Coexisting intracranial meningeal melanocytoma, dermoid tumor, and Dandy–Walker cyst in a patient with neurocutaneous melanosis

Case report

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Abstract

Neurocutaneous melanosis (NCM) associated with Dandy–Walker malformation is a very rare congenital neurodysplasia with the same origin. Primary intracranial melanocytic and dermoid tumors are also benign congenital lesions that usually arise from the leptomeninges and are formed by the inclusion of cutaneous ectoderm at the time of neural tube closure. The authors describe a patient with coexisting intracranial meningeal melanocytoma, NCM with Dandy–Walker malformation, and intraventricular dermoid tumor.

Key Words • Dandy–Walker malformation • cyst • dermoid tumor • melanocytoma • neurocutaneous melanosis

Neurocutaneous melanosis is a rare, nonfamilial syndrome that is characterized by large and/or numerous congenital melanocytic nevi and excessive proliferation of the melanin-containing cells in the leptomeninges. Approximately 8 to 10% of patients with NCM also harbor an associated Dandy–Walker malformation, and this association indicates that these developmental abnormalities share a common origin.

Nevertheless, intracranial dermoid tumor and meningeal melanocytoma are thought to be rare congenital tumors that arise from the inclusion of cutaneous ectoderm at the time of neural tube closure.

The association of primary melanoma with multiple cutaneous nevi, dermoid tumor, and a Dandy–Walker variant has not been previously reported in the medical literature. In this report we discuss the clinical history, neuroimaging findings, and pathological results related to a unique case in which these rare intracranial congenital tumors coexisted with a Dandy–Walker malformation and NCM.

Case Report

History and Examination. This 29-year-old man was admitted to our clinic because of a long-term, severe headache that had been worsening over the last 3 years. After admission, he described the sudden onset of headache and projectile vomiting as symptoms of his increased intracranial pressure. The results of his neurological examination were normal, except for the finding of mental retardation (intelligence quotient 61). On clinical examination, we found multiple disseminated melanocytic nevi on his back and scalp (Fig. 1).

Neuroimaging Studies. The MR imaging studies obtained at admission revealed two large, separate intracranial masses that were combined with a cystic component. The left-sided temporal lesion appeared homogenously hyperintense on the T1-weighted MR image and hypointense on the T2-weighted image. This entity was approximately 7 × 5 × 4.5 cm, and it was a lobulated, solid mass containing multiple cysts (Fig. 2 upper left and center). The right-sided temporal cystic lesion appeared hyperintense on the T1-weighted MR image and hypointense on the T2-weighted image. This mass was approximately 5 × 3 cm and its CT signal was that of lobulated fat containing calcifications (Fig. 2 upper right). The vermis exhibited hypoplasia, with a cystic space that was caused by an open communication with the fourth ventricle (Fig. 2 lower left). The MR images obtained after the addition of Gd contrast material demonstrated enhancement of the tumor-infiltrated meninges (Fig. 2 lower right).

Operation. We performed a unilateral frontotemporal cra-
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Nectomy for removal of the left temporal mass and found melanin pigmentation scattered over the whole brain cortex; the cortex appeared to be dotted with it (Fig. 3). The tumor was so hypervascular that we could not easily remove it. Tumor biopsy sampling and cystotomy were safely performed.

Pathological Findings. The cortical surface showed diffuse proliferation of pigment-laden cells in the leptomeninges and in the Virchow–Robin spaces (Fig. 4 upper). Biopsy sampling of one of the intracranial nodules revealed a melanocytoma arising from the diffuse melanosis. The tumor was composed of densely packed, monomorphic epithelioid or polygonal cells with round vesicular nuclei, prominent nucleoli, and cytoplasm rich in melanin (Fig. 4 lower). Immunostaining for HMB-45 was focally positive, and staining for S100 protein was also positive. The Ki67 labeling index was 5%. Mitoses and necrosis were not found. Biopsy sampling was performed on the skin lesions, and this showed intradermal nevi.

