Blue Nevus and Nevus of Ota Associated With Dural Melanoma

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A 41-year-old woman with a complex congenital nevus that possessed features of both a blue nevus and a nevus of Ota is described. She was found to have melanotic hyperpigmentation of the underlying subcutaneous tissue and dura mater. Two separate melanomas developed in the tissues underlying her nevus (one in the subcutaneous tissue and another intracranially, arising from the dura).


The blue nevus, nevus of Ota or Ito, and mongolian spot are all classified as benign dermal dendritic melanocytoses. They differ in size, depth, anatomic location, and concentration and array of dendritic melanocytes.

In general, the nevus of Ota is not thought to give rise to cutaneous melanoma. However, associated melanomas of the uveal tract, orbit, and brain have been reported. Similarly, a solitary dermal blue nevus only rarely becomes a malignant blue nevus or melanoma.

A case of a congenital plaque-like dermal blue nevus and nevus of Ota with associated ipsilateral subcutaneous and dural melanocytosis is reported. Melanomas developed in the subcutaneous tissue and intracranially, arising from the dura.

Case Report

A 41-year-old, right-handed white female was born with a nevus involving her right frontotemporal scalp and temporal fossa. It gradually enlarged and extended until puberty, when it involved her right cheek and buccal membrane. There was no family history of large congenital nevi or melanoma. The patient presented in August 1987 with a 2-month history of generalized headaches and the development of a firm, nontender subcutaneous nodule in the right temporal fossa.

Physical examination showed a macular, densely pigmented (blue-brown) blue nevus mass over the right frontotemporal scalp and temporal fossa; less dense, bluish-speckled pigmentation with indistinct margins extended over the skin of the cheek and, to a lesser extent, the buccal mucosa (Fig. 1). There was no apparent ocular pigmentation. Neurologic and ophthalmologic exams were normal. A 1.5-cm subcutaneous nodule was palpable below the blue nevus plaque in the right temporal fossa. Otherwise, the general exam was normal.

A head computed tomography (CT) scan with contrast showed a 5 × 6-cm, homogeneous, hyperdense mass in the right middle fossa (Fig. 2). Angiography showed that the mass arose from the floor of the right middle fossa; its vascular supply arose predominantly from the posterior branches of the right meningeal artery.

At surgery, the temporal subcutaneous nodule was removed with the skin incision. The muscle and underlying bone appeared normal. On opening the skull, the dura was found to be pigmented in a distribution corresponding to the overlying nevus (Fig. 3). The mass was approached along the floor of the right middle fossa. The temporal lobe was elevated and a bluish-gray mass was identified approximately 1.5 cm deep to the surface of the brain. A biopsy confirmed melanoma. The tumor had a thick, vascular pedicle that actually arose from the dura overlying Meckel's cave. A gross total removal was achieved. The patient's postoperative course was unremarkable. No additional therapy was given. A follow-up CT scan done 6 months after surgery showed a 1-cm enhancing nodule in the right middle cranial fossa, in the area of the tumor pedicle, consistent with tumor recurrence. The patient was asymptomatic and has been observed expectantly. Her 1-year postoperative scan remains unchanged and she continues to feel well.

Results

Pathologic Examination

Biopsy specimens from skin, the subcutaneous nodule, dura, and the dural mass were obtained. All were formalin-fixed and paraffin-embedded. Sections were cut and stained with hematoxylin and eosin (H & E).

Scalp (Fig. 4): The epidermis was unremarkable. Two types of pigment-bearing cells were found in the dermis: one had small, round nuclei and dendritic cytoplasmic extensions containing fine granular melanin pigment; the other was polygonal with coarse, granular melanin. The
dendritic cells aligned parallel to the collagen fibers in the dermis and subcutaneous fat. Focally within the deep dermis was a collection of small, regular nevus cells containing no pigment with small, round nuclei lacking atypical cytologic features and mitoses. The skin changes were those of a blue nevus with one area of cellular blue nevus.

Subcutaneous nodule (Fig. 5): The specimen contained a portion of subcutaneous fat and connective tissue, within which was a 1.5-cm mass. The connective tissue showed a typical blue nevus with the same two cell types seen in the scalp biopsy specimen. No cellular blue nevus was seen. The tumor showed central necrosis and scattered melanin pigment deposition. The tumor cells were large, round or polyhedral in shape, and many contained melanin. They were arranged loosely in nests. A few cells had dendritic cytoplasmic projections containing pigment. There was moderate nuclear pleomorphism with heterochromatin and prominent nucleoli. Mitotic figures were uncommon (<1 per 10 high-power fields). This tumor was interpreted as being a malignant melanoma.

Dura (Fig. 6): The dura contained bipolar dendritic cells similar to those in the dermis, as well as pigment-containing macrophages.

Dural mass (Fig. 7): The tumor consisted of a vascular neoplasm with melanin pigment scattered throughout. The cells lacked a nesting pattern and formed a syncytial mass. The cells showed nuclear pleomorphism and prominent nucleoli. They were large and polyhedral with clear or melanin-containing cytoplasm. Mitoses were uncommon (<1 per 10 high-power fields) and nuclear atypia was mild. Focal cells showed S-100 positivity. Although showing less cellular atypia than the temporalis mass, this lesion also was interpreted as melanoma.

