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Case Report

MENINGEAL MELANOCYTOMA OF THE MIDDLE CRANIAL FOSSA: A CASE REPORT

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Summary

The term meningeal melanocytoma was first introduced by Limas and Tio in 1972 to figure out lesions of the central nervous system that, under light microscope view, have meningioma characteristics, and ultrastructural characteristics of melanocyte neoplasm, respectively. Meningeal melanocytomas (MM) represent 0.06-0.1% of brain tumors. The annual incidence is about 1 per 10 million. This type of neoplasm is rarely seen in clinical practice. A few cases have been reported in the literature. A 62-year-old man was admitted, complaining of trigeminal pain in the area of the I and II branches of CN V on the left of his face. He underwent two surgeries in a row. First, malignant melanoma, and then meningeal melanocytoma were histologically verified. Neurological examination demonstrated hyperpigmented left iris, neuralgic pain in I and II branches of CN V, latent central hemiparesis and hemihypesthesia for the right extremities, positive Babinski reflex on the right, positive axial pathological reflexes, and partial motor aphasia.
Primary intracranial meningeal melanocytoma is difficult to diagnose preoperatively because of the tumor's non-specific clinical and neuroradiological characteristics. So electron microscopy and immunohistochemical additional diagnostic confirmation are mandatory. Immunohistochemical findings - the presence of S-100 protein, vimentin, and antimelanocyte antigen HMB-45, no reaction for EMA and ultrastructural (melanosomes in different maturation, contacts type zonula adherens) are all in favour of the histopathological diagnosis of MM. Meningeal melanocytoma is a slow-growing tumor, biologically benign, and attempts for surgical radicalism are recommended. It is not entirely clear whether a malignant transformation of these tumors is possible. The role of chemotherapy also remains questionable and has not been documented as effective.
Keywords: intracranial meningeal melanocytoma (IMM), supratentorial, malignant transformation, pigmented tumors of the meninges, middle cranial fossa.

Introduction

The term meningeal melanocytoma was first introduced by Limas and Tio in 1972 to figure out lesions of the central nervous system. These lesions have the characteristics of meningioma under a light microscope view, and the ultrastructural characteristics of melanocyte neoplasm, respectively[1].

Meningeal melanocytoma (MM) appears anywhere along the neuraxis but commonly occurs in the spinal canal near the cranial base (upper cervical spinal cord) and the posterior cranial fossa, Meckel cave, or nuclei adjacent to the cranial nerve. A supratentorial localization is rare. Patients in the fourth and fifth decades are mostly affected, though these tumors have been diagnosed in all age groups. It rarely occurs in children.

Macroscopically, MMs are circumscribed tumors. The degree of pigmentation is variable, from black to dark brown.

Primary intracranial meningeal melanocytoma is difficult to be diagnosed preoperatively because the clinical and neuroradiological characteristics of the tumor are non-specific. On CT, it appears as a well-defined, isodense to hyperdense, homogenous, and contrast-enhancing lesion.

Meningeal melanocytomas represent 0.06-0.1% of brain tumors. The annual incidence is about 1 per 10 million. This type of neoplasm is rarely seen in clinical practice. Less than 200 studies have reported meningeal melanocytoma in the literature (PubMed), and less than 50 studies report evaluation of intracranial meningeal melanocytoma (PubMed) (Fig.1, 2).

Materials and methods

A 62-year-old man was admitted, presenting with trigeminal pain in the area of the I and II branches of CN V on the left of his face. He underwent two surgeries in a row. First, malignant melanoma, and then meningeal melanocytoma were histologically verified. Neurological examination revealed a hyperpigmented left iris, neuralgic pain in I and II branches of CN V. Latent central hemiparesis, hemihypesthesia in the right extremities, a positive Babinski reflex on the right, positive axial pathological reflexes, and partial motor aphasia were also detected.

Surgical treatment: Left frontotemporal craniotomy was performed with transulcal access through the Sylvian fissure. A well-encapsulated multilobulated brown-to-black formation, fused to the middle cranial fossa's dura, was resected under microscopic magnification (Fig.3).

The first pathohistological diagnosis was spindle-cell malignant melanoma. The patient

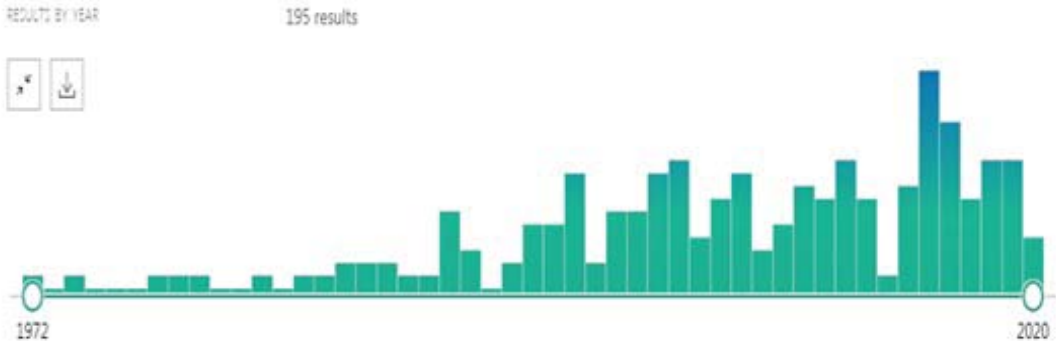

Figure 1. Meningeal melanocytoma (MM) studies reported in the literature (pubmed)

