Case Report

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A Case Report of Neurocutaneous Melanosis with Associated Dandy-Walker Complex

Mohamad Hosein Lookzadeh[1], Razieh Sadat Tabatabaie[2], Maryam Saeida-Ardekani[1], Hanie Bakhshayesh[3*]

[1] Mother and Newborn Health Research Center, Shahid Sadoughi University of Medical Sciences, Yazd, Iran
[2] Department of Obstetrics and Gynecology, Shahid Sadoughi University of Medical Sciences, Yazd, Iran
[3] Department of Pediatrics, Shahid Sadoughi University of Medical Sciences, Yazd, Iran

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Corresponding author:
Hanie Bakhshayesh

Email:
hania.bakhshayesh@yahoo.com

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ABSTRACT

Background: Neurocutaneous melanosis (NCM) and Dandy-Walker malformation (DWM) are two forms of rare congenital neurodysplasia. NCM is a rare dysmorphogenesis characterized by single or multiple giants pigmented cutaneous nevi and the involvement of benign and/or malignant melanocytic tumors of the leptomeninges. DWM is a rare congenital malformation of the brain. Cystic enlargement of the fourth ventricle is its characteristic that communicates with an enlarged posterior fossa, cerebellar dysgenesis, high tentorial insertion, and hydrocephalus. However, these two conditions are rare, and NCM associated with DWC is even more unusual.

Case Report: Here, we report a male newborn with macrocephaly and multiple pigmented nevi over his whole body with regular borders and normal weight, height, and spine. He was finally diagnosed as NCM in association with DWC.

Conclusion: After diagnosing NCM in association with DWC, appropriate follow-up is recommended; however, there is no particular treatment to prevent the malignant change.

Introduction

Studies have demonstrated significant associations between congenital abnormalities of the skin and the central nervous system.[1] A rare congenital neurocutaneous syndrome, named neurocutaneous melanosis (NCM), is characterized by large or multiple pigmented nevi in combination with leptomeningeal melanosis or Melanoma.[2-4] Cutaneous lesions are usually diagnosed at birth - though more may appear later- but neurological

manifestations emerge as the pressure gradient between the cerebrospinal fluid and venous pressure decreases.[5,6] The condition presents in the form of Dandy-Walker malformation (DWM), including the growth of the posterior fossa, high-set tentorium, underdevelopment (small size and abnormal position) of the middle part of the cerebellum (cerebellar vermis), and cystic dilation of the fourth ventricle that is communicating with the posterior fossa and also hydrocephalus.[3] Although shunt placement and chemotherapy may result in temporary alleviation, the prognosis of the association of NCM and DWM is still inferior, and affected children usually die earlier than age four years.[4,5] Here, we present a rare case of NCM in association with DWC.

Case Report

The case is a male newborn in Shahid Sadoughi Hospital with multiple pigmented skin patches. He was a product of a non-consanguineous marriage with an uneventful delivery. There was no family history of a similar condition. In physical examination, he had a macrocephaly and multiple pigmented nevi over his whole body with an average size of 20-50 mm and regular borders (Figure 1).

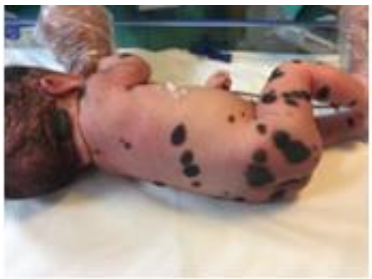

Figure1. Multiple congenital melanocytic nevi

His weight, height, and spine were normal. Routine laboratory investigations at admission to the neonatal intensive care unit showed normal values for his age. Computerized tomography (CT) showed bi-compartmental hydrocephalus with enlargement of the posterior fossa and vermian hypoplasia, suggesting a DWM. There was no evidence of space-occupying lesions and no calcifications or hemorrhages (Figure 2).

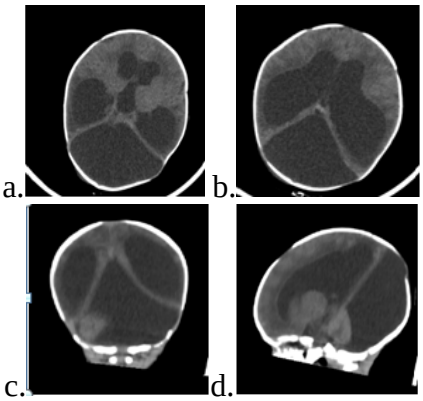

Figure2. Computed tomography demonstrates the hydrocephalus and hypoplasia of cerebellum and enlarged fourth ventricle: a,b: hydrocephalus, c,d: hypoplasia of cerebellum, d: enlarged fourth ventricle

Discussion

NCM is a rare congenital disorder characterized by excessive proliferation of melanin-producing cells in both the skin and leptomeninges. To our knowledge, nearly 300 cases of this non-heritable syndrome are reported in the literature[4] and 10% were associated with DWM.[4,5] The criteria for this lesion were as follows; 1) giant nevus (greater than 20 cm in adults and lesions that are approximately 9 cm in diameter on the head or 6 cm on the body in infants), 2) multiple nevi (greater than or equal to 3 lesions), 3) no evidence of cutaneous Melanoma, except in cases where meningeal lesions are histologically benign, 4) no evidence of meningeal Melanoma except in cases where the cutaneous lesions are benign.[1,7] According to the above criteria, our case was compatible with the diagnosis of neurocutaneous melanosis. Neurocutaneous melanosis may be associated with syndromes such as Sturge-Weber or von Recklinghausen's disease. Associations were also reported with the Dandy-Walker complex, spinal lipoma, and arachnoid cyst.[6,7] The association of DWM with NCM has an inferior prognosis.[8] In all reported cases, the patients showed rapid neurological deterioration and death by four years of age. These two abnormalities show a

phenotypic marker for more profound melanotic infiltration of the leptomeninges, increasing the risk of malignant transformation.[5] An insult to the development of the cerebellar hemisphere and the fourth ventricle may result in the Dandy-Walker complex. The fourth ventricle-cisterna Magna cyst may be formed due to any failure of incorporation between the choroid plexus and the roof of the fourth ventricle or the delayed opening of the foramen Magendie. The meningeal cells influence cerebellar development. In NCM, the development of both the cerebellum and the fourth ventricle may be disrupted because of the melanin-containing abnormal leptomeninges.[8,9]

Conclusion

In this report, we presented a case of NCM with DWM. The association of NCM with DWM is increasingly recognized, and it carries an inferior prognosis.

Conflict of Interest

The authors have no conflict of interest.

Acknowledgments

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The present study was approved by the Ethics Committee of Shahid Sadoughi University of Medical Sciences (IR.SSU.REC.1401.086).

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