Dermatology Practical & Conceptual

Giant Congenital Melanocytic Nevus Accompanied by an Intracranial Arachnoid Cyst

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Introduction

Owing to a somatic *NRAS* mutation at the stage of neural crest formation, patients with congenital melanocytic nevi (CMN) may have coexisting central nervous system (CNS) abnormalities. This risk of CNS abnormalities is higher in CMN with multiple satellite nevi [1]. Frequent CNS abnormalities include melanin deposits, melanocytic proliferations (neurocutaneous melanocytosis [NCM]), Dandy-Walker malformation, and spinal or cranial arachnoid cysts. Moreover, CMN with a diameter >40 cm projected adult size are classified as giant congenital melanocytic nevi (GCMN) and carry an increased melanoma lifetime risk.

Case Presentation

We report a 33-year-old male patient with a GCMN covering his lower trunk and upper legs accompanied by large numbers of disseminated and partially hypertrichotic "satellite" nevi (Figure 1, A-D). There was a history of seizures at the age of 3 years, but no medical reports were available. The patient's current neurological status was unremarkable. Magnetic resonance imaging (MRI) scans of the brain and spinal cord revealed a large arachnoid cyst in the posterior cranial fossa (Figure 1, E-G). The cerebellum appeared compressed but no structural aberrations suggestive of a Dandy-Walker malformation were found (Figure 1, E-G). The brain parenchyma and leptomeninges showed no melanocytic proliferations.

We scheduled the patient for dermatological examinations every 6 months including sequential total body imaging, dermoscopy, and palpation of skin and lymph nodes. We recommended follow-up MRI scans of the brain and spine at 2-year intervals to monitor the arachnoid cyst and to detect any new CNS abnormalities. The patient was instructed to present immediately in case of any new neurological symptoms.

Conclusions

CNS involvement in CMN was termed *neurocutaneous melanocytosis* (NCM) and has long been assigned an unfavorable prognosis. Recent observations showed a dichotomy between "classic" NCM (showing widespread leptomeningeal involvement) and other CNS manifestations, including focal melanin deposits of the brain parenchyma and cysts [1]. Importantly,

