Massive primary cerebellopontine angle melanocytoma in an 11 year old child: A case report and review of literature

Amresh Bhaganagare, Asha Shenoy[1], Naina Goel[1], Atul Goel[2]

Department of Neurosurgery, Bangalore Medical College and Research Institute, Pradhan Mantri Swastha Suraksha Yojana Superspeciality Hospital, Fort Road, Bangalore, Karnataka, [1]Departments of Pathology, [2]Neurosurgery, King Edward Memorial Hospital and Seth G S Medical College, Parel, Mumbai, Maharashtra, India

ABSTRACT

A rare case of meningeal melanocytoma in an 11 year-old girl is reported. The tumor was sub-totally resected. She received no adjuvant treatment and is symptom-free at the follow-up of 8 months.

Key words: Cerebellopontine angle, melanocytoma, skull base tumor

INTRODUCTION

Melanocytomas are rare intracranial lesions. Primary intracranial melanocytic tumors are extremely rare in children. Our literature search revealed reports of seven such cases in children under age of 18 year.[1-7] There is only one case of localized cerebellopontine angle melanocytoma in pediatric age group.[1] We report an additional case of a child of eleven years having a massive cerebellopontine angle melanocytoma and discuss the literature on the subject.

CASE REPORT

An 11-year-old girl presented with chief complaint of headache, vomiting and progressive ataxia for about a month. Additionally, she complained of numbness and weakness of her right side of face. Clinical examination revealed right facial hypoesthesia with depressed corneal reflex, right lower motor facial nerve paresis (Grade 2 House and Brackman) and right sensorineural type of partial hearing loss. Fundoscopy revealed gross papilledema. There were no cutaneous hyperpigmentation or evidence of any other neurocutaneous stigmata.

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Computerized tomography (CT) scan [Figure 1] showed a hyperdense, minimally enhancing lesion in the right cerebellopontine angle, which extended into the middle fossa in the Meckel's cave along the trigeminal nerve root. Magnetic resonance imaging (MRI) showed the mass to be hyperintense on T1-weighted image and relatively hypointense on T2-weighted image [Figure 2a-c]. The lesion was radically excised by right retrosigmoid suboccipital approach. The tumor was jet black in color and was highly vascular. There were dense adhesions of the tumor to the adjoining cranial nerves. All cranial nerves were preserved during operation. The part of the tumor that extended into the Meckel's cave was exposed by drilling the petrous apex. However, despite this, the entire tumor in the middle fossa was not exposed and a part of the tumor could not be resected as it merged inseparably into the trigeminal nerve root. The patient had rapid recovery from her symptoms.

At a follow-up of 8 months, she was entirely asymptomatic. Imaging at this time confirmed sub-total tumor resection [Figure 3a, b]. The patient did not receive any adjuvant therapy. Histological examination [Figure 4] revealed a cellular tumor composed of cells in sheets and in whorls. Cells were polygonal, spindloid and some were epitheloid with moderate cytoplasm with brown black pigment. The cells had vesicular nuclei with prominent nucleoli. Abundant pigment obscured the cellular details. There was no invasion of surrounding brain parenchyma. Mitoses were insignificant. Masson Fontana staining revealed positivity for melanin pigment, which was confirmed after bleaching with potassium permanganate. Clinical and radiological

Address for correspondence: Dr. Amresh S Bhaganagare,
Associate Professor Neurosurgery, BMCRI and PMSSY SSH Victoria Hospital Campus, Fort Road, Bangalore - 560 002, Karnataka, India.
E-mail: amreshsb@yahoo.com

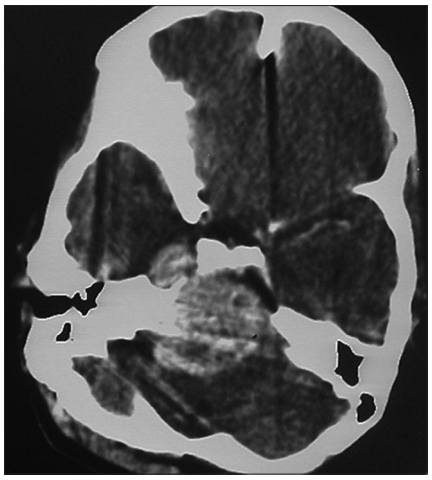

Figure 1: Plain CT scan showing a large predominantly hyperdense cerebellopontine angle lesion extending into the middle cranial fossa

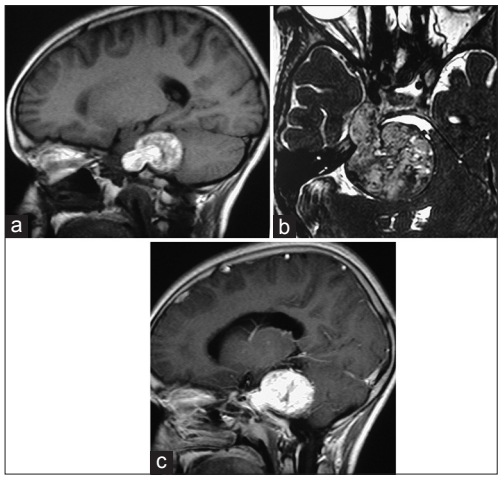

Figure 2: (a) T1-weighted MRI showing the hyperintense tumor and its extensions (b) T2-weighted MRI showing the variegated appearance of the tumor. There are hyperintense and hypointense areas interspersed within the tumor (c) Contrast enhanced T1-weighted MRI showing moderate enhancement of the tumo

