Primary Malignant Melanoma of the Gasserian Ganglion Associated with Neurofibromatosis


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Case Report

History

A 38-year-old single woman presented to one of us (F.S.H.) on June 17, 1985, with a history of episodic numbness over the left side of the face of 1 year's duration. At the onset these episodes lasted only a few minutes. Later, they became more frequent and progressively coalesced to become continuous. Two months prior to admission the patient experienced burning, intermittent pain in the same distribution, which became unbearable, prompting her to seek medical advice.

Examination

The patient exhibited the stigmata of von Recklinghausen's neurofibromatosis, namely, numerous café au lait patches over the trunk, freckles in the axillae and over the face, and subcutaneous nodules on the trunk, breasts, and extremities. Total body inspection revealed no melanomas or giant hairy nevi. Fundoscopy was negative and no elevated lesions, nevi, or papilledema was observed. Eye movements were intact bilaterally. There was a marked decrease in all sensory modalities over the distribution of the left trigeminal nerve. The left temporalis and masseter muscles were atrophied and weak. Upon opening the mouth, the jaw deviated to the left. The left corneal reflex was absent. The palpebral fissure and the pupil on the left side were smaller than on the right. There was a mild enophthalmos and the left side of the face did not perspire. The other cranial nerves were intact. There were no long tract signs.

Awaiting to complete the workup, the patient was started on 200 mg of carbamazepine (Tegretol®, Geigy Pharmaceuticals, Ardsley, N.Y.) orally three times a day in an attempt to relieve her excruciating, intolerable pain. This produced an excellent response with an almost complete relief of pain. Computed tomography (CT) scan revealed a hyperdense mass in the region of the left trigeminal root and Meckel's cave uniformly enhancing...
Melanoma of the Gasserian Ganglion

Part of the tumor invading the cavernous sinus was left in place. The leptomeninges over the temporal lobe looked grossly normal. The wound was closed in layers.

Pathology
The specimen revealed strands of large pleomorphic cells with heavily pigmented cytoplasm (melanin) and pleomorphic and bizarre nuclei having prominent nucleoli. Interspersed among these sheets of cells were nerve fibers. The diagnosis of malignant melanoma was made. This was confirmed by electron microscopy (Figure 2). The temporal lobe biopsy specimen showed focal increase in well-differentiated astrocytes compatible with a very low-grade astrocytoma. The dura mater was intact and was not invaded by the malignant process.

Postoperative Course and Follow-Up
Immediately following surgery the facial pain disappeared totally. The numbness over the left side of the face became more dense. The rest of the neurological examination remained unchanged. The wound healed primarily and the patient was discharged on the 10th postoperative day. Four months later the patient was doing exceptionally well. When seen on January 23, 1986, she had a left homonymous hemianopsia and a mild left sixth nerve paresis. A CT scan revealed a recurrence, much larger than the original tumor (Figure 3).

On March 13, the patient developed a third nerve paralysis. On April 2, she became blind in the left eye and on July 4, she suffered from severe headaches and swelling of the left eyelids, with closure of the left palpebral fissure. This was associated with a decrease in hearing in the left ear.

On September 26, she was readmitted with left exophthalmos and a left “frozen eye.” She was still conscious and oriented, but had difficulty swallowing and incontinence of urine.

On September 30, a CT scan revealed marked enlargement of the tumor with erosion of the floor of the middle cranial fossa, the lateral wall of the orbit, the anterior portion of the petrous pyramid, and an extension of the tumor into the posterior fossa. It had invaded the base of the skull and presented in the nasopharynx and reached the foramen magnum, eroding the anterior arch of C-1. The zygomatic bone was invaded and eroded (Figure 4). The adjacent brain in the left temporo-parietal region revealed excessive edema. The mass had pushed the undersurface of the brain and had proba-

Figure 1. CT scan done preoperatively in June 1985. Note homogeneous brisk enhancement of the mass across the tentorium along the gasserian ganglion and Meckel's cave.

following intravenous contrast administration (Figure 1). The morphologic and topographic radiological features of the mass were suggestive of a meningioma but, because of the clinical presentation, the preoperative diagnosis was a schwannoma of the left trigeminal nerve.

