguishable from metastatic melanoma in radiological and pathological examination. Primary CNS melanoma is an aggressive disease and also prone to metastasis to other organs.[3] Diagnosis of primary intracranial malignant melanoma needs exclusion of the possibility of secondary by through clinical examination, opthalmological and radiological evaluation.[4] Here, we are presenting a rare case primary malignant melanoma of posterior fossa of the brain in a young girl.

Address for correspondence: Dr. Subrata Pal,
Kalpataru Apartment, Sahid Colony, BT Road, P.S. Khardaha,
North 24 Pargana, West Bengal, India.
E-mail: subratapal1985@gmail.com

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CASE REPORT

An 11-year-old girl presented with headache, vomiting, and ataxia for the past 2 months. Her general physical examinations and higher function were within normal limit. Ophthalmic examinations revealed normal visual acuity and right temporal hemianopia. Fundoscopic examination showed normal retina. Routine hematological examination only revealed mild anemia (hemoglobin −10.2 g%). All biochemical tests and serum electrolytes assay were within normal limit. Computed tomography (CT) brain revealed a large spherical discrete dural based hypodense mass lesion at left occipital region extending to the right side with perilesional edema [Figure 1] CT scan diagnosis of the lesion was an infratentorial meningioma.

She was undergone occipital craniotomy, and intra-operative finding was a grayish black vascular tumor infiltrating from left cerebellum to right side. Intra-operative diagnosis was malignant melanoma. Intra-operative squash cytology revealed hypercellular smears comprised large pleomorphic

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neoplastic cells with intra- and extra-cellular pigment, necrosis and hemorrhage [Figure 2a and b]. The neoplastic cells were round and have moderate eosinophilic cytoplasm, intracellular melanin pigment, high N:C ratio, large round hyperchromatic nuclei, and prominent nucleoli. Mitotic figures were frequent in the neoplastic cells. Squash cytology was diagnosed as malignant melanoma.

In histopathology sections, the tumor was composed of pleomorphic tumor cells arranged in solid and papillary pattern. He cells showed fine brown-black pigment in cytoplasm, large round pleomorphic nuclei, prominent macronucleoli, and frequent mitosis [Figure 3]. Depending on histomorphology, we thought three possible differential diagnoses: (1) malignant melanoma, (2) pigmented choroid plexus carcinoma and (3) pigmented papillary medulloblastoma. The tissue section was stained for synaptophysin, and the tumor was synaptophysin negative. Synaptophysin negativity excluded pigmented choroid plexus carcinoma and pigmented papillary medulloblastoma. An extensive search by dermatological, opthalmological and radiological examinations were performed but failed to show any other primary sites. Final diagnosis was given as primary malignant melanoma of cerebellum.

She was treated with postoperative radiotherapy with 4000 cGy. Follow up was done up to 2 years and there was no recurrence within this time.

DISCUSSION

CNS melanomas are rare malignant neoplasm and most of these arise as metastasis from skin or uveal melanoma.[1,4] Following carcinoma of breast and lung, melanoma is third most common malignancy which metastasize to brain.[1,4,5] Melanocytes are found at skin, mucous membrane, leptomeninges, brain parenchyma, and uvea.[1,6] However, primary intracranial melanoma is a very rare tumor.[1,4] Primary CNS melanomas are derived from melanocytic elements of leptomeninges (melanoblasts of neural crest).[1] Sites of primary CNS melanoma are hemispheres, cerebellum, medulla oblongata, cervical spinal cord, and rarely olfactory bulb.[1,4]

Primary CNS melanocytic lesions are divided into four subtypes: (1) diffuse melanosis, (2) primary isolated intracranial melanoma (3) meningeal melanoblastoma with cutaneous pigmentation and (4) discrete spinal cord melanoma.[1] Diffuse leptomeningeal melanomas mainly affect children and may be associated with neurocutaneous melanosis complex or phakomas.[2,4] Female patients are more commonly involved than male (M:F - 1:2). Clinical presentation of CNS melanoma depends on location and growth rate of the tumor. Melanoma of cerebellum presents with cerebellar symptoms such as ataxia, vertigo,