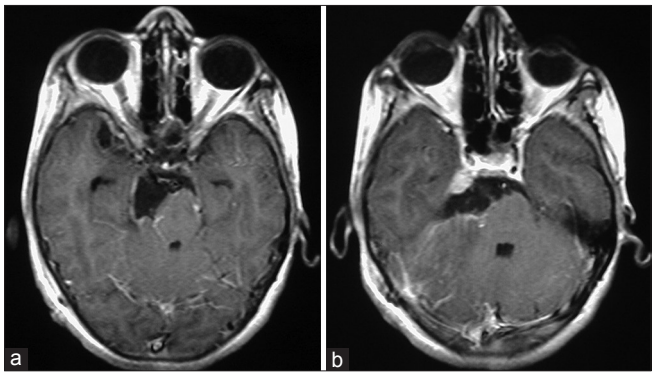

Figure 3: (a) Postoperative MRI showing the tumor resection (b) Postoperative contrast enhanced MRI showing the residual lesion in the middle fossa

search for any other site of melanoma in the body was negative. Investigations included chest radiograph, ocular examination, and ultrasonography of abdomen and barium enema.

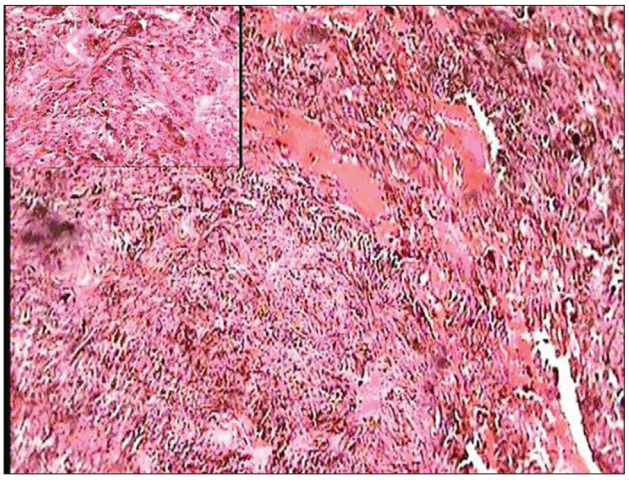

Figure 4: Microphotograph of cerebellopontine angle melanocytoma showing melanocytes in sheets and whirls (H and E, ×100). Inset showing individual melanocytes with dark brown melanin pigment in the cytoplasm (H and E, ×400)

DISCUSSION

Primary intracranial melanocytic lesions are rare, with an estimated incidence is 0.9 per 10 million population.[8] and their occurrence at the cerebellopontine (CPA) is extremely rare.[9] The melanocytic tumors range from benign melanocytomas to malignant melanomas. They originate from melanocytes derived from the neural crest cells during early embryonic development. These cells are commonly distributed in the leptomeninges with a maximum density in the region of the caudal medulla and high cervical cord. The radiological features of melanocytic tumors are characteristic. The lesion is hyperdense on plain CT scan film and enhances on contrast administration. On MRI, the lesion is hyperintense on T1 weighted image and hypointense on T2 weighted image. The lesion enhances on contrast administration. The combination of characters on CT scan and MRI are unique for melanomas. Nevertheless, the MRI signals of these lesions vary widely, depending on the amount of intratumoral hemorrhage. The differential diagnosis of pigmented primary CNS tumors involving the cerebellopontine angle includes melanocytic tumors, pigmented meningiomas, and pigmented schwannomas. Metastatic melanomas are the third most common CNS metastasis, representing 9% of all CNS metastases.[10] Other sites of origin for the melanoma must be eliminated through a thorough physical and radiographic examination. The lesion in our patient had features consistent with a primary melanocytoma, which is the most benign form of melanocytic tumor. Meningeal melanocytomas follow a benign course and radical surgical resection has been associated with excellent long-term outcome.[11]

Histologically melanocytoma cells are uniform and well–differentiated, and they have a low nucleus to cytoplasm ratio. They also exhibit relatively low cellularity, mild nuclear atypia, low mitotic activity, and a low MIB–1 labeling index, and are not associated with necrosis or invasion. In contrast, melanomas are anaplastic and exhibit high cellularity and a high nucleus–to–cytoplasm ratio, marked nuclear atypia, high mitotic activity, a high MIB–1 labeling index, and are usually associated with necrosis or invasion. Histopathologically, intermediate–grade melanocytic tumors are classified between these two extremes.

Treatment options for primary melanocytic tumors vary. However, for melanocytomas, complete tumour resection is considered the best treatment option. The usefulness of radiotherapy, chemotherapy, and immunotherapy has not yet been established. Immunotherapy with agents such as interleukin–2 and IFα (interferon α) has mostly been ineffective because of the difficulty in crossing the blood–brain barrier.[2] Some authors have recommended radiation treatment after an incomplete tumor resection.[11] Despite the incomplete resection of the tumor in our case, adjuvant treatment was not considered as its utility is not conclusive.

CONCLUSION

Primary solitary cerebellopontine angle melanocytoma is rare especially in children. Careful review of preoperative imaging may give important clues to the diagnosis. Given the good postoperative survival of patients with meningeal melanocytoma they should not be misdiagnosed and effort should be made for total tumor removal.

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