Operation
On July 16, 1985, the tumor was approached through a left temporal craniotomy. The tip of the left temporal lobe (4 cm) was excised for exposure and to obviate retraction on the dominant temporal lobe. The white matter core was hard and gliotic; a biopsy specimen was taken.

The dura mater of the middle fossa in the region of Meckel's cave was raised by a tumor measuring 2.5 cm in diameter. The dura mater was incised revealing a black mass. The undersurface of the dura mater was neither attached to nor invaded by the tumor. The latter was debulked and nerve rootlets were observed coursing through it. The tumor was followed into the posterior fossa and excised with the attached rootlets of the tri-
Figure 2. (A) Ganglion cells are on the left. Tumor with intracytoplasmic dark granules is seen on the right (hematoxylin and eosin stain × 160). (B) Representative field of the tumor showing intracytoplasmic dark granules (melanin). Nuclear atypia and mitotic figures are easily seen, denoting a malignant tumor (hematoxylin and eosin stain × 160). (C) Electron microscopic picture showing the inclusion bodies to be melanin.
bly invaded the brain parenchyma. There was hydrocepha-
lus and a shift of the midline structures to the right
side. The patient's condition deteriorated rapidly, and
she died on October 16, 1986. No autopsy was per-
mitted.

Discussion

Tumors of the fifth nerve in general are extremely rare
and comprise 0.2% of all primary and secondary intra-
cranial neoplasms [12]. Melanomas of the fifth nerve are
even rarer [10,38]. The only reported cases are associ-
ated with primary or secondary meningeal melanoma-
tosis [2,5,6,28,34,35], or in patients suffering from mel-
anoctyoma [5,6,10,21,23,35,37,39], which seems to have
a predilection for this site. Metastatic melanoma of the
fifth nerve has not been described, although it has been
in the eighth cranial nerve [18]. Other malignant extra-
cranial tumors have been reported to involve secondarily
the intracranial portion of the trigeminal nerve either by
direct extension or through distant metastasis [13,27].

In von Recklinghausen's disease, primary malignant mel-
anomas have been described in the skin as well as in the
uvea [3,9,30], iris [17], and meninges [14,19,25]; none
have been reported to involve the fifth nerve as far as
we know.

Cranial nerves are commonly affected in neurofi-
bromatosis by a variety of tumors: schwannomas, glio-
mas of the optic nerve, and meningiomas of the optic
sheath. The cranial nerve that is affected most commonly
is the vestibuloacoustic nerve. The trigeminal nerve can
be the seat of such a tumor, independently or in associa-
tion with other intracranial nerves.

Our case is one rare example of a tumor of melanocy-
cytic origin of the fifth cranial nerve. The only other case
of possible association of a melanoma of a cranial nerve
and neurofibromatosis is that of Dastur et al [11], in
which a melanotic tumor of the acoustic nerve was re-
ported in a 38-year-old man with café au lait spots but
“no peripheral neurofibromata.”

Though no autopsy was obtained, we considered the
melanoma in our case as primary because:
Figure 3. CT scan done 6 months following operation reveals a local recurrence larger than the original tumor. Note the spread of the tumor anteriorly and laterally.

1. Clinically, there was no evidence of any other melanoma over the skin or in the retina.
2. There were no associated giant hairy nevi [20, 24, 26, 32].
3. Bone scan, abdominal ultrasound, chest x-ray, and hematologic examination failed to reveal any extracranial malignancy.
4. Although the interval between the original complaint and death was 28 months, no other melanoma was diagnosed in another organ.

The CT picture deserves some comment. Meningiomas and schwannomas of the region of Meckel’s cave are not uncommon; their CT appearance may be similar to ours except that, on plain CT scan, melanomas usually appear hyperdense [16] (130 Hounsfield units in our patient). It is important to keep in mind the possibility of such a tumor while discussing the prognosis of patients harboring an enhancing lesion in this area.

The association of cerebral gliosis and von Recklinghausen’s disease has been described involving especially the limbic system and periaqueductal area [7]. In our case gliosis was found in the former site.

An interesting observation was that carbamazepine
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Figure 4 (Continued)

(Tegretol) did relieve the trigeminal pain, although it was organic in origin. This fact should warn against treating trigeminal neuralgias with this drug before full investigation is carried out and the cause of the neuralgia is ascertained.

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References