Meningeal melanocytoma

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Meningeal melanocytoma is a rare type of tumor that originates from melanocytes, which are the pigment-producing cells usually found in the skin and eyes. In some cases, these cells can be found in other parts of the body, including the meninges, which are the protective membranes that surround the brain and spinal cord. When a melanocytoma forms in the meninges, it is referred to as a meningeal melanocytoma.

Here are some key points about meningeal melanocytomas:

Rare Occurrence: Meningeal melanocytomas are considered rare tumors. They account for only a small percentage of all central nervous system tumors.

Benign Nature: Meningeal melanocytomas are typically benign (non-cancerous) tumors, which means they do not spread to other parts of the body (metastasize). However, they can cause symptoms by compressing nearby brain or spinal cord structures.

Location: These tumors are most commonly found along the base of the skull or in the spinal cord, although they can occur in other parts of the central nervous system.

Symptoms: The symptoms of meningeal melanocytomas can vary depending on their location and size. Common symptoms may include headaches, seizures, changes in vision, and neurological deficits such as weakness or numbness.

Diagnosis: Diagnosis is typically made through a combination of imaging studies, such as MRI or CT scans, and a biopsy to confirm the presence of melanocytic cells.

Treatment: Treatment of meningeal melanocytomas often involves surgical removal of the tumor, which can provide both diagnostic information and symptom relief. If the tumor is completely resected, it may not recur. However, in some cases, additional treatment such as radiation therapy may be considered.

Prognosis: The prognosis for meningeal melanocytoma is generally good when the tumor is benign and can be completely removed. The long-term outcome can be excellent, with a low likelihood of recurrence.

Meningeal Melanocytoma can occur along the neural axis most commonly in posterior fossa, adjacent to the cranial nerve nuclei, and Meckel's cave and in the foramen magnum. Within the spine, it present as intradural extramedullary masses, mostly found in the upper cervical region, as the melanocytes are most concentrated in this region. These tumours can be both intra-/extradural and intra-/extramedullary [1)].

Meningeal Melanocytoma were first described in 1972 by Limas and Tio as primary melanotic tumours of the leptomeninges [2)]

Classification

Intracranial Meningeal Melanocytoma

Spinal Meningeal Melanocytoma

Clinical features

They can present as diffuse disseminations within the subarachnoid spaces, space occupying solid masses within the central nervous system. They present as slowly growing mass lesions with focal neurological deficits due to the mass effect on the adjacent tissues [3)]

Differential diagnosis

Melanocytic tumors may be secondary to melanoma or be histologically benign, however, their diffuse nature makes them impossible to cure. Melanocytosis is a diffuse tumour that can form solitary extra-axial tumours, which invades the parenchyma and presents signs of malignancy with increased mitosis and Ki67, observed in 1 to 6% of immunopathological exams. Melanoma of the leptomeninges, presents signs of malignancy with anaplastic cells, which cluster in fascicles of melanin in the cytoplasm, with more than 3 atypical mitoses per field and Ki67 presenting in more than 6% of the immunopathological fields analysed [4)].

Meningeal melanomatosis is an infrequent tumor originating from the melanocytes in the leptomeninges and one of the recognized primary melanocytic tumors of the central nervous system. The average survival has known to be about 5 months. It can be associated with solid tumors, such as

meningeal melanocytomas [5].

Meningeal melanomatosis is an extra-axial well-encapsulated malignant tumour with diffuse meningeal growth and dark coloration (due to high melanin contents), while meningeal melanocytoma is the focalized benign variant. Melanocytic lesions may be secondary to melanoma or be histologically benign, however, their diffuse nature makes them impossible to cure. Melanocytosis is a diffuse tumour that can form solitary extra-axial tumours, which invades the parenchyma and presents signs of malignancy with increased mitosis and Ki67, observed in 1 to 6% of immunopathological exams. Melanoma of the leptomeninges, presents signs of malignancy with anaplastic cells, which cluster in fascicles of melanin in the cytoplasm, with more than 3 atypical mitoses per field and Ki67 presenting in more than 6% of the immunopathological fields analysed [6]

A preoperative differential of spinal meningeal melanocytomas is challenging due to their non-specific clinical and neurological presentations [7]

Treatment

Surgery is the preferred therapeutic approach, and total resection is associated with the best outcome. Patients with partial resection or tumor recurrence benefit from adjuvant radiotherapy, whereas chemo- or immunotherapies do not improve the disease course [8].

Outcome

Malignant transformation was described in 18 patients. Of these, 11 patients developed metastasis [9]

They typically behave as slow-growing, well-circumscribed tumours in the spinal cord, however, can also be found in the brain [10].

These tumors have better prognosis than their malignant counterparts [11].

Literature review

A literature review was performed using PubMed and Web of Science. All published cases were evaluated for location, sex, age, therapeutic approach, and outcome. In total, we included 201 patient cases in our meta-analysis.

The majority of MM was diagnosed more frequently in men between the third and fifth decade of life.

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Ricchizzi present the first retrospective meta-analysis of all MM cases published in the English language, including an evaluation of different treatment strategies allowing us to suggest a novel treatment guideline highlighting the importance of total resection for recurrence-free survival and characterizing those cases which benefit from adjuvant radiotherapy [12)]

Case reports

2023

Intermediate grade meningeal melanocytoma of the posterior fossa, GNAQ mutation-positive [13)].

2017

Padilla-Vázquez et al. present the case of a patient with long-term meningeal melanomatosis, with progressive neurologic deficit and characteristic radiologic features, and another case of meningeal melanocytoma.

Benign melanocytic neoplasms of the central nervous system must be treated aggressively in the early phases with strict follow-up to avoid progression to advanced phases that do not respond to any treatment method. Unfortunately, the prognosis for malignant melanocytic lesions is very poor irrespective of the method of treatment given [14)].

2015

A 21-year-old man with a previous history of recurrent lipothymia was admitted to the emergency department because of generalized seizures. Death occurred despite resuscitation. A medico-legal autopsy was performed. External examination of the body showed nonspecific asphyxia signs without any violence evidence. Necropsy noticed a brain edema with a dark color of the meninges especially in the frontal part. Histological examination concluded to diffuse meningeal melanocytoma with cerebral edema [15)].

A 31-year-old woman presenting with a 2-year history of amenorrhea and an intrasellar mass with suprasellar extension, suggestive of hemorrhagic pituitary neuroendocrine tumor.

Transsphenoidal surgical excision was difficult due to extensive bleeding from the lesion, and at the time, the tumor could not be diagnosed histopathologically. Six years later, Sakata et al. operated again because of tumor regrowth. Angiography revealed a hypervascular tumor, which was fed from the dorsal sellar floor. We had difficulty resecting the tumor, but achieved total removal. These case had typical radiographic characteristics of melanocytoma, revealed by both magnetic resonance

imaging and angiography. However, it was difficult to reach a final diagnosis. Further histopathological examination, including immunohistochemical and ultrastructural studies, was helpful for diagnosis of melanocytoma.

Primary sellar melanocytic tumors are derived from melanocytes in the meningeal lining of the sellar floor or in the diaphragm sellae, based on both embryological assumptions and the clinical findings of these case [16].

2011

Meningeal melanocytoma with malignant behaviour [17]

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