PIGMENTED MENINGIOMAS: AN UNUSUAL VARIANT

REPORT OF A CASE WITH REVIEW OF THE LITERATURE

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Recently, Kiel et al.2 reviewed the literature of primary melanin-pigmented tumors in the central nervous system and found what they believed to be 85 valid cases and they presented another case, bringing the total to 86. Of the 14 patients with malignant tumors treated surgically, the median postoperative survival was only 1 month. Their case showed the longest postoperative survival which was 19 months. There is still another group in which the survival extends into many years. In these the original diagnosis was also melanoblastoma but because of the long survival the histological material was reviewed and the diagnosis was changed to meningothelialomatous meningioma containing melanin pigment.

This histological picture was mentioned by Russell and Rubinstein4 in their discussion of meningiomas. They described 2 examples in the cervical region and cerebellum respectively, but they did not amplify the case histories. In their cases the constituent cells were described as short and spindle-shaped and, apart from the presence of melanin granules, were not remarkable. Ray and Foot3 in 1940 described 2 cases. Their first patient was a 29-year-old female with a 5-year history of low-back pain with radiation into the legs and associated paresthesias. She was found to have a well encapsulated tumor, black in color, measuring 1 X 3½ cm., lying intradurally at L3. Microscopic sections showed that the tumor was composed of cells resembling those of meningiomas, containing melanin pigment and without mitotic figures. The patient made an excellent recovery and was alive and well 5 years later. Kiel et al.2 cited this case and stated further, by personal communication from Dr. Ray, that this patient was well in 1960, 25 years after the operation.

The second case of Ray and Foot was that of a 45-year-old white female who had headaches of 1 year’s duration, recent vomiting and increasing sluggishness and stupor. The patient was found to have a tumor filling the cisterna magna. It was described as having a tough capsule and weighing 25 grams. Again the cells were described as meningocytes with no mitotic figures. The cytoplasm was dense and filled with melanin granules. In their original report the patient was said to have been alive and well 2 years postoperatively. However, subsequently the patient died but no autopsy was performed, the cause of death being ascribed to recurrence of tumor.

In 1950 Bakody et al.1 described an intradural tumor in a 45-year-old white male. This tumor extended from L2 to L4. The tumor was well encapsulated and some 5 cm. in length. Eight years postoperatively the patient was alive and quite well. Kiel et al. stated that Dr. Hazard reported this patient alive and well 19 years following operation.

In view of the rarity of this tumor, we shall present our case in detail.

CASE REPORT

The patient, a 51-year-old white male, was seen at the University of Chicago Hospitals and Clinics first in July 1958. He stated that he was well until 6 years prior to admission when he noticed the onset of numbness over the left side of the face and the top of the head on arising one morning. The previous night he had driven for two hours with the wind blowing against the left side of his face. The “numbness” was described as a pins-and-needles sensation which, over a number of months, progressed to complete anesthesia, thereafter remaining stationary. Five years before admission the patient noticed a drawing sensation on the left side of the face. Two or three years later he had a tooth extracted and following this he noted double vision and unsteady gait with a tendency to fall to the left. A year prior to admission he noted difficulty with chewing and swallowing and he felt that his mouth was excessively dry. Also he noted that there was some change in his voice, this now being somewhat higher-pitched. For the last 6 months he had been hospitalized in another institution where lumbar puncture, roentgenograms of the skull and angiograms were done. These were reportedly normal. The region of the sensory root of the trigeminal nerve was explored through a left temporal craniectomy and apparently no abnormality was found. Following the operation the pulling-and-drawing sensation in the left side of the face became worse and subsequently the patient experienced intermittent suboccipital pains.

Examination. When first seen at this clinic, it was

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Fig. 1. Photomicrograph of tumor showing tendency to whorl formation. The large dense bodies are calcospheres; the smaller bodies are melanin granules. Hematoxylin and eosin, X350.

Fig. 2. Photomicrograph to show the large accumulation of melanin in the tumor. Fontana-Mason, X350.
found that the left pupil was greater than the right and there was slight ptosis of the left lid. The patient had diplopia in all directions and it was noted that there was distinct limitation of movement of the left eyeball, especially upwards. The corneal sensation was diminished and there was hypalgesia over the left side of the face. Hearing was normal. Voluntary movements of the face were normal. The palate deviated to the right and gag sensation was absent. Strength and reflexes in the limbs were normal. There was some ataxia of the right heel to knee bilaterally. On walking the patient tended to fall to the left. Following admission, a lumbar puncture semifromogram was obtained, by means of air in the subarachnoid space, a soft-tissue density anterior and to the left of the pons. Also it seemed that the 4th ventricle was minimally displaced backwards and to the right.

Operation. On July 18, 1958, a suboccipital craniectomy was performed. Above the left 7th and 8th nerves a smooth-walled tumor was encountered. On aspirating it with a needle, there was no blood. Then on incising it, a tar-colored material was found. The tumor extended widely in front of the pons across to the other side. The total volume was described as being larger than a walnut. This was removed subtotally and the 5th nerve was not visualized at any time.

Histologic Report. A frozen section of the surgical specimen was done and melanin pigment was recognized easily. In the permanent sections streams and whorls of cells were noted, many of which contained melanin particles in the cytoplasm. The nuclei contained delicately stained chromatin and were elongated in shape. There was some proliferation of blood vessels, especially the capillaries. Many calcifications were noted in all areas. No definite mitotic figures were seen. There were many multinucleated giant cells. The initial impression was that this was most likely a melanoblastoma.

Postoperative Course. There was little change in the patient's neurological status except for some increase in the ataxia. He subsequently has been followed at the clinic at 3-month intervals and there has been essentially no change in his neurological status since the time of operation 3½ years ago.

Because of the fact that the tumor had been removed subtotally it was feared that his postoperative survival would be rather short. Roentgen-ray therapy was not given. As time passed without any change in his neurological status, we became dubious of the original histological diagnosis. As a result original slides were reviewed and new sections were made. Fig. 1 demonstrates the tendency to whorl formation and the regularity of the nuclei. The large dense bodies are calcospheres and the smaller bodies which appear to be located within the cytoplasm are melanin granules.

DISCUSSION

In general primary pigmented tumors arising in the central nervous system are unusual forms of melanomas. Rarely one encounters a pigmented meningioma. Initially this is misdiagnosed as a primary melanoma and the correct diagnosis is made after the patient pursues an unexpectedly benign course. An unusually long survival leads to a re-examination of the material and to the correct diagnosis.

SUMMARY

A review of the literature of pigmented meningiomas of the central nervous system is presented and an additional case is reported. The patient is a 51-year-old male who had a tumor of the clivus with extension to the left. The tumor was removed subtotally; the initial diagnosis was that of melanoblastoma. After 3½ years, the patient's neurological status had not changed. Review of the histology determined the inclusion of this tumor in the unusual variety of melanin-pigmented meningiomas.

REFERENCES