Case report:

A rare case of Spinal Papillary Meningioma

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Abstract:

Meningiomas are common intracranial neoplasms, while papillary mengiomas are the rare variety of neoplasms which are mostly located in the cerebral convexity. Papillary meningiomas in spinal cord is an extremely rare entity. We report a case of intramedullary, extradural, spinal papillary mengioma at lumbar region in a 49 year old female patient.

Keywords: Meningioma, Spinal meningioma, Papillary Meningioma

Introduction:

Meningiomas are fairly common Central Nervous System (CNS) neoplasms, arising from the arachnoidal cells, have a large variety of histopathologic appearances. Amongst these papillary meningioma (PM) is a rare variant that represents less than 1% of all meningiomas, with a higher gradation due to the likelihood of aggressive behaviour and recurrence. Until now, less than 100 intracranial PM cases have been reported in the English literature since 1938, very few cases of spinal papillary mengiomas have been reported till now[1]. Here, we report a rare case of papillary meningioma arising from the meninges of the spinal cord level L1-L2 in a middle-aged lady.

Case report:

A 49 year old lady presented with a history of low backache persisting for a year, radiating to bilateral lower limbs since a week. On the day of admission, she had difficulty standing in an upright posture and walking and had diminished sensations below bilateral knee. Patient was moderately built and nourished. Her vitals were stable. CNS examination revealed complete loss of sensations below the knee and motor power was 0/5 with absent knee, ankle jerks. Rest of the systemic examination were normal. The biochemical and haematological investigations were within the normal limits. A Magnetic Resonance Imaging (MRI) done showed an intradural, extramedullary, Gadolinium enhancing mass lesion centred at L1-L2 level, measuring 38 (craniocaudal)x 14 (mediolateral) x 11(anteroposterior) millimetre. The mass was situated just below the conus medullaris, distorting and compressing the cauda equina, and completely effacing the subarachnoid space.[Fig-1a] The patient underwent L2-L3 laminectomy and mass excision under the working diagnosis of Neurofibroma. A total excision specimen was received. The lesion was well circumcised, pale brown tissue mass measuring 3x1.5x0.5cm with attached linear fragment fibrous tissue measuring 0.5cm. Outer surface was congested. Cut surface appeared pale brown with an irregular pale white area measuring 1x0.5cm along with areas of haemorrhage. A few cystic areas were identified at the periphery, each measuring 0.3x0.3cm.[Fig-1b] Microscopy showed lesional tissue comprising of tumour cells arranged in numerous papillary and pseudopapillary formation around the blood vessels and in sheets. Tumour cells clusters in sheets were separated by thin walled blood vessels. Tumour cells

were uniform round to oval, having round to oval nucleus and fine chromatin, with moderate to abundant granular cytoplasm. Some of the tumour cells were arranged around blood vessels giving pseudorosette appearance. Numerous mitotic figures were seen. Amidst the tumour cells, there were numerous blood vessels and areas of haemorrhage, necrosis and cystic change. [Fig-1c] Immunohistochemistry showed Progesterone receptor positivity of 50-60% [Fig-1d], and MiB-1 labelling index of 4-6%. Features were consistent with Papillary Meningioma. Post operative period was uneventful. There was no recurrence of the tumour during the follow up period of one year.

Discussion:

Meningioma is one of the most common tumours of the CNS which accounts for 22.8%-34.7%. There are 15 subtypes in the 2007 World Health Organization (WHO) classification of meningioma of CNS[2,3].

Papillary meningioma (PM) is a rare variant of meningioma, representing less than 1% of all meningiomas. They are defined as a subtype of "malignant meningiomas" (WHO Grade III). The first case of PM was described by Cushing and Eisenhardt in the year 1938[1]. Since then less than 100 intracranial PM cases have been reported as isolated case reports in the English literature, but there are hardly few cases which have been reported as spinal PM. The biological behaviour of spinal PM is poorly understood, partly due to limited number of reported cases.

PM are more commonly seen in male patients and tend to occur in younger patients[4,5]. They are predominantly located in the supratentorial compartment though rare locations like posterior fossa, jugular foramen and occulomotor nerve have been described[6)]. Patients with PM usually manifest symptoms caused by intracranial hypertension, such as severe headache, vomiting, dizziness, blurred vision, gait disturbance and limb weakness[5]. PM are known to have an aggressive behaviour and recurrences.

