Familial Central Nervous Malignancies Focus on Spinal Meningioma: Incidence in First Degree Relative

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Abstract

Meningiomas are extramedullary, intradural tumors arising from meningothelial arachnoid cap cells within the spinal dura. Spinal meningiomas are the most common spinal tumors in adults, accounting for up to 38% of intradural spinal tumors but only for 6.5% of overall craniospinal tumors in the adult age group. Similar histological subtypes are observed in both intracranial and spinal meningiomas, including meningothelial, metaplastic, psammomatous, transitional, atypical, and clear cell types. Multiple genes have been associated with spinal meningiomas like deletion of chromosome 22q and of its associated gene NF2 in cases of spinal meningioma. We are presenting a case of Intradural meningioma in a 30 year old female and her 50 year old mother. Keywords: Meningiomas, meningothelial, metaplastic, psammomatous.

INTRODUCTION

Spinal meningiomas usually occur after the fourth decade of life, over 70% of the patients are between the ages of 40 and 70 years with a mean age of 50 years, with similar frequency as the nerve sheath tumors, representing approximately 25% of all spinal cord tumors, 40% of intradural extramedullary tumors, without invading pia mater [1-8]. Most meningiomas have a significant predilection for females 75% - 85% of cases; arising primarily in the thoracic region - approximately 80%; the cervical region is affected less often; lumbar and sacral tumors are relatively rare [9]. Meningiomas typically grow slowly and usually with benign character, with a region of dural attachment, often seen dorsal-lateral, in a globoid configuration; rarely “en plaque meningiomas” - as a carpetlike [10, 11]. Clinical findings vary from mild to significant neurologic dysfunction; the most frequent clinical findings are back pain, sensori-motor deficit and sphincter dysfunction [9].

CASE REPORT

30 year old female presented with the history of lower back pain radiating down to both legs, numbness and weakness in both legs with loss of sensation of touch, and temperature variations since 3 months, and difficulty maintaining balance which was progressive for 2 months. On examination, patient had spastic paraparesis of bilateral lower limb with power 2/5. MRI revealed D9-D10 intradural tumor of spine. D9-D10 laminectomy with gross total excision of tumor was done which revealed firm/hard tumor densely attached with dura and surrounding nerve roots. The patient showed some improvement in her neurological deficits after surgery and was discharged after 1 week with close follow up.

5 years ago, the patient’s 50 year old mother had similar symptoms of weakness in both legs which progressively worsened. On examination, patient had spastic paraparesis of bilateral lower limb with power 2/5. MRI showed D4-D5 intradural extramedullary lesion in left lateral thecal sac with dural enhancement. D4-D5 laminectomy with partial excision of tumor was done. Histo-pathology showed meningotheliomatous meningioma (WHO grade-1). Patient was discharged in stable condition with slight improvement in the neurological deficits. Follow up showed substantial improvement in the patient’s weakness.
Fig A,B: MRI Dorso-Lumbar spine showing intensely enhancing Intradural extramedullary focal lesion on left side of spinal canal at D9-10 level compressing/dispacing the cord with cord edema. Fig C,D: Before and after removal of tumor surgically.

Fig E,F: MRI Dorso-lumbar spine shows well defined homogenously enhancing intradural extramedullary lesion in left lateral thecal sac at D4/D5 level with dural enhancement, causing right-wards displacement and compression of the cord with edema from D3 to D7 levels.
**DISCUSSION**

Familial nervous system cancers are rare and limited data on familial aspects are available particularly on site-specific tumours. Studies have been conducted to analyse familial risks of nervous system tumors using standardized incidence ratios.

Standardised incidence ratios (SIRs) are calculated for offspring of affected relatives compared with offspring of non-affected relatives. Standardised incidence ratios (SIRs) are used to measure the cancer risks for offspring according to occurrence of cancers in their families [12]. According to the study, standardised incidence ratios were calculated for offspring whose parent, sibling or parent and sibling had the same, concordant cancer, that is using parents or sibling as probands. The SIR of brain tumours was 1.7 in offspring of affected parents; it was 2.0 in siblings and 9.4 in families with a parent and sibling affected. For spinal tumours, the SIRs were much higher for offspring of early onset tumours, 14.0 for offspring of affected parents and 22.7 for siblings. The SIRs for peripheral nerve tumours were 16.3 in offspring of affected parents, 27.7 in siblings and 943.9 in multiplex families. Brain cancer accounted for 87.6% of all offspring cases and 93.7% of the familial cases, spinal tumours accounted for 6.7% of all and 1.3% of the familial cases, peripheral tumours accounted for 5.7% of all and 6.0% of the familial cases. The early onset cases were a minority of all cases at each anatomic site but among familial spinal and peripheral nerve patients they were the majority. For concordant anatomic sites, peripheral nerves showed the highest risk, followed by the spine and the brain.

**CONCLUSION**

Some 90% of nervous system tumours are located in the brain whereas spinal and peripheral nerve tumours account for the remainder [13]. Recently large epidemiological studies on nervous system cancer have been carried out but a few environmental risk factors have consistently been identified [14]. Therapeutic and low-level irradiation, hereditary syndromes and family history remain as the established risk factors of nervous system tumours [15-18]. Less than 3% of patients with nervous system tumours have a first-degree family member diagnosed by these neoplasms [18]. Because of the low incidence and rarity of familial cancers many of the published genetic epidemiological studies have not been able to distinguish anatomic locations or tumour types with sufficient numbers of cases [19-21]. But some studies have shown a promising relation of familial spinal cancers especially meningiomas.

**REFERENCES**


