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CASE REPORT

Spinal meningeal melanocytoma

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ABSTRACT

Primary melanotic meningeal neoplasms are extremely rare lesions and benign forms are even rarer though with better prognosis than the malignant ones. We describe a 40-year-old male with a history of gradually progressive weakness of both lower limbs with normal bowel, bladder control, and an intradural mass measuring 1.5×1.0 cm on radiologic investigations. The lesion was surgically excised. Histopathologic examination revealed heavily melanin-pigmented cells, nuclei with reticulogranular chromatin and small nucleoli, moderate amount of eosinophillic cytoplasm with indistinct cell boundaries, and symplasmic appearance. A probable diagnosis of meningeal melanocytoma was made. The diagnosis was confirmed on immunohistochemical analysis which revealed strongly positive expression of HMB-45 in the tumor cells. Vimentin and S-100 were also diffusely positive while neuron specific enolase showed focal and patchy positivity; however, epithelial membrane antigen was distinctly negative.

Key words: Melanocytoma, melanotic neoplasm, spinal meningeal neoplasm

Introduction

Melanotic meningeal neoplasms can be either metastatic or primary, and can also be divided as benign or malignant. Metastatic lesions are by far the most common ones where malignant melanoma comprises 12–16% of all tumors metastatic to the central nervous system.[1] However primary ones are extremely rare. Primary pigmented tumors of the leptomeninges include pigmented meningioma, malignant melanoma, meningeal melanocytoma, melanotic shwannoma, and melanoblastosis.[2,3] Among the reported primary meningeal melanocytic tumors, the majority are the malignant forms. Benign melanocytic tumors have been designated as "meningeal melanocytoma" which were first proposed by Limas and Tio (1972).[1] The differential diagnosis is often difficult among these tumors owing to their similar appearance on computed tomography (CT) and magnetic resonance imaging (MRI) studies, thus necessitating additional diagnostic confirmation by electron microscopy or immunohistochemical analysis. Furthermore, since the biological behavior, treatment, and prognosis of these lesions are different, it is important to make the correct pathological diagnosis. Here, we present a case of meningeal melanocytoma with clinicopathological, histological, and immunohistochemical studies.

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Case Report

A 40-year-old male presented with a history of gradually progressing weakness of both lower limbs and paresthesias since 1 year. On physical examination, motor power was 4/5 in both lower limbs with slightly increased reflexes. Bladder and bowel control was normal. On radiological investigations (CT and MRI), an intradural extramedullary mass measuring 1.5×1.0 cm was observed. The patient was operated and the excised mass was sent for histologic examination. Biopsy was received in multiple pieces and on gross examination the tumor was dark brown to tan in color, measuring together 1.8×1.0×0.8 cm. Microscopic examination revealed heavily melanin-pigmented cells, nuclei with reticulogranular chromatin and small nucleoli, moderate amount of eosinophillic cytoplasm with indistinct cell boundaries, and symplasmic appearance. A probable diagnosis of meningeal melanocytoma was made [Figures 1 and 2].

The diagnosis was confirmed on immunohistochemical analysis which revealed strongly positive expression of HMB-45 in the tumor cells. Vimentin and S-100 were also diffusely positive while neuron specific enolase showed focal and patchy positivity; however, epithelial membrane antigen (EMA) was distinctly negative [Figure 3].

