Melanotic Meningioma

Case Report

MICHAEL ABBOTT, M.D., FRED A. KILLEFFER, M.D., AND PAUL H. CRANDALL, M.D.
Department of Surgery/Neurosurgery, University of California at Los Angeles,
School of Medicine, Los Angeles, California

Melanin-containing tumors of the central nervous system are encountered only occasionally in neurosurgical operations, Metastases from malignant melanoma make up the majority of these lesions. Primary melanoma of the leptomeninges may produce similar tumors but represents a more unusual condition. To this list may be added the rare melanotic meningioma which is almost a curiosity for only seven cases have been recorded. In this paper we report an additional case and discuss this benign tumor in relation to its highly malignant counterparts.

Case Report

This 57-year-old, white man was admitted to the UCLA Hospital because of progressive weakness and hypesthesia in the legs.

Pain in the spine and left leg had been noted 5 years before admission; with symptomatic treatment it had subsided. However, 18 months before admission the patient noted hypesthesia below both knees. Shortly thereafter, while running, he experienced an attack when he was momentarily unaware of his body below the umbilicus; this loss of sensation caused a fall and right tibial fracture. After 6 months of immobilization, disproportionate weakness and numbness of the right leg were noted. Intermittent rectal incontinence began 6 months before admission, followed by urinary hesitancy 3 months later. One month before admission there was a definite increase in weakness of the right leg and hypesthesia of the left leg. There was no history of any type of skin tumor.

Examination. No spinal tenderness or peripheral or ocular melanotic lesions could be found. Weakness in the distal musculature and reflex hyperactivity of the right leg were confirmed. The left analcutaneous reflex was absent. Hypalgesia below T-6 on the left, mild impairment of the light touch sensation in both legs, and absence of position sense in the right foot were noted. The clinical impression was that of a mass lesion affecting the thoracic spinal cord. Plain x-rays were normal, but Pantopaque myelography revealed an intradural, extramedullary mass on the right at the T-4 spinal cord level (Fig. 1). At this time the spinal fluid was clear, with a protein of 87 mg%.

Operation. At laminectomy a glistening black tumor was found dorsolateral to the cord on the right. It was dumbbell-shaped, extruding through the intervertebral foram into the paravertebral space. The intradural portion was 2 cm in diameter, encapsulated, and attached to the dura. This portion was totally removed. The extradural and extraforaminal portions were somewhat larger.
and removed by curettage. No additional tumors were seen.

The patient's postoperative course was smooth, and his neurological status improved rapidly. He has been followed at regular intervals and is well at 1 year postoperatively.

Histological Examinations. On cut section the tumor was soft, black, and surrounded by a firm, fibrous capsule. The tumor was cellular and well vascularized. Golden brown, granular pigment was scattered throughout, being so prominent in some areas as to obscure the cellular details. Multiple areas of punctate calcification were present. There was a loose architectural pattern to the tumor cells which were arranged in a streaming fibrous pattern with occasional loose whorls (Fig. 2 left). In some areas the cells lacked definite arrangement and appeared as a syncitium. The oval nuclei became elongated when involved in the fibrous configurations (Fig. 2 right). There was moderate nuclear anisomorphism but no indication of dedifferentiation. The nuclei contained a fine chromatin pattern without conspicuous nucleoli; often the nuclei were empty, giving a vacuolar appearance. No mitoses were seen. Special stains confirmed the presence of melanin and absence of iron. The histological diagnosis was melanotic meningioma.

Discussion

The differential diagnosis of melanotic lesions of the nervous system includes secondary malignant melanoma, primary melanoma of the meninges, and pigmented meningioma. Secondary malignant melanomas frequently involve the brain in the metastatic phase of the disease. Several excellent reviews of the neuropathological findings have been presented. The histological changes are those of an obviously malignant neoplasm, usually with a high mitotic index.

One form of secondary melanoma deserves special mention because of its peculiar histology. Malignant melanoma of the eye may assume varied histologic patterns; Callender has presented their appearances very

![Fig. 2. Left: Low-power photomicrograph demonstrates the flowing, fibrous pattern of the tumor. Melanin pigment is seen occurring in a focal fashion with non-pigmented areas interspersed. H&E., ×100. Right: higher power photomicrograph shows elongated, oval nuclei arranged in streaming configurations typical of fibrous meningiomas. Melanin pigment is prominent, occurring in particles of various sizes and shapes. H&E., ×400.](image-url)
Melanotic Meningioma

well. In the spindle category of ocular melanoma one may see an architecture similar to that of the meningiomas. Type B spindle tumors show single prominent nucleoli, while Type A tumors show intranuclear bars running parallel to the long axis of the nucleus.

