Malignant Meningioma - Clinicopathological Study of an Unusual Presentation of a Rare Malignancy

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Introduction

Malignancy in meningiomas is rare and very rapid extracranial metastasis to the cervical and abdominal lymphnodes is even rarer. Presently the aggressive or malignant phenotype of meningiomas are difficult to characterize due to broad spectrum of behaviour exhibited by meningiomas. In practical use, the diagnosis of malignant meningioma is reserved for those tumours displaying histologic evidence of brain invasion or evidence of distant metastasis, features that are not easily assessed. Traditionally, pathologists are reluctant to classify meningiomas as malignant unless definite brain invasion is present. Less than 1% of all meningiomas and less than one third of those designated as malignant, metastasize and would by definition need vascular invasion, occult or primary, to accomplish distant spread. A very unusual case is presented who was operated as a case of typical meningioma, was reported histologically as malignant meningioma and developed very rapid, wide spread metastasis to all lymph nodes and lungs in the immediate postoperative period.

Case Report

A 45 year old male, known case of hypertension, presented with complaints of headache, slurring of speech, progressively increasing weakness of right lower limb and incontinence of urine, of two months duration. Fundus examination revealed gross bilateral papilloedema. Spiral CT scan brain showed a 5.5 cm well defined heterogenous density mass lesion with perilesional oedema in left frontal parasagittal region, suggestive of meningioma. Frontal craniotomy with piecemeal excision of left parasagittal meningioma was done, tumour was found to be attached to dura and superior sagittal sinus, was pinkish in colour with moderate vascularity, firm to soft in consistency and had a cavity. Tumour was excised in toto.

Histopathological examination revealed a hypercellular, anaplastic tumour with extensive geographical necrosis (Fig 1). There was cytological atypia with moderate nuclear pleomorphism, prominent nucleoli, frequent mitoses (>20/10 HPF) and haemorrhages (Fig 2). Focal areas of

meningothelial whorls and few syncytial cell clusters could be made out in some of the sections while others revealed evidence of glial reaction. The tumour did not show any psammoma bodies, papillary structures or capillary-like channels, no brain invasion could be made out because adjacent cortex was not included in the specimen. Opinion of malignant meningioma (WHO Grade III) was given. Immunohistochemistry revealed tumour to be vimentin and epithelial membrane antigen positive and negative for GFAP. Focal positivity was also observed with cytokeratin.

On examination in the immediate post-operative period, patient was detected to have a firm to hard mass in the neck near the left thyroid region. Possibility of a primary thyroid malignancy with metastasis to the brain was thought of. But FNAC of this swelling revealed metastatic deposits in cervical lymph node with cytomorphology similar to that of primary in meninges rather than that of a thyroid. USG identified both thyroid lobes clearly.

At this stage, CT chest and abdomen was done and it revealed multiple lung metastasis and extensive metastasis to supraclavicular, para-aortic and paracaval lymphnodes. Biopsy of neck mass was done and it confirmed metastasis of malignant meningioma with lymphatic embolisation (Fig 3). The metastatic deposits showed meningeothelial differentiation immunohistochemically and immunocytochemistry for thyroglobin was negative.

Discussion

A concise definition of malignant meningioma remains elusive and controversial. The situation is complicated by the fact that a histologic and biologic continuum probably exists between atypical or aggressive meningiomas and malignant meningiomas. Nevertheless, the modified WHO system of classification has proved useful in identifying meningiomas with potential for either aggressive future behaviour or for recurrence. The term malignant is usually applied to both WHO grade III and grade IV tumours. The system of classification proposed by WHO in 1993 [1], revised in 1997 and again in 2000, is based on cytological features of anaplasia rather than purely histopathologic descriptions. This system classifies meningiomas as meningioma (typical),

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Fig. 1 : Malignant meningioma. Architectural disarray, hypercellularity and extensive necrosis (H&E, low power)

Fig. 2 : Mitotically active malignant meningioma shows frank anaplasia, nuclear pleomorphism and prominent nucleoli (H&E, high power)

atypical or malignant meningiomas based on six individual features of anaplasia : loss of architecture, hypercellularity, nuclear pleomorphism, mitotic figures, focal necrosis and brain invasion.

Due to rarity of malignant meningioma, little data exists on its exact incidence and behaviour. Using WHO classification and its modifications mentioned above, benign meningiomas represent 93% (grade I) and atypical tumours 5% (grade II); about 2% are anaplastic or sarcomatous (grade III & IV) [2]. Papillary, haemangiopericytic and angioblastic varieties of meningiomas show more aggressive growth and recurrences. Arie Perry et al [3] examined 116 patients diagnosed as malignant meningioma and found that the first diagnosis of malignant meningioma was based on brain invasion in 86% of patients, frank anaplasia in 26% of patients and extracranial metastasis in 2% of patients, the numbers adding up to more than 100% because some tumours were both brain invasive and anaplastic. Overall, anaplastic grade applies only to less than 1% of all meningiomas.

Meningiomas pose definite diagnostic and

Fig. 3 : Microphotograph cervical lymph node showing metastatic deposits of malignant meningioma (H&E, low power)

therapeutic challenges to pathologists, neurosurgeons and oncologists. Recent advances in neuropathology of meningiomas have produced more specific histopathologic and cytological criteria for classifying these tumours and more sophisticated markers of aggressive behaviour. Only those neoplasms exhibiting morphologic or immunophenotypic evidence of an origin from meningotheial cells should be labelled as meningiomas.

Distinction of the meningiomas from potential counterfeits sometimes requires electron microscopy or immunocytochemical assay. The former's most constant and distinctive ultrastructural feature is the complex interdigitation of tumour cell processes without intervening basal lamina material. Immunohistochemistry regularly demonstrates vimentin positively, regardless of a given meningioma histologic pattern [4]. Of particular interest is the observation that a large majority of meningiomas exhibit membrane as well as diffuse cytoplasmic immunolabelling for epithelial membrane antigen, a feature foreign to tumours of pericytic, schwannian or fibrocytic derivation. Reactivity for S-100 protein, if present, is weak and cytokeratin expression is exceptional.

Distant blood-borne metastases reflect biologic malignant behaviour. A few malignant meningiomas have been reported to have metastasized through the cerebrospinal fluid, while pulmonary, abdominal or bone metastasis are very rare [5]. The distant metastases recorded in the literature are those derived from angioblastic varieties that would now be classified as meningeal haemangiopericytomas [6]. Venous sinus invasion is a feature of many meningiomas and is not predictive of haematogenous dissemination or germane to the diagnosis of a given lesion as atypical or malignant.

In our case the diagnosis could be made on histology on the basis of the WHO criteria. Finding of cervical

mass in the immediate postoperative period raised the possibility of a thyroid malignancy but morphology of metastatic deposits and results of immunohistochemistry confirmed the dissemination of malignant meningioma. Extracranial wide spread lymphatic metastasis of a meningioma is very uncommon and has not been mentioned in literature.

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