Discussion

All melanin-producing cells are thought to be derived from the neural tube and neural crest. In normal humans, the

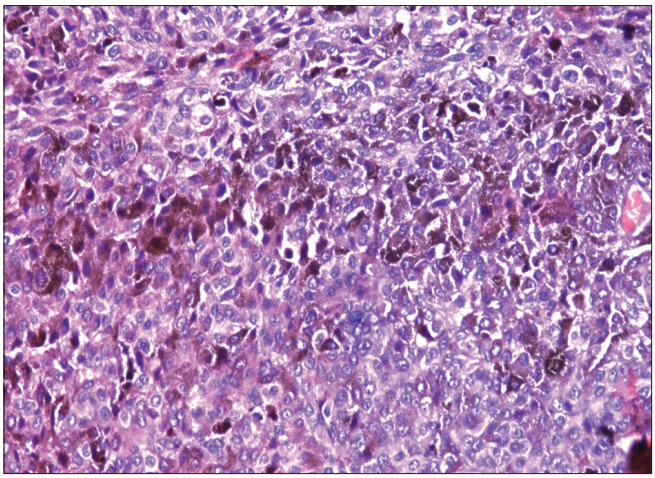

Figure 1: Microscopic section revealing melanin-pigmented cells with symplasmic cytoplasm (H and E; ×200)

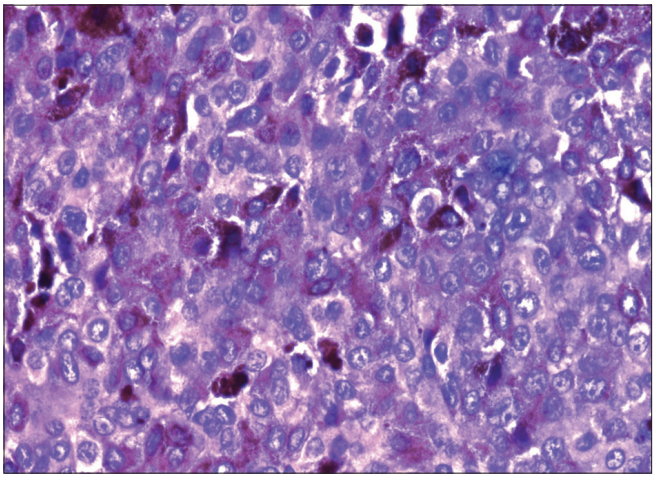

Figure 2: Cells show reticulo-granular nuclear chromatin and small nucleoli, cytoplasm contains melanin pigment (H and E; ×400)

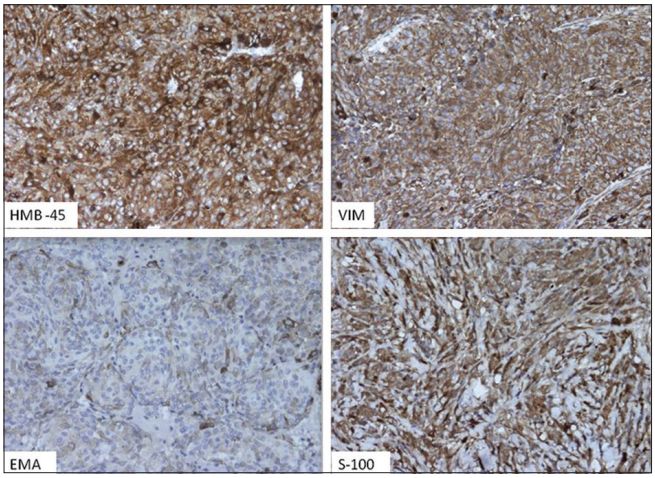

Figure 3: Immunohistochemical analysis revealing strong positive expression by HMB-45. Vimentin and S-100 show diffuse positivity and EMA is distinctly negative

melanin-producing cells of neural tube origin are found in the pigmented epithelial cell of the eye. Those of neural crest origin are melanocytes and rarely Schwann cells. These melanocytes are normally found in the leptomeninges covering the base of the brain and the brain stem. Consequently, the areas most commonly involved are the pons, cerebellum, cerebral peduncles, medulla, interpeduncular fossa, and inferior surfaces of the frontal, temporal and occipital lobes.[4,5] Melanin-producing tumors of neural crest derived cells found in meninges are (1) Blue nevi, (2) meningeal melanocytoma, (3) malignant melanoma, and (4) melanotic schwannoma.[1]