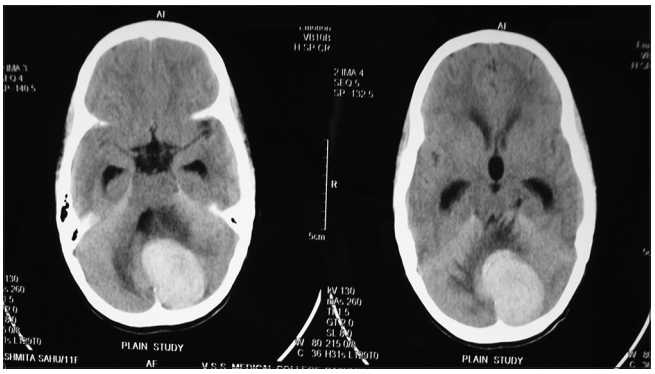

Figure 1: Computed tomography brain revealed a large spherical discrete dural based hypodense mass lesion at left occipital region extending to right side with perilesional edema

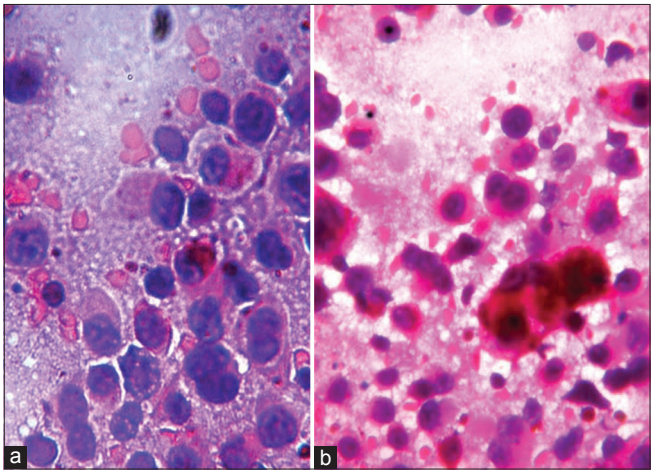

Figure 2: (a and b) Photomicrograph of squash cytology revealed hypercellular smears comprising large pleomorphic neoplastic cells arranged discretely with intra- and extra-cellular pigment ([a] Leishman and Giemsa, ×40 view; [b] PAP, ×40 view)

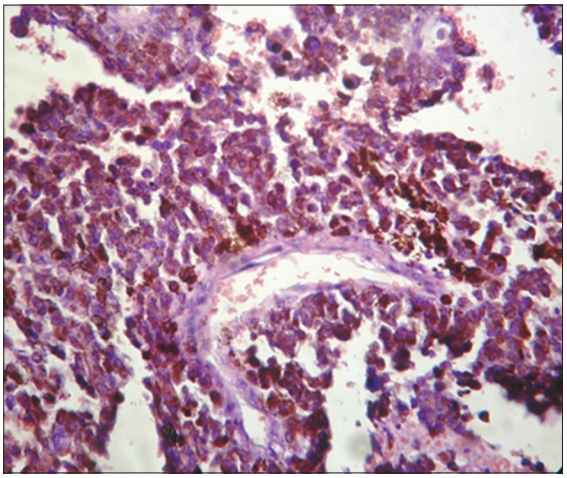

Figure 3: Photomicrograph showing histology of the tumor comprised of pleomorphic tumor cells arranged predominantly in papillary pattern showing fine brown-black pigment in cytoplasm, large round pleomorphic nuclei, prominent macronucleoli and frequent mitosis (H and E, high power view)

increased intracranial pressure, cranial nerve palsy, and meningism.[2] In our case, the presenting features were increased intracranial tension (ICT) and ataxia. In our case, radiological (CT scan) diagnosis was meningioma because it was a dural-based mass. Radiologically, differential diagnoses of malignant melanoma are meningioma, meningeal melanocytoma, metastatic tumor, and sarcoma.[1,7] Magnetic resonance imaging (MRI) brain can differentiate melanotic melanoma from amelanotic melanoma. Melanotic melanomas appear hyperintense in T1 and hypointense in T2-weighted images because of shortening of both T1 and T2 relaxation times. Amelanotic melanomas give reverse images.[1,2] However, MRI cannot differentiate melanoma from intratumoral hemorrhage and other melanotic lesions.[1,2]

