Spinal Meningeal Melanocytoma

- Meningeal melanocytoma of the central nervous system in children
- A rare case of multifocal craniospinal leptomeningeal melanocytoma: A case report and scoping review
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- Meningeal Melanocytoma- A Clinicohistological Masquerader
- Primary meningeal melanoma masquerading as neurofibromatosis type 2: illustrative case

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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].

Epidemiology

Meningeal melanocytomas are extremely rare benign tumours arising from the leptomeninges; 1 case presents per 10 million people every year [2]

They are most commonly located in the upper cervical region of the spinal cord due to the greater concentration of melanocytes [3]

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 [4].

Case reports

2023

A 60-year-old Chinese female who presented with numbness and weakness of the limbs for approximately 6 months. computed tomography (CT) and magnetic resonance imaging (MRI) revealed a dumbbell-shaped tumor inside and outside the cervical (C) spinal canal.

The patient was using CT and MRI. Subsequently, the patient underwent surgery, and low-grade melanocytoma was diagnosed pathologically.

Subsequently, the patient underwent a surgery, and the tumor was completely removed.

The tumor did not recur after 6 months.

This case suggested 2 “take-away” lessons: first, spinal meningeal melanocytomas may be dumbbell-shaped; and second, melanocytoma could appear as hyperintense, isointense, or hypointense on T2-weighted MRI [5].

A 56-year-old man who presented to our unit with a 4-month history of lower limb weakness and a sensory level at T6. Magnetic resonance imaging shows an intradural extramedullary tumour. The patient underwent a thoracic debulking of the lesion with neurophysiological monitoring. Histopathology confirmed the diagnosis of melanocytoma of meningeal origin, with a low mitotic count. Our patient recovered well post-operatively with no complications. Surgical resection is an effective method to manage this tumour; however, adjuvant radiotherapy is advised due to the risk of recurrence and malignant transformation [6].

2022

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. The patient present was diagnosed of a meningeal melanomatosis that developed two solid tumors related to an in vitro fertilization. The clinical course was rapidly fatal. Although the use of comprehensive diagnostic procedures, usually the final diagnosis of primary diffuse meningeal melanomatosis is postmortem, it would be advisable for the appropriate management of the patient to make a differential diagnosis and to be aware of the behavior of the tumor [7]

A case of intraspinal melanocytoma in a 57-year-old female, which clinically as well as radiologically mimicked other spinal lesions. The final diagnosis was confirmed on histopathology [8].

A case of a patient who reported constipation and abdominal pain around the umbilicus, which progressed into cord compression with lower extremity weakness and gait instability. Spinal magnetic resonance imaging (MRI) revealed a tumor at the level of T11, and she underwent gross total resection of the mass. Pathology demonstrated a meningeal melanocytoma with intermediate features. She received post-operative radiation therapy and had stable disease for three years, at which time she developed new weakness and drop metastases. This case represents a rare presentation of a rare disease, in which a spinal cord tumor presented with constipation and abdominal distress. Intradural-extramedullary tumors of the thoracic spine are most commonly nerve sheath tumors or meningiomas, but rare entities such as melanocytomas can present in this location; even more rarely, these tumors can have an aggressive course with delayed recurrence [9)]

2021

The case of a thoracic intramedullary meningeal melanocytoma in a patient unable to undergo an MRI.

This is the first reported S-100-negative case with genetic testing to support the diagnosis of a rare intramedullary melanocytoma [10)]

2020

A 30-year-old female harboring a C6-T1 ventrally located intradural extramedullary lesion compressing the cord anteriorly. The lesion was totally resected via an anterior approach with oblique corpectomy even if the usual treatment involves surgical removal through a posterior approach using a laminectomy or laminotomy.

There is no evidence of recurrence at 4-year follow-up records of the patient. We discuss the surgical approach of these rare lesions. [11)].

2016

A 41-year-old woman with primary multifocal meningeal melanocytoma in the spinal canal. Contrary to earlier reports, the tumors presented with a scattered appearance mimicking neurofibromatosis.

On admission, the cerebral MR images of the patient were normal, whereas the spinal MR images showed scattered multifocal nodules mimicking neurofibromatosis. Surgical resection of the responsible lesions was scheduled. In addition to this case presentation, relevant previous reports were reviewed, and the challenging diagnosis, management, and prognosis of meningeal melanocytoma are discussed.

Gross total resection of the two largest lesions was achieved, and histopathological examinations confirmed the diagnosis. Despite the benign histopathological findings, the patient had an aggressive clinical course. On follow-up at 18 months after surgery, she succumbed to the disease.

Clinicians should be alert to a potential aggressive clinical course of meningeal melanocytoma, despite its benign histopathological nature. Of particular note is multifocality and diffuse leptomeningeal hyperpigmentation, which may suggest a poor prognosis. A combined treatment including surgical resection and adjuvant radiotherapy should be considered, and long-term close

follow-up is necessary [12].

2015

A case of meningeal melanocytoma of the cervical region presenting with superficial siderosis. Extensive neuroradiological examination is necessary to locate the source of the bleeding in such patients. Usually, the patient will be cured by the complete surgical excision of the lesion [13].

2011

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 [14].

2010

Spinal meningeal melanocytoma: A rare meningeal tumor [15].

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