Meningeal melanocytoma and primary malignant melanoma of the leptomeninges are similar in their origin from leptomeningeal melanocytes, but actually represent both ends of the spectrum, ranging from a lesion that is benign in appearance and behavior to one that is malignant. However neither of these entities is associated with pigmented lesion elsewhere. Meningeal melanocytoma has a much better prognosis than their malignant counterparts. A variety of neurological and clinical features may be seen with meningeal melanocytoma, including frequent occurrence of hydrocephalus. Hydrocephalus is usually treated with placement of a ventriculoperitoneal shunt, but a filter must be added to the apparatus to prevent spread in the rare event of malignant transformation. Other clinical features include seizures, chronic basal meningitis, multiple cranial nerve palsies, chronic spinal arachanoiditis, psychiatric disturbances, still birth, intracranial hemorrhage, myelopathy, and radiculopathy.[5,6]

The gross appearance of meningeal melanocytoma as seen during surgery or at autopsy is that of a well-encapsulated, nodular, dark brown or black lesion that is firmly attached to the underlying leptomeninges. A meningioma may mimic this gross appearance if large amounts of haemosiderin are present within the lesion from previous episodes of hemorrhage.[3]

Histological examination of cells obtained from pigmented tumors is of limited value other than to demonstrate intracytoplasmic melanin. The cells have a uniform cytological appearance, without areas of anaplasia, necrosis, or significant mitotic activity. Ultrastructurally, meningeal melanocytoma contains a large number of melanosomes and premelanosomes at different stages of differentiation.[3,5] Additionally, meningeal melanocytoma cells lack desmosomes and interdigitating cytoplasmic processes characteristic of meningioma.[3]

Immunohistochemical analysis is indispensible in differentiating meningeal melanocytoma from other similar pigmented lesions.[7]

Meningeal melanocytoma is characterized by a positive immunohistochemical reaction to antimelanoma antibody (HMB45), S-100 protein, and vimentin antibodies and by a negative reaction to EMA.[2,3] Melanocytic meningioma is characterized by a positive reaction to EMA and vimentin,

Table 1: Immunohistochemical features of pigmented tumors of the meninges

	Meningeal melanocytoma	Melanocytic shwannoma	Melanocytic meningioma	Melanoma
HMB45	+	±	–	++
S-100	+	++	–	+
Vimentin	+	+	+	–
EMA	–	–	±	–
GFAP	–	±	–	–
Leu7	–	+	–	–

EMA = Epithelial membrane antigen; GFAP = Glial fibrillary acidic protein; + = Positive, ++ = Diffuse positivity, ± = Focal positivity, – = Negative

and by a negative reaction to HMB-45 and S-100 protein. The histological differentiation between malignant melanoma and melanocytoma can be even more difficult. The overall lack of mitotic activity, lack of nuclear pleomorphism and hyperchromaticity, and the indolent growth of the mass spanning more than 4 years all point to melanocytoma versus melanoma [Table 1].[5]

The imaging appearance of meningeal melanocytoma is variable depending on the degree of mineralization, and thus of limited value in differential diagnosis. The CT appearance is characterized by iso- to hyperattenuating lesions with variable contrast enhancement. The MRI appearance is strongly influenced by the paramagnetic effects of melanin, which cause shortening of T1 and T2 relaxation times. Therefore, the MRI appearance of these lesions is generally that of high signal intensity on T1- weighted images and diminished on T2-weighted images, with enhancement after contrast administration.[5]

Considering the good postoperative survival rate of patients with meningeal melanocytoma, the surgeon should be advised of this possible diagnosis in suspected cases of meningioma, especially when it involves the posterior fossa or Meckel's cave.[2] Accordingly, a maximum effort at tumor resection should be made, with expectations of good postoperative outcome. According to the results of some works of the last years, it seems appropriate to use postoperative radiotherapy for those patients with symptomatic residual, progressive or recurrent tumors not amenable to further resection.[8,9]

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