DOI: https://doi.org/10.4038/jdp.v19i1.7863 *Journal of Diagnostic Pathology 2024;19(1):8-11.*

Case report

A rare occurrence of a metastatic deposit of meningioma in a cervical lymph node

K.V. Chathurika Wijegunarathna, S. Roshana Constantine

Department of Histopathology, National Hospital of Sri Lanka

Submitted on 17.07.2024. Accepted for publication on 18.05.2025.

Abstract

Meningioma is a common primary central nervous system (CNS) tumour, in which recurrences and local invasion can occur depending on the CNS World Health Organisation (WHO) grade. Extracranial spread of meningioma is rare and mostly includes metastasis to liver, lung, mediastinum and bone. Cervical lymph node involvement is extremely rare and only a few cases have been reported. This is a case of a 57-year-old man, with a diagnosis of CNS WHO grade 2 meningioma who presented with cervical lymph node (level II) enlargement. Microscopically whorls and sheets of cells resembling a meningioma were observed with diffuse EMA positivity. Although it is a rare occurrence, the possibility of metastatic meningioma should be considered in the differential diagnosis of cases of cervical lymph node metastasis, in patients with primary meningioma. Extracranial spread of meningioma carries a relatively poor prognosis. Early diagnosis is important as the excision of primary/secondary disease followed radiotherapy is the currently recommended treatment.

Keywords: meningioma, extracranial spread, cervical lymph nodes

Introduction

Meningioma is a common tumour of the central nervous system (CNS), arising in the meningothelial cells from the arachnoid mater (1). The CNS World Health Organisation classification, classifies meningioma into three grades, which show increasing rates of recurrences and progression(2). Metastatic deposits from meningioma are rare, and reported in 1 in 1000 meningiomas (3). The most common sites are the liver, lung, mediastinum and bone (4). Cervical lymph node involvement is extremely rare (4). The few case reports available indicate that metastasis mostly develops from grade 2 and 3 meningioma, with it occuring more frequently in grade 3 meningioma (4). Meningioma usually carries a favourable prognosis with a five year survival of 88% (2). However in a metastatic setting, the five year survival rate is 65% and with increasing grade of the meningioma the prognosis becomes worse (4) (5). Due to its limited incidence, the treatment approach to metastatic meningioma is still under debate (5). When feasible, complete surgical removal of both the primary and metastatic tumors is currently the preferred initial approach, typically followed by radiotherapy (5).

Case report

A 57-year-old man, previously diagnosed one year ago with a CNS WHO grade 2 meningioma located in the left sphenoid wing, presented with a neck lump that had been present for one month. Magnetic resonance imaging showed an enlarged left level II cervical lymph node. A recurrence of the tumor was observed at its original location in the left sphenoid wing. No additional lymph node enlargement was detected. left cervical lymph node dissection was performed.

A 5cm lymph node was identified in the level II legion. Microscopy (Figure 1) showed that the lymph node was largely replaced by a tumour composed of whorls and sheets of cells with monomorphic round to oval vesicular nuclei and moderate eosinophilic cytoplasm. Nuclear inclusions were present. Psammomatous

Corresponding author: Dr Chathurika Wijegunarathna
Department of Histopathology, National Hospital of Sri Lanka, Colombo, Sri Lanka
wijegunarathnac@yahoo.com

calcifications were not seen. The mitotic count was 1/2mm^2.

Review of the previous histology (Figure 2) revealed a tumour with similar morphology, characterized by whorls and sheets of cells. Necrosis, sheeting architecture and small cell areas with high nuclear:cytoplasmic ratio were present. Mitoses were 3/2mm^2. Hence the tumour belonged to CNS WHO grade 2.

Immunohistochemistry for EMA (Figure 1), which is expressed in 80% of meningiomas (2), was performed in the lymph node deposit and demonstrated strong diffuse membranous positivity in the tumour cells. Since EMA can be positive in the epithelial tumours, a pancytokeratin (Pan CK) stain was performed(6), which showed complete negativity (Figure 1). PR, which is positive in approximately 66% of meningiomas(2), was negative. SSTR2A immunohistochemical marker which is expressed in almost all meningiomas (2) is not available in our setting. Facilities to identify molecular features such as monosomy of Chromosome 22 were not available.

