Nevus of Ota associated with meningeal melanosis and intracranial melanoma

Case report


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A case is reported of a patient in whom an intracranial extracerebral melanoma developed in association with a life-long nevus of Ota. Melanotic pigmentation of the skull, periosteum, and meninges beneath the nevus was found at operation.

Key Words □ melanoma □ nevus of Ota □ skull □ meninges □ scalp □ iris

The nevus of Ota is an irregular discoloration of facial skin and scleral connective tissue. It is frequently unilateral, occurring most commonly in an area that corresponds to the distribution of the trigeminal nerve. In contrast to the giant hairy nevi that are thought to be precancerous lesions, Ota's nevus rarely undergoes malignant change; however, it is occasionally associated with non-cutaneous neoplasms of neural crest origin. These generally arise from the pigmented parts of the iris, choroid, orbit, and uveal tract but rarely from leptomeninges. This case describes a patient with Ota's nevus, diffuse melanosis of the skull, dura, and leptomeninges, and an intracranial extracerebral melanoma.

Case Report

This 23-year-old right-handed Caucasian woman had had a nevus of Ota since birth. Initially, it involved only the skin adjacent to the right eye, but spread gradually over the first 13 years of life to involve the neighboring sclera, conjunctiva, forehead, ear, and scalp. A nodule, which was histologically non-malignant, developed within the nevus in 1978, but there was otherwise no change in the external appearance of the nevus after the age of 13 years (Fig. 1). The distribution was unilateral and trigeminal, with a slight involvement of the C2–3 dermatomes.

Admission. In May, 1980, immediately following the birth of her first child, the patient developed stereotyped attacks of involuntary irrelevant thought and feelings of unreality, each lasting a few seconds and occurring up to 20 times per day. These were diagnosed as partial seizures. Physical examination did not reveal any abnormality apart from the nevus, mild inturning of the terminal phalanges of both little fingers, and multiple benign ectopia of the cardiac ventricles. Computed tomography with contrast enhancement, however, showed a right temporal lobe mass (Fig. 2).

Operation. Craniotomy, with removal of the mass, and biopsy of the nevus were performed in March, 1981. At operation, the periosteum, bone, dura, and, to a lesser extent, leptomeninges underlying the nevus were found to be heavily pigmented with melanin. Beneath the right temporal lobe, a 2- to 3-cm black vascular nodule was seen arising from the dura along the anteromedial aspect of the tentorial notch. The mass was indented but not infiltrating the brain, and was removed completely along a well defined plane. The patient made an uneventful postoperative recovery and has been well, with no seizures, over the 12 months since surgery.

Pathological Examination. Skin biopsy revealed a nevus of Ota resembling a typical blue nevus with varying numbers of melanocytes, occasional melanophages, and some free melanin in the dermis (Fig. 3). These were scattered mainly between collagen bun-
Melanoma and nevus of Ota

FIG. 1. Photograph showing unilateral scleral hyperpigmentation (left) and ipsilateral nevus of the periorbital skin, forehead, and scalp (right).

FIG. 2. Computerized tomography scan with contrast enhancement showing a mass in the right temporal lobe next to the tentorium.

dles but also around skin adnexae. Mitotic figures, hemorrhage, necrosis, junctional change, epidermal melanosis, and deep fatty infiltration were not seen.

The intracerebral mass was a smooth-surfaced slightly lobulated black nodule, 3 × 2 × 2 cm in size. Grossly, it was focally necrotic and hemorrhagic. There appeared to be a 0.1- to 0.2-cm thick capsular wall. One small section of the mass was found next to the meninges and brain, and, although the brain was not infiltrated, tumor was found in the venous sinuses.

FIG. 3. Photomicrograph of the skin biopsy from the scalp nevus showing dermal melanocytes. H & E, × 42.
Most tumor cells were spindle-shaped (Fig. 4), but others were somewhat epithelioid. There were moderate nuclear pleomorphism, rare mitotic figures, pial infiltration, and areas of hemorrhage and necrosis. Together, these changes suggested that this was a malignant neoplasm.

Discussion

The development of intracranial melanoma as a complication of neurocutaneous melanosis is well recognized in association with giant hairy nevi; however, it has also been described rarely with the nevus of Ota. Intracranial melanomas associated with Ota's nevus have been found in the contralateral cerebral hemisphere, the optic chiasm, and in the pineal gland, but only two other cases of an ipsilateral intracranial melanoma comparable to this one have been reported.

The pathogenesis of the melanoma in these cases is not clearly established. Recent embryological experiments elegantly demonstrate the lineage of neural crest derivatives. The findings of such studies clearly support the idea that the leptomeninges, cranial vasculature, dermal connective tissue, corneal stroma, and supportive cells of the peripheral nervous system (including the glia and Schwann cells) arise from common stem cell pools. This would suggest that various neurocutaneous syndromes, such as von Recklinghausen's neurofibromatosis, Sturge-Weber angiomatosis, and neurocutaneous melanosis, may represent a generalized dysplasia of neural crest cell origin in which one or more elements of skin and central nervous system are differentially affected. These findings also partly reconcile rare instances where several neurocutaneous syndromes have been found in association with one another (such as giant nevi with neurofibromatosis, Sturge-Weber disease, and pial telangiectasia).

The direct continuity of pigmentation in the nevus, skull, and dura of this case is not typical of the neurocutaneous melanotic syndromes, and resembles that seen in a case of aggressive cellular blue nevus which had been assumed to spread intracranially by direct extension. The tumor in our patient was, however, histologically similar to primary melanomas of the brain and cord, which may also show signs of cellular malignancy. The histology of the skin biopsy was not that of the cellular blue nevus. Furthermore, the patient has remained well 12 months postoperatively, despite the lack of radical excision of the nevus which was considered necessary in the case of aggressive cellular blue nevus reported by Findler, et al.

We emphasize that a diagnosis of primary intracranial melanoma should be considered in patients with the nevus of Ota who present with features of an intracranial lesion.

Acknowledgments

We should like to thank Dr. R. C. D. Greenhall, Dr. R. Logan, and Dr. M. Walshe for permission to report a patient under their care and for helpful discussion. We also thank...
Melanoma and nevus of Ota

Dr. J. T. Hughes for criticism of the manuscript, Mr. B. Brophy for provision of many of the photographs, and Miss J. Prior for preparation of the manuscript.

References


Manuscript received June 1, 1982.