Although this tumor is malignant, Callender found it outside the globe in only one of 35 cases. The over-all histological picture is similar but not identical to that of meningioma and might give rise to histological confusion in the metastatic phase of ocular melanoma.

Primary melanoma of the nervous system, although less common than the secondary form, is a well-described entity. It is believed to arise from areas of the meninges which normally contain melanin (areas adjacent to the upper spinal cord and ventral surface of the brain). To accept the diagnosis, it is advisable to have evidence eliminating a possible distant primary tumor. The clinical picture has been outlined, and it is apparent that this is a highly malignant condition. Hirano and Carton surveyed 19 surgical cases of primary melanoma of the spinal cord and found the mean duration of preoperative symptoms to be 6 months while the mean postoperative survival was approximately 4 months. Kiel, et al., found the mean total survival to be 5 months, although two cases were mentioned with postoperative periods of 8 and 10 years.

The histological picture of primary melanoma of the central nervous system is clearly that of a malignant tumor. The correct histological diagnosis has apparently been made in all but one case. Schnitker and Ayer described a 49-year-old woman with a 3-year history of back pain in whom an intramedullary melanotic tumor was removed from the T-10 area; she died 6 months later with scattered melanosis of the nervous system and a single metastasis in the liver. This patient had had failing vision in the left eye which was not examined pathologically. Although the biopsy was interpreted as an atypical meningioma, many of the features of spindle cell melanoma were present. In view of the diffuse metastatic nature and unusual histology it is likely that this melanotic tumor was not of meningeal origin but a metastatic ocular tumor.

Melanotic meningiomas are truly rare; only seven cases have been reported. Bailey and Bucy first popularized the phrase “melanoblastic meningioma” in describing a diffuse tumor in a 9-year-old girl, which caused death in 1 year. Review of their description, however, prompts exclusion of this highly malignant, alveolar tumor. Russell and Rubinstein quoted two cases, cervical and cerebellar in position, but gave little elaboration other than their histological benignity.

Ray and Foot described two cases. In the first, a melanotic tumor was removed from the upper lumbar spinal canal; a 25-year follow-up of this case was later recorded by Keil, et al. In the second, a midline posterior fossa tumor was removed after 1 year of preoperative symptoms; death occurred 2 years later, without autopsy. In both cases, the histology was described as that of benign meningioma. Bakody, et al., described a tumor removed from the L-3 spinal canal in a patient who had been symptomatic for 2 years, and who was followed for 6 years postoperatively. They stated that the histological differentiation from malignant meningioma was quite obvious.

Turnbull and Tom described a melanotic tumor of Meckel’s cave with a 2-year preoperative period and a 19-year postoperative follow-up. Keegan and Mullan presented a cerebellopontine angle tumor symptomatic for 6 years, with a 3½-year postoperative survival. In both of the latter cases, the diagnosis was made retrospectively when the long survival period prompted review of the original pathological specimens. At the time of review, the histological appearances were felt to be those of meningioma.

The case described by Schnitker and Ayer casts doubt that pathological examination of biopsied material can be used prognostically. The atypical cytologic picture reminiscent of spindle cell melanoma, and the lack of an ocular examination, make their diagnosis of melanotic meningioma doubtful. In none of the other cases of melanoma of the leptomeninges has such histology been found, and, conversely, all of the reported melanotic meningiomas have had recognizably benign histologies and correspondingly long survivals.

Our case has all the features of a melanotic meningioma. The preoperative period of
18 months and postoperative survival of 12 months without signs of recurrence are not diagnostic but exceed the mean survival time in primary leptomeningeal melanoma or secondary malignant melanoma. The tumor was encapsulated, growing in a typical dumbbell fashion. Finally, the histology was entirely benign and was typical of meningioma except for the addition of melanin pigment.

We believe that the histological characteristics of melanotic meningioma are specific and offer a guide to prognosis. Primary leptomeningeal melanoma and secondary melanoma, including those of ocular origin, also have a characteristic histological picture; however, the outlook for these neoplasms is decidedly different since only brief periods of survival may be expected. It is important to realize that a benign melanotic tumor may occur in the central nervous system. If such a tumor is found at surgery, it should be dealt with as a benign lesion and not merely biopsied as a presumed metastatic deposit.

Summary

We have reported a case of melanotic meningioma occurring in the region of the thoracic spine, have reviewed related reports, and have emphasized the benign nature of this tumor.

References