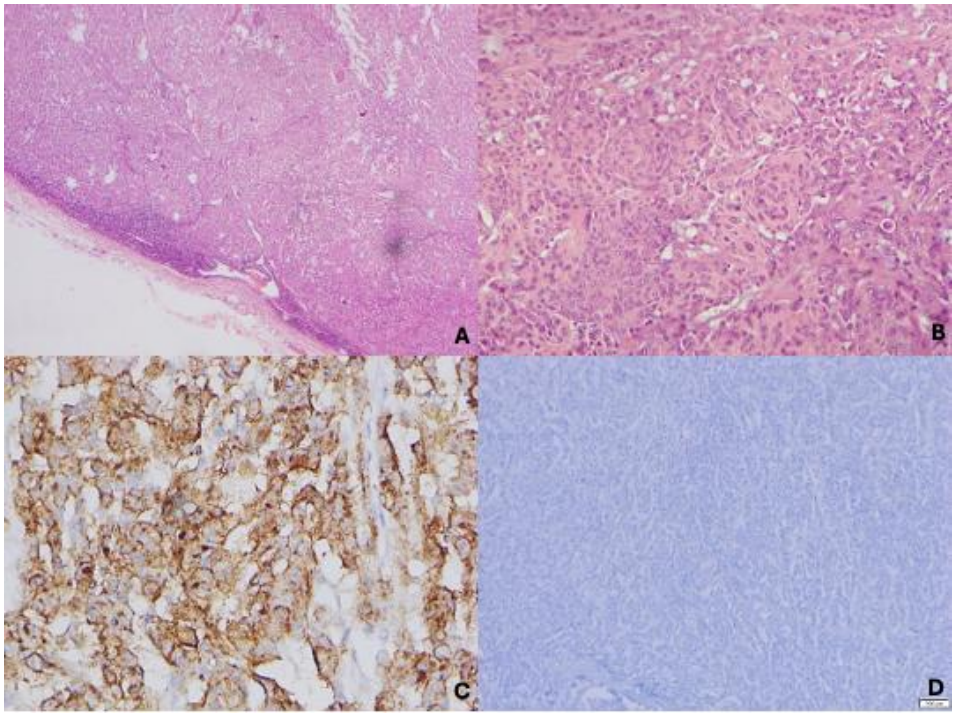

Figure 1: A) The lymph node was replaced by a tumour (Haematoxylin and eosin (H&E) x40); B) composed of cells arranged in whorls (H&E x100) C) The cells showed diffuse membranous positivity for EMA (EMA x100) D) and negativity for pancytokeratin (PanCK x40)

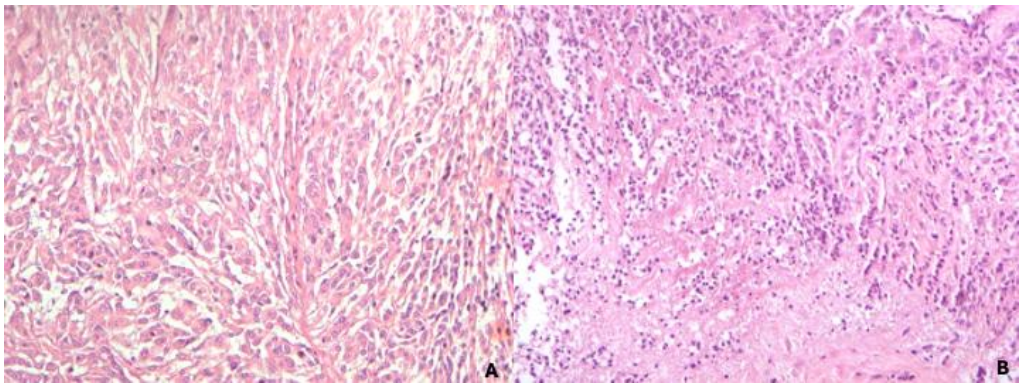

Figure 2: Sphenoid wing meningioma (CNS WHO grade 2). A) The tumour showed sheeting of cells (H&EX100) B) necrosis and small cells with high N:C ratio (H&EX100)

Radiological and clinical evaluation revealed no evidence of additional lymph node enlargement, nor any lesions in the lungs, liver, or bones, and no signs of an underlying epithelial malignancy. The tumour showed a strong morphological resemblance to a meningioma with EMA positivity. Pan CK negativity excluded a tumour of epithelial origin (6). The strong diffuse positivity of EMA (7) and the absence of morphological features of a paraganglioma, excluded the possibility a paraganglioma. Since this tumour did not show morphological resemblance to a lymphoma, it was not considered as a differential diagnosis. Based on these clinical and immunomorphological features it was diagnosed as a metastatic deposit of the previously diagnosed meningioma.

Following the diagnosis, the patient underwent re-excision of the recurrent meningioma at the left sphenoid wing followed by radiotherapy to the site. The patient is currently disease free at seven months of follow up.

Discussion

Meningioma is a primary CNS tumour, which arises from arachnoid cap cells and comprises 37.6% of all primary CNS tumours (2). They most often occur in adults and the incidence increases with age. The median age of diagnosis is 66 years and females are affected more than males (2). Currently the most established risk factor for meningioma is ionizing radiation (2). Molecularly it is associated with monosomy of chromosome 22 in approximately 80% of cases (2).

Grading of the meningioma is done by CNS WHO grading system which assigns three grades (2). They are categorized by mitotic count, brain invasion, specific morphologic subtype, necrosis, increase cellularity, sheeting, small cells with high N:C ratio, prominent nucleoli and anaplasia (2). The presence of *TERT* promoter mutation and/or homozygous deletion of *CDKN2A/CDKN2B* automatically makes it a CNS WHO grade 3 tumour (2). CNS WHO grade 2 and 3 tumours have increasing rates of recurrences and progression (local spread), and CNS WHO grade 3 tumours are associated with a worse prognosis. However, most meningiomas are WHO grade 1 tumours and have a benign clinical course (2).

