Melanotic Tumor of the Acoustic Nerve  
Case Report  

Indian Council of Medical Research Neuropathology Unit, and the Department of Neurosurgery,  
Sir J.J. Group of Hospitals, Bombay, India

Since the earlier observations of Masson (1932), it has become increasingly clear that nerve sheath tumors are derived from Schwann cells rather than from any other component of the peripheral or cranial nerves. This has been abundantly substantiated by the tissue-culture studies of Murray and Stout and Lumusden. Although reports have been published describing melanomas of the central nervous system, melanosis of the leptomeninges, and melanotic neurofibromas, or schwannomas and intracranial melanotic schwannoma has not been reported. The present report deals with a case of schwannoma of the left acoustic nerve characterized by gross and microscopic evidence of melanosis.

Case Report

History. A 38-year-old Sikh man came to our neurosurgical clinic complaining of progressive headache of 4 months’ duration. The headache was diffuse but maximal in the frontal region, increasing in severity, and had become continuous for the previous 3 weeks. The patient had also suffered from giddiness and vomiting for 3 months, the vomiting being projectile. Two months before, he had noticed a whistling noise in the left ear, coming on at odd intervals and lasting for 1 or 2 hours. After a few weeks it disappeared, and then he noticed that hearing in that ear had become very poor. There was no right aural complaint. The patient was investigated elsewhere. Lumbar puncture gave the patient relief from his headache, and no unctoward symptoms occurred after the spinal taps. Radiography at that hospital had revealed erosion of the apex of the left petrous ridge.

Direct questioning revealed a history of an old, blunt, closed injury to the left temporal region, unaccompanied by unconsciousness or any sequelae such as otorrhea.

Examination. There was a mild ptosis of the left eye. The left abducens was partially paralysed. Nystagmus with the quick component to the left was noticed. The left facial muscles were weaker than the right. There was total deafness in the left ear. There was no motor or sensory abnormality of the face or body. Romberg’s test was positive; the patient swayed and tended to fall to his left. There were no abnormal pulsations or bruit. Mental function was normal. The corrected vision, pupillary reactions, optic fundi, and corneal reflexes were all normal.

Café-au-lait spots were noted, a large one being situated on the skin over the left anterior abdominal wall. There were no peripheral neurofibromata. There were no pigmented moles, nevi, or other abnormal pigmentation. The chest and abdomen revealed no abnormal findings. The liver was not enlarged. Routine blood and urine tests were noncontributory.

The clinical impression of a left acoustic nerve tumor was strengthened by the radiological findings, which demonstrated dilatation of the left internal auditory meatus (Fig. 1) and erosion of the left petrous ridge. Caloric tests evoked no response from the left acoustic nerve.

Operation. A suboccipital craniectomy was carried out 4 days after admission. A black vascular tumor 4 X 2 cm was encountered in the left cerebellopontine angle. It was encapsulated and was noted to enter a dilated left internal auditory meatus. It surrounded the acoustic nerve, with the facial nerve stretched over its capsule and densely adherent to it. The acoustic nerve had to be sacrificed, and even the separation from the facial nerve was a difficult task. The tumor extended along the 9th, 10th, 11th, and 12th cranial nerves through their various foram...
Ent. The internal auditory artery was adherent to the tumor and had to be sacrificed. The tumor was excised piecemeal, leaving behind only the parts adherent to the various cranial foramina. The meninges were not abnormally pigmented, nor were any seedling deposits of black tumor tissue discovered anywhere in the operative field. The lateral one-third of the left cerebellar lobe had to be sacrificed in order to get at the tumor, but the cerebellar cortex had not been invaded by tumor.

Postoperative Course. The patient recovered well from the operation, but developed a mild fifth nerve paralysis, in addition to the nystagmus and the facial nerve weakness already present. Gross left-sided ataxia was noticed in the immediate postoperative period; this improved partially later. The fifth nerve paralysis improved and, when the patient was discharged, he showed only a mild ataxia, mild left facial paralysis, and nystagmus.