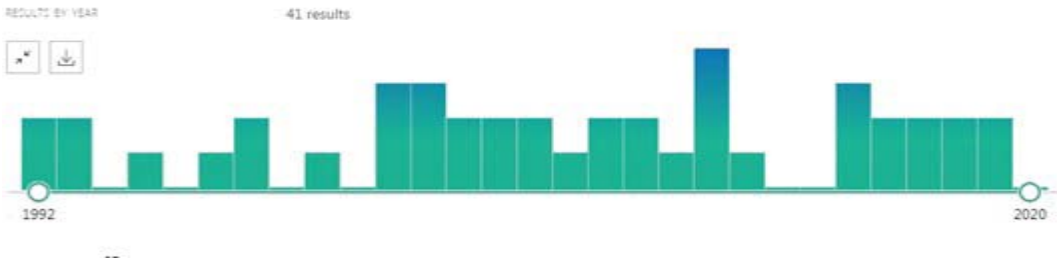

Figure 2. Intracranial meningeal melanocytoma (IMM) studies reported in the literature (pubmed)

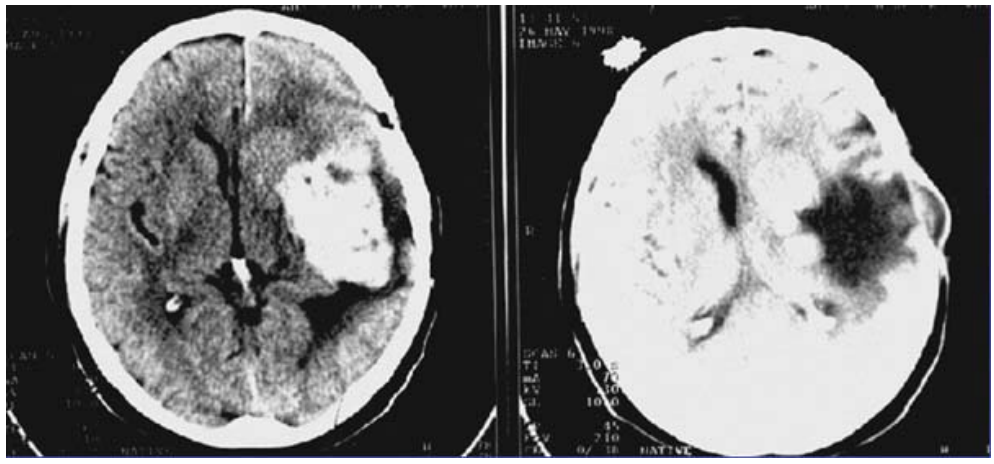

Figure 3. Pre and postoperative CT

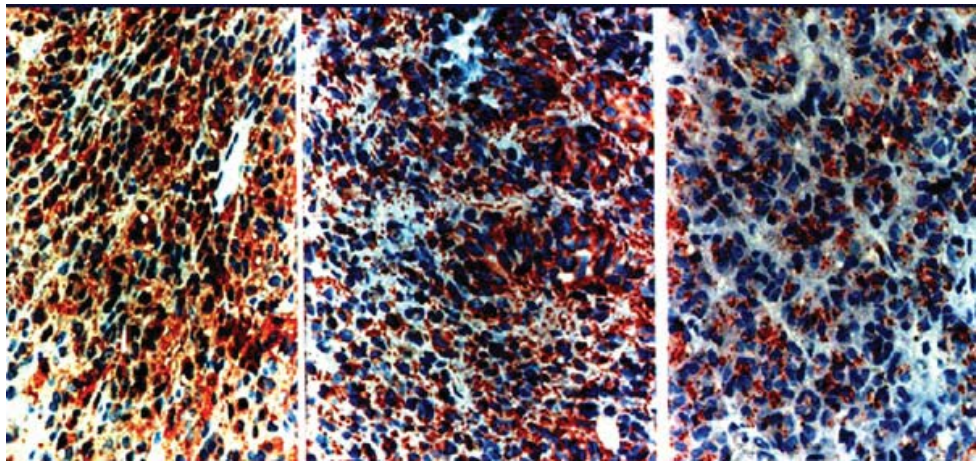

Figure 4. Positive immunohistochemical staining for S-100 protein, vimentin and melanin (nmv-45). Tumor cells were not positive for EMA (epithelial membrane antigen)

was referred for radio- and chemotherapy.

