Secretory Meningioma Mimicking Malignancy

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Introduction

Meningiomas are common tumours of the central nervous system (CNS) and majority of them are benign (World Health Organisation Grade 1) [1]. Among the variants, secretory meningioma (SM) is recognized by its gland-like formations [2], which mimics metastasis of adenocarcinoma to the brain. It is also known to be vascular and produces significant peritumoural brain oedema (PTBE) leading to increased perioperative morbidity. These characteristics justify its recognition, to prevent misdiagnosis by the pathologist and care during resection by the neurosurgeon.

Case Report

A 45 year old lady presented with right sided headache, unsteady gait, numbness right side of face with pain and watering of the right eye for six months. She had diplopia for distant objects since two months, irritability and forgetfulness for one month. Clinical evaluation revealed an ataxic gait. There was no evidence of raised intracranial tension. She had V and VII cranial nerve deficits on the right side.

The clinical diagnosis was a left cerebellopontine angle space occupying lesion (meningioma/schwannoma trigeminal nerve). Contrast enhanced computed tomography (CECT) scan of the brain revealed a brightly enhancing, extra-axial mass lesion arising from the petrous apex on the left side. This mass was compressing and displacing the midbrain to the right side and had associated perilesional oedema. Based on the CT findings a diagnosis of meningioma of the petrous apex was made (Fig. 1). At operation, a vascular tumour near the petrous apex (left) was excised. Post operative CECT showed a mild hydrocephalus with brain stem compression. The patient had persistent V nerve sensory loss and lower motor neuron palsies of the VII, VIII and IX cranial nerves.

On microscopic examination, the excised tissue showed a meningothelial pattern. The cells had little pleomorphism and no mitosis; focally nuclei showed intranuclear clearing. At many places cells were arranged around luminal spaces containing eosinophilic amorphous material

(‘pseudopsammomma’ bodies) (Fig. 2). These were positive for periodic acid schiff (PAS) stain and immunoreactive to carcinoembryonic antigen (CEA) (Fig. 2 Inset). The tumour cells showed strong positivity for cytokeratin (CK) and a diagnosis of secretory meningioma was made. The patient is gradually recovering, on follow-up over a four month period but has persistent V nerve (sensory) deficit.

Discussion

Meningiomas are benign tumours originating from the meninges of the brain and spinal cord. They occasionally demonstrate malignant potential (WHO grades II and III). A large number of histological variants have been identified, some prognosticating a more aggressive course, while others pose a diagnostic dilemma [1]. Secretory meningioma is one such variant with a unique appearance on imaging and microscopic evaluation [2]. These tumours have a marked female predominance (male:female ratio of 9:1) and preferentially involve the frontal meninges and sphenoid ridge [3]. On clinicoradiologic evaluation, this subtype may present with PTBE giving an ominous picture [4].

The tumour represents a highly vascular meningothelial meningioma with glandular differentiation and perivascular arrangement of cells. It is marked by the presence of small, round, eosinophilic, strongly PAS positive, diastase-resistant and ‘pseudopsammomma’ bodies [4]. Ultrastructurally, these are extracellular amorphous debris, within well developed lumina formed by cells with typical epithelial features like tight junctions and villi. The tumour cells are positive for CK while other meningiomas are weakly positive for CK, but strongly express epithelial membrane antigen (EMA). The eosinophilic bodies stain positive for CEA, IgA and IgM. Serum CEA levels may also be high (which is an occasional occurrence in meningiomas), demonstrating the true secretory nature of the tumour [5].

The pitfall in diagnosis lies in the morphologic

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Fig. 1 : Axial section of contrast enhanced CT scan of the brain showing a brightly enhancing extra-axial mass lesion arising from the petrous apex on the left side.

‘epithelial’ resemblance to metastatic carcinoma (including expression of cytokeratin and CEA) commonly from breast (which has a predilection for dural metastasis) or colon. This is compounded by the possibility of elevated serum CEA in both conditions. Ironically, coexistence of breast carcinoma and meningioma is not unusual [6]. Careful evaluation for cytologic features of malignancy (pleomorphism and atypia) have to be relied upon to favour a metastatic diagnosis. On the other hand, vimentin coexpression is not seen with breast carcinoma but is constantly present in meningiomas. Combination of cytokeratin subsets may differentiate this primary benign tumour of CNS from a metastatic adenocarcinoma (CK7+,CK20- for SM and CK7-,CK20+ for adenocarcinoma) [7].

Peritumoural brain oedema is an adverse prognostic factor in these otherwise benign tumours and is a harbinger of perioperative morbidity and mortality [8]. The oedema is probably the result of a defective efferent venous drainage from the tumour [9].

Recent interest in molecular pathways of tumorigenesis has revealed association of some histologic types of meningioma with neurofibromatosis 2 (NF2) mutations. These are seen in 30-60% meningiomas but are consistently absent in SM, indicating alternative pathways [10].

Secretory meningioma is a unique variant with a distinctive histologic phenotype which is likely to pose a diagnostic dilemma and adversely affect peri and postoperative recovery due to the associated vascularity and oedema.

Fig. 2 : Meningotheelial cells in sheets with gland-like lumina enclosing eosinophilic ‘pseudopsammoma’ bodies (Haematoxylin and Eosin x 200). Inset: Secretions are strongly immunoreactive to carcinoembryonic antigen (CEA) (IHC with DAB x 40).

Conflicts of Interest

None identified

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