Discussion

Our patient’s congenital nevus exhibited features of both a common blue nevus and a nevus of Ota. The facial pigmentation was more consistent with a nevus of Ota, a form of benign dermal melanocytosis described by Ota in 1939.11 Many of these patients have associated scleral hyperpigmentation,2 which our patient lacked. However, the single skin biopsy specimen available (taken from the temporal area) was more consistent with a common blue nevus in that the dendritic melanocytes were numerous and were located deeply in the dermis. Classically, a nevus of Ota contains relatively few bipolar melanocytes scattered throughout the upper layers of the dermis.2 It is unlikely that the blue nevus and nevus of Ota represent distinctly separate entities since transitional states between them have been seen clinically and histologically.2,12,13

The cutaneous lesion in our patient represented only the visible portion of a more widespread abnormality, for she also exhibited dendritic melanocytosis in the underlying subcutaneous tissue and dura mater. The involved tissues were located predominantly in the area served by the maxillary branch of the trigeminal nerve. Melanoma was documented in both the subcutaneous tissue and intracranially, arising from the dura. A gross total removal of the tumor was performed.
FIG. 3. At surgery, the dura underlying the nevus was densely pigmented.

FIG. 5. (Top) The subcutaneous nodule consisted of tumor cells loosely arranged in nests. There was moderate nuclear pleomorphism with heterochromatin and prominent nucleoli. Mitoses were uncommon. Fig. 6. (Bottom left) Dendritic melanocytes were interspersed throughout the dura. Fig. 7. (Bottom right) The cells comprising the dural mass exhibited less cellular and nuclear atypia than did those of the subcutaneous mass. Scattered melanin was present. Nucleoli were prominent. Focal cells showed S-100 positivity.
of tumor was effected. Postoperative irradiation was considered for this patient but was not pursued because of the following: (1) the low-grade nature of the neoplasms, (2) the extensive "field defect" that presumably gave rise to the neoplasms, and (3) the questionable sensitivity of melanoma to standard radiotherapy. Follow-up CT scans have been suggestive of local recurrence but have remained stable over a 6-month period, consistent with an indolent process. No treatment is planned until there is evidence of significant tumor progression or symptoms ensue.

Several possible explanations must be considered to account for the findings in this patient. One is that a portion of her large congenital nevus underwent malignant transformation and resulted in simultaneous subcutaneous and dural metastases. However, close examination of the nevus showed no evidence of malignancy. Moreover, this would not explain the presence of a diffuse background of subcutaneous and dural melanocytes apart from the actual masses, nor would metastases be likely to be distributed to separate tissue layers within a restricted anatomic area. There have been a few reports of malignant blue nevi occurring in the scalp that have invaded directly into the underlying structures. However, these lesions uniformly show the background microscopic features of cellular blue nevi with superimposed malignant changes (i.e., frequent mitoses and cellular pleomorphism). Moreover, in these cases the pigmentation and pathologic condition obviously extended contiguously from skin to deeper structures, which was not the case with our patient.

Meningeal melanomas are known to occur in neurocutaneous melanosis, a rare phakomatosis in which pigmented congenital cellular nevi are associated with leptomeningeal melanocytosis. However, in this disorder the pia arachnoid is the chief site of melanocytic infiltration (the dura mater is spared). In our patient, the dura was densely pigmented. Moreover, the gross and histologic appearance of our patient's cutaneous lesion was that of a blue nevus and nevus of Ota, not the typical congenital giant, pigmented, hairy cellular nevi seen in neurocutaneous melanosis.

Unlike other large congenital nevi, the nevus of Ota has only rarely been reported to undergo malignant change. However, scattered case reports in the literature document the association of the nevus of Ota with melanomas arising in other sites, particularly the uveal tract. There have been a few reports of intracranial melanoma complicating a nevus of Ota. These have involved dura, Meckel's cave, the pineal gland, the cortical surface, and the optic chiasm. In 1983, Sagar et al. reported a patient with a nevus of Ota and an ipsilateral dural melanoma; the bone and dura underlying the nevus were hyperpigmented. Our patient seems most closely allied with this group.

A localized anomaly of neural crest development can explain our patient's findings. Complex experimental work by Le Douarin, using chimeric animals, has elucidated a number of descendants of the neural crest. Pigment cells, supporting cells of the peripheral nervous system, autonomic ganglia, several endocrine cells, and the

Fig. 4. Notice the fairly numerous dendritic melanocytes present throughout the dermis with extension into subcutaneous tissue.
skeleton and connective tissue of the head and face (among others) are now known to derive from this fascinating stem cell pool. As generalized neurocristopathies exist (i.e., neurofibromatosis), so do more restricted abnormalities (i.e., the common congenital malformations of the face and mouth). In this patient, we found dendritic melanocytes in the dermis, the subcutaneous tissue, and the dura, all in a localized anatomic area. Melanomas were found at two separate levels.

We propose that some aberration of the complex interactions determining the migration, arrest, and differentiation of the cranial neural crest occurred in this patient. These dendritic melanocytes were abnormal not only in their location; they also displayed a tendency to transform.

REFERENCES