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International Meeting on "Pigmentary Disorders from a Global Perspective," Bali, Indonesia
June 22-24, 1997
GIANT CONGENITAL NEVOMELANOCYTIC NEVUS AND INTRACEREBRAL CALCIFIED LESION

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A 7-month-old Caucasian boy was admitted for recurrent generalized seizures since the age of 1 month. Prenatal and postnatal history and developmental milestones were normal. There was nothing in the past medical history that could have accounted for the seizures. The patient was the fourth child of healthy parents, who were first cousins. He had three completely normal brothers. There was no family history of congenital nevus, vascular pathology, or seizures. Physical and neurologic examination was normal. Dermatologic examination revealed a distinct, dark-brown, elevated plaque measuring 17 x 22 cm, situated with small papules and coarse hairs, covering the lower thoracic, lumbar, and sacral regions in a garment distribution (Fig. 1). This giant nevus was associated with multiple scattered, darkly pigmented satellite nevomelanocytic nevi. Histopathologic examination of this lesion was suggestive of congenital nevomelanocytic nevus (CMN). An electroencephalogram (EEG) was normal. Magnetic resonance imaging (MRI) revealed a 2.3 x 2.0 cm, irregularly shaped lesion deep in the right parietal region adjacent to the thalamus, which could be a hamartoma (Fig. 2). Cranial computed tomography (CT) with 55 Hounsfied unit confirmed a calcified hamartomatous lesion in the right hippocampus (Fig. 3). The parents did not consent to brain biopsy. A partial remission was achieved with phenytoin treatment during 3 months of follow-up.

DISCUSSION

Giant congenital nevomelanocytic nevus, also known as garment nevus or giant pigmented nevus, is an uncommon nevocellular lesion. It is commonly associated with satellite lesions and leptomeningeal involvement. Giant CMN of the head and neck or posterior mediastinal area have been reported in association with underlying cranial and/or spinal leptomeningeal melanocytosis in about 60% of cases. Moreover, central nervous system tumors associated with CMN, other than those related to melanocytic proliferations, are much more rare. Only one case of giant CMN has been reported with a benign papilloma of the choroid plexus.

A large CMN is present in approximately 1 in 1000 to 1 in 20,000 newborns. The garment distribution that involves a very large body surface area is much more rare with an incidence of 1 in 300,000. All congenital or acquired nevomelanocytic nevi probably originate from neural crest-derived melanoblasts or from their progeny, and the number and size of nevomelanocytic nevi in a given individual are probably determined by genetic and environmental factors. Familial aggregation has been demonstrated for both the large and small varieties of CMN.

Numerous cases of very large CMN have been reported in association with several conditions such as ocular melanoma, choroidal lesions, neurofibromatosis, neuroblastoma, and leptomeningeal melanocytosis. Melanocytic proliferations affecting the central nervous system of patients with giant CMN may be classified as meningocellular melanocytoma, primary melanoma, or metastatic melanoma.

The association of CMN and malignant melanoma is the most serious condition. The lifetime risk of developing melanoma within a large CMN has been estimated at 18%. Malignant melanomas arise early, with approximately one half being diagnosed in the first 3 years of life.

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Figure 1. Garment distribution of the giant CMN on the dorsal area of the patient covering the lower thoracic and lumbar-sacral regions.
to 5 years of life. In addition to melanoma, a variety of other benign and malignant tumors may develop within a large 2430.

In our patient, imaging studies revealed an intracerebral mass lesion that was not biopsied. A possible explanation of mixed neoplasms containing melanocytic and neuronal elements in CNN is that the neural crest is the origin of melanoblasts, Schwann cells, and sensory ganglia.4-6 As a result, CNN and associated tumors might be related to pathologic events developing from the same embryologic origin, namely the neural crest.

We did not observe any melanomas, either primary or metastatic, on the cranial CT and MRI; the attenuation coefficient of the cerebral lesion situated in the right hippocampus in CT was suggestive of calcification in our case. This appearance may be due to extensive parenchymal melanocytosis mixed with calcification. To our knowledge, no calcified intracerebral lesion with CNN has previously been reported. Meningeal biopsy is required for histologic confirmation is required for detecting leptomeningeal melanocytosis in the absence of mass lesions; however, we could not perform either cyologic examination of the cerebrospinal fluid or a meningeal biopsy. Although tumors have been reported in the absence of meningeal melanocytosis,4-11 we believe that these tumors are due to the lesion in the right deep temporal region.

CONCLUSION

All cases with giant CNN must be followed-up carefully for the development of malignant melanoma and extensive investigations should be performed for leptomeningeal melanocytosis and other central nervous system tumors that originate from the neural crest.

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