Melanoma with a Coincidental Acoustic Neurinoma

Case Report

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At least 80 cases of primary melanoma of the leptomeninges have been reported,\(^3\)\(^-\)\(^5\) about a quarter of them in infants or children, with extensive involvement of the skin by nevi. However, there do not seem to be any recorded instances of such a tumor accompanied by another intracranial neoplasm, as in the present case.

Case Report

The patient, a 54-year-old man, admitted with a 13-month history of deafness and continual buzzing in the right ear. Before this he had had no significant illness, and there was no family history of neurological disease. Seven months before admission he developed recurrent short episodes (about 1 min) of severe right facial pain, maximal around the eye and over the cheek. Oral medication was ineffective, but alcohol injected into the infraorbital nerve gave some relief. More recently, he had noticed some blurring of vision. He suffered two periods of unconsciousness (at 10 and 3 weeks before admission) each lasting 2 to 3 hours and followed by amnesia, vomiting, and subjective weakness. Between these attacks he had dizzy spells but did not faint.

Examination. There was left anosmia, difficulty in reading fine print, a small area of anesthesia to the right of the nose, and right hearing loss. The eardrums and visual fields were normal, as were the eardrums and bone conduction. Complete physical examination, routine hematological testing, and urinalysis showed no other abnormalities. On the day before operation the patient had a generalized convulsive seizure. Brain scan showed a small area of concentration in the right middle fossa, just above and in front of the pinna (Fig. 1). Pneumoencephalography demonstrated deficient filling of the right cerebellopontine angle.

Operation. Fluid removed at pneumoencephalography contained 490 mg of protein per 100 ml, 63 mg of sugar, and 195 cells, mainly mononuclears. A tumor was removed through a right occipital craniotomy. It was about the size of a golfball and extended both toward the foramen magnum and to the midline, with displacement of the fifth nerve. The tumor was reported as a typical acoustic neurinoma.

Postoperative Course. Two days after operation the patient went into shock and vomited coffee-ground material; although vital signs were stabilized by medical treatment including transfusion, he remained in poor general condition. Twenty-three days after operation the patient had another seizure. Lumbar puncture 6 weeks after operation showed normal pressure, 32 cells, mononuclear, and 184 mg of protein per 100 ml. Two days later the patient rather suddenly became unresponsive, with fixed dilated pupils, and in spite of supportive measures died the following day.

Autopsy. No source was found for the apparent bleeding episode that occurred 2 days postoperatively. Because of the acoustic neurinoma, a careful search was made at autopsy for skin tumors and nevi, but none was found. The brain was swollen, with bilateral uncal and cerebellar herniation, and appeared to show two areas of blood clot, each about 1.5 × 2 cm in extent, situated re-
pectively in the medial margin of the left superior frontal gyrus and in the right middle frontal gyrus. These firm masses were raised above the cortex as slight humps abutted directly on the leptomeninges. No undue pigmentation or other meningeal change was seen. No residual tumor was found in the posterior fossa, but there was some softening of the inferior and lateral parts of the right cerebellum adjacent to the site of the acoustic neurinoma. Sections in the coronal plane showed an apparent large hemorrhage in the right centrum semiovale (Fig. 2). The spinal cord appeared normal.

**Microscopic Examination.** Most of the large mass on the right side was verified as blood clot, but the inferior part was composed of malignant melanoma. The two subpial masses were both melanomas, and a few smaller (1 to 2 mm) foci were found in the superficial white matter on the right. The melanoma tissue was composed of masses of generally discrete cells, rounded or polygonal, varying from 10 to 30 μm in diameter (Fig. 3 left). The cells had one, often two, and rarely more, nuclei, most of which were rounded and often vesicular. Some bizarre forms were present, but mitotic figures were

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**Fig. 2.** Section through the anterior basal ganglia, showing large hemorrhage with melanoma below, and subpial foci in left superior and right middle frontal gyri.

**Fig. 3.** *Left:* Section showing pleomorphism, and mixed pigmented and nonpigmented cells. H&E, ×320. *Right:* Section from the edge of the tumor showing perivascular spread with small cell clumps in parenchyma. H&E, ×80.
few. Although the subpial foci were more deeply pigmented, any high-power field showed cells without pigment, as well as the presence of fine and very coarse granules. The latter may have been in microphages, but the morphology was not distinct. The pigment could be bleached by hydrogen peroxide, and stained as melanin with the Fontana and Ferric ferricyanide techniques. Iron stains showed very few positive-reacting granules. In areas of hemorrhage, the tumor cells were plump and rounded with pale, often slightly kidney-shaped nuclei and prominent nucleoli; pigment was scanty in these cells. At the edge of the tumor masses there was vascular cuffing with melanoma cells, and parenchymal invasion appeared to be occurring from these foci (Fig. 3 right).

Following the diagnosis of malignant melanoma, the original acoustic nerve tumor was reexamined. Numerous sections were stained by the Fontana method. However, the tumor showed no variation from the usual histology of acoustic neurinoma.

Discussion

This patient had a typical acoustic neurinoma with no evidence of pigment formation and, quite separately, several foci of subpial and deep melanin-forming tumor. These foci were invading the cerebral parenchyma and must be regarded as multicentric, intracranial and malignant melanomas of histological type 1 by Gibson’s criteria. Several ophthalmological examinations had shown normal fundi and visual fields. According to Hogan and Zimmerman, most ocular melanomas present evidence of either frank tumor or glaucoma before metastasis, and the metastatic pattern is of general visceral involvement, often most severe in the liver. Direct spread with meningeal involvement is uncommon, and occurs most often with posterior fundal tumors visible on fundoscopy; in this case, there was no involvement in the vicinity of the optic nerves. Nasal melanomas usually spread to the viscera or cervical lymph nodes. Rarely is invasion through the base of the skull. Melanomas are highly unpredictable, and the possibility of an extra-cranial primary tumor in this case cannot be completely excluded, although it seems unlikely.

Reports of primary, pigmented intracranial tumors have included, in addition to melanomas, about six pigmented meningiomas, a pigmented acoustic neurinoma, and an obscure intracerebellar primitive neoplasm. Except for the last one, all of these tumors probably came from neural crest derivatives. Benign neoplasms of melanin-forming meningeal and Schwann cell origin are commonly associated in central and peripheral forms of neurofibromatosis, and malignant change may occur at least in the Schwann cell tumors.

Summary

We have reported a patient who had primary leptomeningeal melanomas and an acoustic neuroma.

Acknowledgment

I am indebted to Mr. D. A. Gibson, Director of the Audio-Visual Facility, for the photography.

References