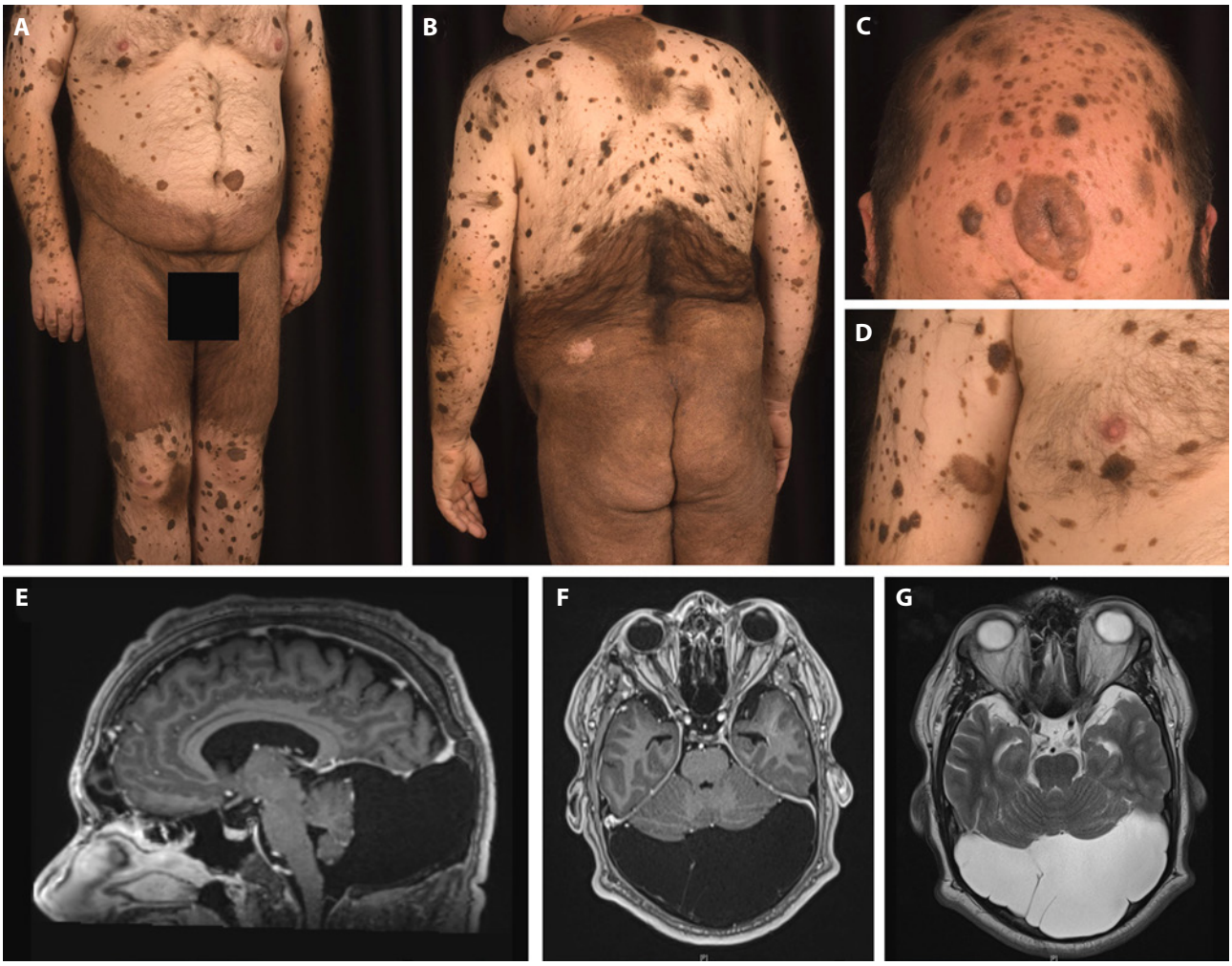

Figure 1. Clinical presentation and MRI scans of the brain. The lower trunk and both upper legs are covered by a GCMN (A). A second large melanocytic lesion is situated on the thoracic posterior midline (B). Many accompanying heterogeneous, partially hypertrichotic and nodular "satellite" nevi are spread over the whole body (C, D). Sagittal T1-weighted magnetization-prepared rapid gradient-echo (MP-RAGE) MRI of the brain revealed a large arachnoid cyst in the posterior cranial fossa (E). Axial T1-weighted MP-RAGE MRI scan of the brain (F) and axial T2-weighted turbo spin-echo (TSE) MRI of the brain (G) showed compression of the cerebellum but no structural aberrations. [Copyright: ©2019 Dusel et al.]

in the case of "classic" NCM the risk of severe complications is considerably higher. Arachnoid cysts are frequent incidental findings in CNS imaging and for the most part do not require any medical intervention. It has been hypothesized that leptomeningeal melanosis may interfere with the formation of cerebrospinal fluid resorption pathways, resulting in the formation of posterior fossa or spinal arachnoid cysts. Table 1 lists previous publications reporting the rare association of GCMN with or without NCM and cerebral or spinal arachnoid cysts. However, in our patient MRI scans revealed no melanocytic

Table 1

Author, Year [Reference]	Observation
Frieden et al, 1994 [2]	Case series of 20 neurologically asymptomatic patients with GCMN; brain abnormalities in 9 patients, 1-month-old girl without NCM but left middle cranial fossa arachnoid cyst
Foster et al, 2001 [3]	Case series of 46 neurologically asymptomatic patients with GCMN; brain abnormalities in 14 patients, 1 patient without signs of NCM by MRI but middle cranial fossa arachnoid cyst
Holmes et al, 2001 [4]	43-year-old woman with GCMN and increasing thoracic pain, MRI without signs of NCM but spinal arachnoid cyst
Acosta et al, 2005 [5]	5-month-old girl, GCMN, NCM, symptomatic hydrocephalus, spinal arachnoid cyst
Ramaswamy et al 2012 [6]	Case series of 14 patients with GCMN and NCM; 3 patients had extensive dorsal spinal arachnoid cysts

deposits of leptomeninges [2-6]. Further studies are needed to establish whether there is a true pathogenic association between GCMN and arachnoid cysts. In a large, prospective cohort published by Kinsler et al [7], the risk of melanoma (CNS or cutaneous) was 12% in the group of children with an abnormal baseline MRI, compared with 2% in those with an unremarkable MRI. When stratified according to the largest diameter of the cutaneous nevus, the melanoma risk was 8% for CMN >60 cm and 1% for CMN of all other sizes.

MRI screening is justified in infants with CMN >40 cm in diameter and/or >20 satellite nevi at birth. The clinical management of CMN patients with symptomatic CNS abnormalities includes neurosurgery, anticonvulsant therapy, and shunt placement in cases of increased intracranial pressure. In general, problematic melanocytic lesions in children include large congenital nevi and Spitz nevi that may be viewed as either potential melanoma precursors or melanoma simulators, respectively [8]. Recommendations for management of the CMN and satellite nevi itself should incorporate the risk of malignant transformation as well as psychological implications of any disfigurement. Because of the limited benefit of any prophylactic excisions for melanoma prevention, dermatological follow-up examinations alone are a valuable option in many cases.

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