Second Admission. The patient was readmitted 8 months after the first operation with the complaints of pain in the neck and left eye and diplopia of 15 days' duration. Examination now revealed an absent left corneal reflex, with anesthesia of the left half of the face and paralysis of the muscles supplied by the left mandibular nerve. The left sixth and seventh cranial nerves were now totally paralyzed; mild left nystagmus and ataxia persisted. The other findings remained unchanged. Growth and extension of the tumor were obvious. There was no evidence of systemic spread of the tumor to other regions or organs.

Second Operation. At reoperation the dura was tense and bulging. A black, shining, arachnoidal cyst was noticed as soon as the dura was opened. This cyst occupied the space created by partial excision of the cerebellum during the previous operation; 15 ml of straw-colored fluid were aspirated from it. The cut surface of the cerebellum and the neighboring meninges demonstrated a few minute black melanotic deposits. Solid black tumor tissue was seen over the apex of the petrous ridge, eroding it, and extending forward in front of the brain stem and into the middle cranial fossa. It was very friable, and rather densely adherent to the bone, meninges, and the nerves in the jugular fossa and the hypoglossal canal. Once again it was excised piecemeal, with as much as possible being removed.

The patient did not recover from the operation, remained unconscious, and died on the 15th postoperative day. Autopsy was not permitted.

Pathological Examination. The gross and microscopic appearances of the black, friable tumor removed at the second operation were identical in all respects to those of the first specimen, with the exception that the melanosis was a little less intense and permitted histological examination without bleaching of the sections. Histological examination of paraffin sections of the first specimen stained with H. & E. did not reveal clear cellular outlines or details, the tissue mostly showing black masses and clumps. Hence, bleaching of the sections was carried out with various agents such as oxalic acid with hydrogen peroxide and ferric chloride or potassium permanganate (Pearse19) and potassium permanganate with sodium metabisulphide or in addition with sodium thiosulphate. Of these, the first and the last gave the best results and permitted the sort of partial bleaching which seemed to bring out most of the histological details. Total bleaching rendered subsequent staining difficult and inadequate except for the Fontana stain for melanin.

Thin sections (6μ) stained with H. & E. and Picro Mallory stains revealed large and small

---

Fig. 1. Plain skiagram of skull, Towne's view, showing dilatation and funneling of left internal auditory meatus without any destruction of bone. (Report of Dr. J. N. Sidhva, Honr. Neuroradiologist.)
groups of tumor cells around fibrovascular cores (Fig. 2). In some places the cells had a parallel arrangement. The nuclei of these were seen clearly, after complete bleaching, to be oval, vesicular, and very lightly chromatinated, often with a central chromatin dot. The outlines of these cells, which resembled Schwann cells, were delineated by fine brownish-black granules (Fig. 2). There were no mitotic figures or tumor giant cells in any of the several sections examined. There were no whorls or psammoma bodies that would have suggested a meningiomatous tumor.

Among these cells were irregular groups of larger disuniform cells, which were still heavily loaded with pigment and revealed no intracellular details (Figs. 2 and 3). These cells were often found in clusters or in the form of a collar around blood vessels (Fig. 3) which, however, remained free of the pigment. They appeared different in character from the cells first described, being histiocytes, perhaps actively macrophagic Schwann cells.

Fontana’s stain carried out after bleaching the sections (see Fig. 3) revealed nothing additional, merely confirming that all pig-
Melanotic Tumor of the Acoustic Nerve

Acoustic deposits, whether dense or fine, were melanin. Unbleached sections as well as partially bleached sections stained with H. & E. or Fontana's silver nitrate revealed parts of tumor tissue made up of a spongy network of branching cells, their cytoplasmic processes being defined by the melanin granules (Fig. 4). Their nuclei were lost in the granular cytoplasm and were not in the center of the clear spaces, as would have been expected if this were Antoni-type-B schwannoma tissue. The cells under consideration were probably dendritic melanoblasts.

There were no hemorrhages, stainable iron pigment, or cystic degeneration. At one spot there was a small amount of cerebellar cortical tissue compressed by the tumor but in no way infiltrated by it.