Six months later, the patient underwent a second operation. Histological analysis revealed a highly cellular tumor formation composed of epithelioid and spindle cells with numerous dark-brown to black cytoplasmic inclusions.

Fontana-Mason staining for melanin was positive, and hemosiderin staining was negative. Gomori staining found reticulin fibers in the perivascular spaces.

The immunohistochemical study demonstrated the presence of vimentin, S-100 protein, and HMB-45 antigen. Tumor cells were negative for epithelial membrane antigen (EMA). (Fig.4).

Electron microscopic studies showed the presence of pre-melanosomes at different stages of maturation and the presence of numerous melanosomes (Fig.5).

The differential diagnostic criteria that

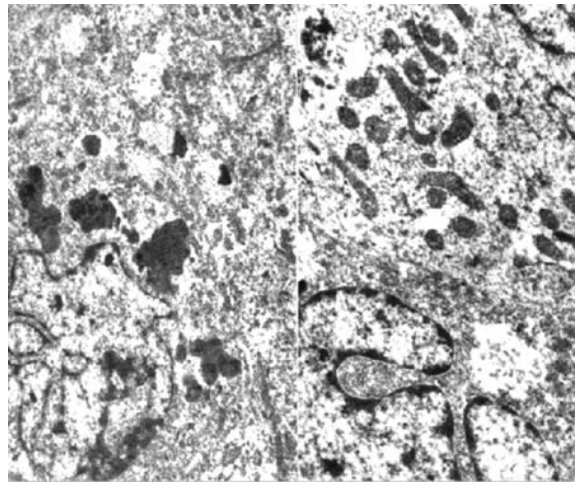

Figure 5. Electron microscopic study

distinguish this rare, benign tumor from other melanotic tumors in the CNS:

1. Immunohistochemical (presence of S-100

	Meningeal Melanocytoma	Melanocytic Schwannoma	Melanocytic Meningioma	Melanoma
HMB-45	+	+-	-	++
S-100 PROTEIN	+	++	-	+
VIMENTIN	+	+	+	-
EMA	-	-	+-	-
GFAP	-	+-	-	-
Leu7	-	+	-	-

Figure 6. Immunohistochemical features of pigmented tumors of the meninges

protein, vimentin, and antimelanocyte antigen HMB-45.) No reaction for EMA.

2. Ultrastructural (melanosomes in different maturation, contacts type zonula adherens.) [2,3]

Discussion

Primary meningeal melanin-containing neoplasms are quite uncommon and rare. They include malignant melanoma, meningeal melanocytoma, pigmented meningioma, melanotic schwannoma, and melanoblasts and have different biological behaviours, characteristics, treatment, and prognosis [1,4].

Similar appearance in CT and MRI investigations is often confusing in making a diagnosis, so additional diagnostic confirmation with electron microscopy and immunohistochemical analyses are needed [5,6].

Meningeal melanocytomas are benign tumors, and possible aggressiveness has not been proved. Radical surgical resection is mandatory. Radiotherapy will prevent tumor regrowth when radical resection is not possible. There are no definite guidelines for the management of MM. The prognosis in MMs is better than the malignant variants of melanin-containing neoplasms [7].

Radiotherapy (RT) is significant for this type of tumour. Rades et al. have shown that radical resection is superior to subtotal resection, with a statistical significance at 1st, 2nd, 3rd, and 4th-year follow-up intervals ($p<0.05$). Subtotal resection with radiotherapy (RT) is superior to the subtotal resection alone at 2-years follow-up ($p<0.05$) [8]. Gamma-knife radiosurgery reduces tumour volume and improves clinical outcomes. Nowadays, the patients' best local control and survival period are provided by gross total resection (GTR) with adjuvant radiotherapy [9].

RT and local control are essential endpoints because recurrent MMs may transform into malignant melanoma. The 5-year local control rate is quite better (86%) for patients treated with a dose of 45–55 Gy versus (27%) with a dose of 30 – 40 Gy [10,11].

In our opinion, radiotherapy is necessary for incompletely resected tumors. There is an ongoing debate about whether RT should be performed to gross totally resected tumors. There is no evidence of benefits from chemotherapy [12].

As a cell proliferation marker, the Ki-67 antigen is an excellent marker for determining a given cell population's growth fraction. Roser et al. have reported an incomplete resection of MM, in which only 3% of cells were stained with Ki-67. RT was not administered. Malignant transformation and tumor recidivism occurred after 12 years. At the time of recurrence, 5% of cells stained positive for Ki-67 [13]. So, MM's slow growth makes low Ki-67 staining significant for recurrence and malignization [14].

Conclusion

Meningeal melanocytoma is a slow-growing, biologically benign tumor, and attempts for surgical radicalism are recommended. It is not entirely clear what percentage malignant transformation of these tumors is possible. The role of chemotherapy also remains questionable and not documented as effective.

Acknowledgements

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