Extracranial metastasis of meningioma is rare and is reported in 1 in 1000 meningiomas (3). It was first reported in 1886, and since then there have been reports describing varied presentations of this rare occurrence (3, 4). It usually occurs with CNS WHO grade 2 and 3 tumours and is most often associated with CNS WHO grade 3 tumours (8). The common sites of extracranial spread are the liver, lung, mediastinum and bone (9). Metastatic involvement of the cervical lymph nodes is exceedingly rare, with only 21 cases reported to date (9).

The pathogenic pathways of extracranial metastases from meningioma are still largely unknown (10). Historically it is assumed that iatrogenic seeding might play a role, where the disruption of the tumour during resection of the primary lesion could lead to metastatic spread via the lymphatics and venous sinuses (10).

Risk factors for distant metastasis of meningioma include history of open cranial neurosurgery, venous sinus infiltration, local recurrence, and histological malignancy (CNS WHO grade 3 tumours)(10).

Patients with metastatic meningioma carry an unfavorable prognosis when associated with CNS WHO grade 2 and 3 tumour (5). Due to their rarity, there are no specific guidelines on the screening and management of these patients (4).

Improved and more targeted detection of these cases is required, as extracranial metastasis can manifest decades after initial diagnosis and are often detected by incidental imaging, at which time management is frequently limited to palliative care (3). Several tailored imaging modalities have been suggested for screening. Nuclear medicine techniques such as 68Ga-DOTANOC PET/CT imaging have been used to detect extracranial metastases with high signal intensity, as there is high expression of somatostatin receptors in meningiomas (4). Similarly, somatostatin

receptor scintigraphy tagged with the ^{111}indium-octreotide and ^{111}In -pentreotide radioisotopes has shown to be useful for identifying extracranial masses (4).

Due to the low incidence of extracranial metastases, there is a lack of consensus on the most effective management strategies (4). This dire situation is also present in the management of recurrent intracranial WHO grade 2 and 3 meningiomas and reflects the paucity of drug therapies for these tumours. While metastatic meningiomas represent an aggressive and extreme end of the spectrum, the failure to manage them is also reflective of the failure to control the primary disease (5).

A limitation in the diagnostic work-up of this case is the absence of molecular pathological analysis, owing to the unavailability of the necessary facilities. Notably, SSTR2A—a marker commonly expressed in the majority of meningiomas—is not accessible at our institution.

In conclusion, it is important to consider a metastasizing meningioma when a patient with a history of meningioma presents with cervical lymph node enlargement. Since these patients go undiagnosed for years it's important to establish better screening modalities as progressive metastatic disease carries a bad prognosis.

References

1. Ostrom QT, Cioffi G, Gittleman H, et al. CBTRUS statistical report: primary brain and other central nervous system tumors diagnosed in the United States in 2012–2016. *Neuro-oncology*. 2019;21(Supplement_5):v1-v100.

2. World Health Organization Classification of Tumours of the Central Nervous System. 5 ed ed. Lyon: International Agency for Research on Cancer; 2021.

3. Watanabe G, Young K, Rauber E, et al. A systematic review of extraneural meningioma metastasis: timing, evolution and outlook. *J Neurooncol*. 2024;168(2):187-196. doi:10.1007/s11060-024-04659-6

4. Himič V, Burman RJ, Fountain DM, Hofer M, Livermore LJ, Jeyaretna DS. Metastatic meningioma: a case series and systematic review. *Acta Neurochir (Wien)*. 2023;165(10):2873-2883. doi:10.1007/s00701-023-05687-3

5. Khan MF, Mohamed M, Young K, et al. Extracranial Meningioma Metastasis: A Systematic Review of Clinical Characteristics, Management Strategies, and Outcomes. *Proceedings of IMPRS.* 2023;6(1).

6. Menz A, Gorbokon N, Viehweger F, et al. Pan-keratin Immunostaining in Human Tumors: A Tissue Microarray Study of 15,940 Tumors. *Int J Surg Pathol*. 2023;31(6):927-938. doi:10.1177/10668969221117243

7. Williams MD. Paragangliomas of the Head and Neck: An Overview from Diagnosis to Genetics. *Head Neck Pathol*. 2017;11(3):278-287. doi:10.1007/s12105-017-0803-4

8. Ruff C, Bombach P, Gohla G, et al. Evolution of a Meningothelial Meningioma: From WHO Grade 1 to Anaplastic Grade 3 with Extracranial Metastasis Including Extensive Liver Metastasis. *Diagnostics*. 2024;14(7):676. doi:10.3390/diagnostics14070676

9. Coca-Pelaz A, Bishop JA, Zidar N, et al. Cervical Lymph Node Metastases from Central Nervous System Tumors: A Systematic Review. *Cancer Manag Res*. 2022;14:1099-1111. doi:10.2147/CMAR.S348102

10. Lee GC, Choi SW, Kim SH, Kwon HJ. Multiple extracranial metastases of atypical meningiomas. *J Korean Neurosurg Soc*. 2009;45(2):107-111. doi:10.3340/jkns.2009.45.2.10