Discussion

At both operations this predominantly acoustic tumor was seen to be infiltrating other cranial nerves through their respective foramina. The melanotic meningeal deposits seen over the cerebellum at the second operation indicated a behavior not consistent with the usual type of schwannoma. Moreover, the tumor recurred within 8 months, and the patient died within 13 months of onset of symptoms.

A tentative diagnosis of a "melanotic acoustic schwannoma" was offered after histological examination of the first surgical specimen. There were no dissemination or generalized metastases characteristic of malignant melanoma, nor was there the diffuse or spotty meningeal staining without a large localized mass, suggesting meningeal melanosis. Moreover, the histological pattern observed in the second tumor specimen 9 months after the first continued to be characteristic of schwannoma. The concurrence of other irregular or perivascular cell groups avidly holding on to their pigment (histiocytic Schwann cells) and of spongy areas with branching cells (melanoblasts) created the impression of pleomorphism.

This raises the question of the histogenesis of this tumor. The common ectodermal origin of the Schwann cell as well as of the melanoblast is well known, but the reason for their concurrence in one nerve is not clear. Stout, in describing the rare occurrence of melanin in peripheral nerve tumors, reaffirms the views of Masson that when the Schwann cells come from the neural crest they probably bring with them melanoblasts. This would account not only for the pigmented moles and malignant melanomas but also for the presence of melanoblasts in some neurofibromas. Willis and others have described the association of peripheral neurofibromas and neurocutaneous melanosis.

The only published report, however, which bears some resemblance to ours is Hodson's remarkable case of a melanotic schwannoma of the mandibular nerve occurring with an adamantinoma in the mandible. In the schwannoma, Hodson described two types of cells, the Schwann cell and the dendritic "melanocyte," the melanin preponderating in the latter as in our case. He does not mention, however, the third type of cell we observed here, the large polygonal cell with its heavy melanin content. The possibility that all cells in our tumor are Schwann cells seems unlikely from both the cytological and histogenetic points of view. Melanogenesis would be difficult to explain on the basis of Schwann-cell activity alone. Multipotent though this cell is, there is no evidence of its playing any role in melanin production, although Shillitoe suggests that it may do this, too. In his case of melanotic schwannoma of the gluteal nerve, the few dendritic cells detected contained hemosiderin and not melanin. He also observed heavily melanotic macrophages. Comparable cells have been reported in a meningeal melanoma.

Phagocytosis, one of the many functions ascribed to the ubiquitous Schwann cell, may explain the presence of melanin. Hodson postulated the formation of melanin by residual foci of melanocytes in the schwannoma and its subsequent transfer to the Schwann cells by "cytocrine activity." Shillitoe found it unnecessary to postulate the presence of melanoblasts in all pigmented tumors. In discussing their case of a benign melanotic meningioma, Turnbull and Tom suggested that melanoblasts may supply melanin to, or induce formation of melanin in, adjacent nonmelanoblastic neoplastic tissue. It appears that if there is any particulate matter in the immediate vicinity of Schwann cells, be it intrinsically-derived cell debris or externally-introduced biological or physical material, they will take it up avidly. This
activity is, of course, prominent during nerve degeneration and regeneration. The Schwann cells can ingest India-ink particles introduced underneath the perineurium,\textsuperscript{15} and they invariably phagocytose Mycobacterium leprae in lepromatous neuritis, whether present in large or small numbers.\textsuperscript{1} When introduced into the medium, these bacilli are taken up even by Schwann cells in tissue culture.\textsuperscript{5}

It seems quite plausible that in our case melanin granules produced by the melanoblasts within this predominantly Schwannian tumor were readily ingested by the rapidly proliferating neoplastic Schwann cells which then looked like macrophages. The pigment debris may even provide an added incentive to the growth and multiplication of the Schwann cells as seems to be the case with bacillary material in lepromatous neuritis.

Summary

We have reported the ease of a melanotic acoustic schwannoma which was partially re-removed, recurred locally, and caused death within 9 months of the initial operation. The detailed histological studies suggest the probability of phagocytic Schwann-cell ingestion of melanin granules produced by melanoblasts.

Acknowledgment

Thanks are due to Miss D. K. Manghani, M. Sc., and Mr. V. Talwadkar for their histological technical assistance.

References