Discussion

Neurocutaneous melanosis is a rare, congenital syndrome in which benign and malignant melanocytic tumors of the leptomeninges are found, along with the development of large and/or numerous congenital melanocytic nevi. The original description of a case of NCM was reported by Rokitansky in 1861. The term NCM was coined by Van Bogaert in 1948 to describe a phakomatosis that was characterized by melanotic pigmentation of various areas of the skin in association with a profuse, dusky melanosis of the leptomeninges. In 1991, Kadonaga and Frieden established the current diagnostic criteria, as follows: 1) large or multiple congenital melanocytic nevi in association with meningeal melanosis or melanoma; 2) no evidence of cutaneous melanoma, except in patients for whom the examined portions of the meningeal lesions are benign according to histological findings; and 3) no evidence of meningeal melanoma, except in patients for whom the examined areas of the cutaneous lesions are benign according to histological findings.

The Dandy–Walker malformation is also a rare developmental abnormality of the central nervous system that is characterized by hypoplasia or aplasia of the cerebellar vermis, cystic dilation of the posterior fossa, and an enlarged fourth ventricle. Hydrocephalus is also usually present and is due to maldevelopment of the rostral embryonic roof of the rhombencephalon. Although the origin of Dandy–Walker cysts is not known, it has been theorized that they are caused by an induction failure of the opposing cerebellar plates, with the persistence of the membrane area of the fourth ventricle at or before the 7th week of embryonic development.

Intracranial lipomas are usually associated with partial or complete agenesis of the corpus callosum. They have a characteristic appearance on unenhanced CT scans, with attenuation values ranging close to −100 Hounsfield units. These tumors can demonstrate punctate rim calcifications, and the lesions appear as a fat signal on all MR imaging studies. In addition, MR imaging studies may also demonstrate the associated callosal agenesis.

Intracranial dermoid tumors originate from ectodermal inclusions of primitive pluripotential cells, and this is due to defects of neural tube closure at approximately 3 to 5 weeks of gestation. They are rare lesions and account for less than 1% of all intracranial tumors. These lesions are lined with stratified squamous epithelium, and they enlarge by the desquamation of normal cells and the secretion of dermal elements into a cystic cavity. Dermoid tumors have a characteristic appearance on CT and MR images. They tend to occur adjacent to the midline, and they appear round or lobulated on CT scans, with attenuation values from −150 to 0 Hounsfield units. Additionally, they usually have a slight mass effect and foci of calcification with no contrast enhancement or surrounding edema. They have a high signal intensity on T2-weighted MR images because of their lipid content, and they have a heterogeneous signal intensity on T1-weighted images due to the mixed composition of the tumor. The right-sided temporal mass in our patient corresponded to the characteristic CT and MR imaging findings in dermoid tumors.

The association of these entities (NCM and Dandy–Walker cyst) is very unusual, and only nine previous cases have been reported in the literature. Our patient with NCM was unique because he had a Dandy–Walker cyst, a dermoid cyst, and melanocytoma all at the same time. Dermoid tumors are thought to result from the development of totipotent ectodermal cells that remain within the
developing neural tube and may be found along the neuraxis. Therefore, the NCM, Dandy–Walker cyst, and dermoid tumor may be the result of errors in the neuroectoderm. There has been no report until now of these three different congenital tumors and anomalies being associated with each other.

On MR images, the signal of an intracranial meningeal melanocytoma is strongly related to the amount of melanin pigment; the more melanin, the more shortening of $T_1$ and $T_2$ relaxation time. The increase in signal intensity on the $T_2$-weighted images in our case might be explained by the malignant transformation and ensuing necrosis. Follow-up evaluations with serial MR imaging studies are being performed in this patient.

**Conclusions**

Dandy–Walker malformation and meningeal melanocytoma accompanied by intraventricular dermoid tumor and...
The tumor cells comprised melanocytoma and dermoid tumor. The neural tube may give rise to nonneoplastic lesions such as melanocytoma and dermoid tumor. Therefore, the totipotent ectodermal cells of primitive meningeal development, but also with ectodermal differentiation interferes not only with the normal inductive effects of melanin. Our case support the possibility that leptomeningeal melanosis associated with Dandy-Walker malformation. NCM have not been previously reported. The findings in our case support the possibility that leptomeningeal melanosis interferes not only with the normal inductive effects of primitive meningeal development, but also with ectodermal development. Therefore, the totipotent ectodermal cells of the neural tube may give rise to nonneoplastic lesions such as melanocytoma and dermoid tumor.

References


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