Cytology reveals highly cellular smears comprised discohesive sheets of neoplastic cells with abundant intracytoplasmic melanin pigment. The neoplastic cells have dense eosinophilic cytoplasm, large round eccentric nuclei, high N:C ratio, intranuclear inclusion, and prominent macronucleoli. Mitotic figures are frequent, and necrosis is also present.[8] Squash cytology of primary malignant melanoma is indistinguishable from metastatic melanoma.[8] In squash cytology smears, differential diagnoses of CNS melanomas are pigmented choroid plexus carcinoma, pigmented papillary neuroblastoma and melanocytoma.[8,9] On histopathology, the tumor was composed of round to polygonal cells predominantly in solid sheets and papillae with necrosis. The neoplastic cells had cytoplasmic melanin pigmentation, high N:C ratio, pleomorphism, and macronucleoli. Depending on the location of the tumor, squash cytology, and histology, we found three probabilities: (1) malignant melanoma, (2) pigmented choroid plexus carcinoma and (3) pigmented papillary medulloblastoma. Immunohistochemically malignant melanoma of CNS is positive for HMB-45, melan A, and S-100. Other two differential diagnoses are also positive for S-100 and melan A. However, both the pigmented choroid plexus carcinoma and pigmented papillary medulloblastoma are synaptophysin positive, whereas malignant melanomas are synaptophysin negative. In our case, the tumor was negative for synaptophysin, and that excluded the close differential diagnoses.[10]

Before diagnosis of primary CNS melanoma, extensive workup should be done to exclude metastasis from extracranial primary.[2,4] Immunohistochemistry does not help to differentiate primary melanoma from metastatic tumor.[2]

Complete surgical removal with or without radiation is the treatment approach. Radiation is important in cases of incomplete removal or in cases where surgery is not possible.[2,4,8] High-grade external beam radiotherapy to posterior fossa (40–60 cGy) and adjuvant chemotherapy are given for long disease-free survival. Prognosis of primary malignant melanoma of CNS depends on extent of involvement, signs of raised ICT, neurodeficit, extent of tumor removal and use of adjuvant therapies.

CONCLUSION

Diagnosis of primary CNS melanoma needs exclusive workup to exclude metastatic melanoma and clinical, radiological, and squash cytology data cannot exclude possibilities of other pigmented malignancy of CNS.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Liao WC, Ho JT, Lai PH, Tseng HH. Primary intracranial melanoma: Report of case and review of the literature. Mid Taiwan J Med 2002;7:118-23.
2. Shah I, Imran M, Akram R, Rafat S, Zia K, Emaduddin M. Primary intracranial malignant melanoma. J Coll Physicians Surg Pak 2013;23:157-9.
3. Balakrishnan R, Porag R, Asif DS, Satter AM, Taufiq M, Gaddam SS. Primary intracranial melanoma with early leptomeningeal spread: A case report and treatment options available. Case Rep Oncol Med 2015;2015:293802.
4. Onal C, Ozyigit G, Akyorek S, Selek U, Ozyar E, Atahan IL. Primary intracranial solitary melanoma: A rare case with long survival. Turk J Cancer 2006;36:185-7.
5. Gupta G, Robertson AG, MacKie RM. Cerebral metastases of cutaneous melanoma. Br J Cancer 1997;76:256-9.
6. Greco Crasto S, Soffietti R, Bradac GB, Boldorini R. Primitive cerebral melanoma: Case report and review of the literature. Surg Neurol 2001;55:163-8.
7. Rodriguez y Baena R, Gaetani P, Danova M, Bosi F, Zappoli F. Primary solitary intracranial melanoma: Case report and review of the literature. Surg Neurol 1992;38:26-37.
8. Azarpira N, Asadi N, Torabineghad S, Taghipour M. Metastatic malignant melanoma intraoperative imprint cytology of brain tumor. J Cytol 2012;29:192-3.
9. Halder A, Bera S, Datta C, Chatterjee U, Choudhuri MK. Squash cytopathology of primary meningeal melanocytoma. Neurol India 2016;64:152-4.
10. Tripathy K, Misra A, Misra D, Pujari S, Nayak M, Rath J. Melanotic choroid plexus carcinoma of the posterior fossa. J Clin Neurol 2011;7:105-6.