CT or MR images of PM show irregular tumour shape, severe peritumoral brain edema, heterogeneous enhancement with gadolinium, tumour invasion into the adjacent brain and abundant blood supply to the tumour. The presence of tumoral cystic change is a rare finding in a typical benign meningioma, although it has been frequently reported in PM .There were area of cystic change in our case. PM frequently metastasise to pleura[7], lung[8] and liver[1], or most commonly diffuse cerebrospinal metastasis[9]. Extracranial metastasis is reported to occur in approximately 0.1% of meningiomas of all grades[10]. Ludwin et al analysed 17 cases of PM. According to these authors, it frequently occurs in children (41%) showing frequent mitosis, local recurrence and brain invasion[1]. There was no evidence of metastasis or recurrence of the tumour in our case.

Histologically, the tumour shows meningothelial cells in sheets, perivascular pseudo papillary pattern and focal well-formed papillae surrounding a central fibrovascular core, which is characteristic. Intratumoral necrosis and mitosis was frequently observed on routinely stained sections. The neoplasms most likely to be confused with a papillary meningioma are metastatic carcinoma, chemodectoma, ependymoma, choroid plexus papilloma, astroblastoma, and amelanotic melanoma. There could be a diagnostic overlap with papillary ependymoma which could be resolved by Immunohistochemistry for Vimentin, EMA,GFAP, Progesterone Receptor positivity and MiB-1 labelling index[11,12]. GFAP expression is rarely seen in PM[13]. In this particular case, the latter two were done, which favoured the diagnosis of Papillary Meningioma.

Conclusion:

Papillary meningioma arising from the spinal meninges is a very rare entity. PM has a tendency to present in middle-aged patients, and immunohistochemistry is helpful to differentiate from other primary and metastatic

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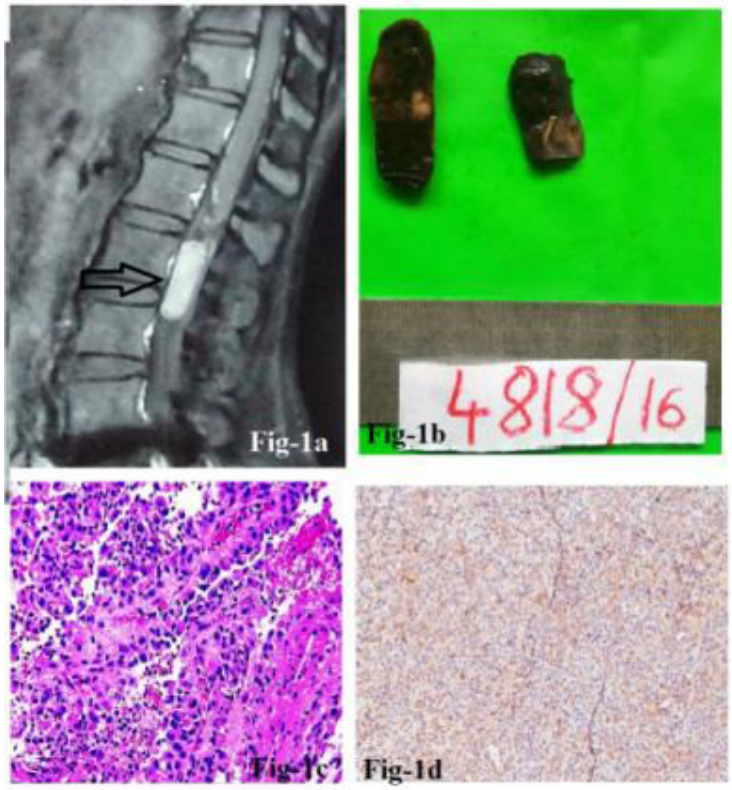
tumours. Follow up of the patient is essential as these tumours are very aggressive, rapidly metastasising and are known for recurrences. MIB-1 labelling index is likely to predict the recurrence.



Legends:

Fig-1a: Magnetic Resonance Imaging of Spinal Cord showing an intradural, extramedullary enhancing mass lesion (Papillary Meningioma) located at L1-L2 level of spinal cord. (Marked by the arrow)

Fig-1b: Gross of Papillary Meningioma showing grey white lesion and attached fibrous tissue fragment

Fig-1c: Microscopy of Papillary Meningioma showing perivascular papillary pattern (H&E x100)

Fig-1d: Tumour cells showing Progesterone Receptor positivity

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