

Intramedullary Spinal Schwannoma – A Case Report

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Abstract

Intramedullary spinal schwannomas are rare tumours and are usually associated with neurofibromatosis types 1 and 2. Intramedullary spinal schwannomas without neurofibromatosis are very uncommon. These tumours may present with slowly progressing sensory or motor syndrome. We report a case of 50 years female patient presenting with Intramedullary spinal schwannoma at D11 to L1 levels.

Keywords: Schwannoma, Benign, Spinal, Intramedullary.

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INTRODUCTION

Spinal Schwannomas constitute 30% of spinal cord tumor which are usually intradural and extramedullary in location. Intraparenchymal schwannomas occurring in the central nervous system without neurofibromatosis is extremely rare. They can occur in various parts of central nervous system like cerebellum, brainstem and spinal cord [1]. Intramedullary schwannoma represents 0.3% of all medullary tumors. These tumors are slow growing benign spinal nerve sheath tumors which are diagnosed by imaging studies or by progressive neurological deficit [2]. These tumors mostly present in 4th to 6th decades in life. Presenting as acute emergency is very rare [3].

Case Report

50 years female patient came to the neurosurgery department with the chief complaints of paraparesis of lower limbs. Patient was known hypertensive and diabetic. On examination her blood

pressure was 140/70mm/Hg, Pulse-76/min. Haematological investigations showed Hb-12.7g%, total WBC count-8900/ μ L, Platelet count:4,36000/ μ L. Biochemical investigations showed fasting blood sugar level - 250mg/dl and random sugar level - 437mg/dl. Serum creatinine was 1.4mg/dl, blood urea was 68.9mg/dl. Serum electrolytes were sodium-138mmol/l, potassium-4.5mEq/l and chloride-99mEq/L.

Plain Magnetic resonance imaging study of the lumbosacral spine showed Grade-I anterolisthesis of L4 over L5 with pseudo disc bulge causing neural foramina narrowing and compression over bilateral traversing nerve roots. L5-S1 diffuse disc bulge causing neural foramina narrowing and compression over bilateral traversing nerve roots was noted. Mild dense bulge was seen at L3-L4 causing mild neural foramina narrowing. Well defined mixed heterogenous signal intensity intramedullary lesion measuring 5.7x2.1cms involving the dorsal spinal cord at D11 to L1 levels was noted (Figure-1).



Fig-1: MRI showing well defined mixed heterogenous signal intensity intramedullary spinal lesion

Clinical diagnosis of paraganglioma or ependymomas was made. D11 to L1 laminectomy was done and tumor was excised in piece meal which was sent for histopathological examination. Microscopic examination revealed lesion composed of spindle

shaped cells arranged in fascicles and whorls. Few foci showed palisading of cell nuclei (Antoni A areas) with intervening eosinophilic zones (Verocay bodies) (Figure-2).

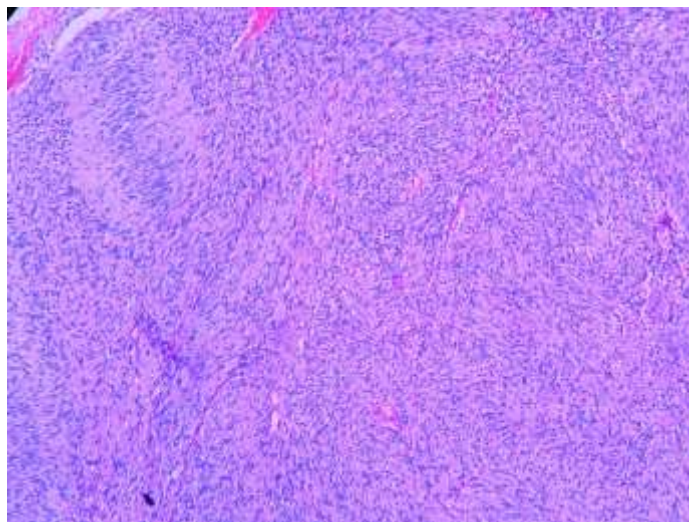


Fig-2: Spindle shaped tumor cells showing nuclear palisading (Antoni A areas) (H&E, X100)

Mild anisokaryosis was noted in some foci. Hypocellular areas were seen. Immunohistochemistry

was performed and tumor cells were positive for S-100 (Figure-3).

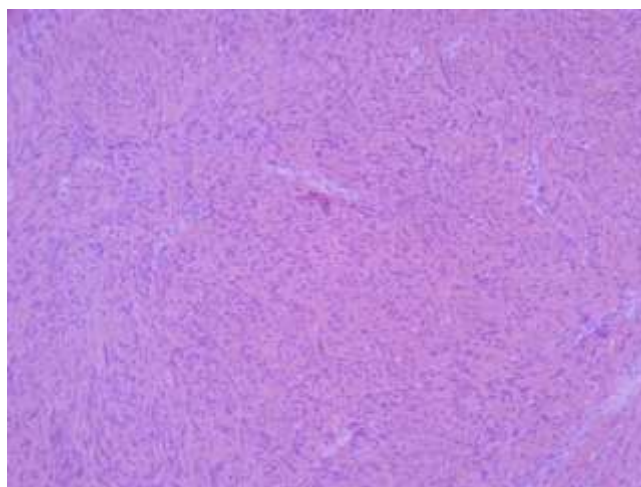


Fig-3: Tumor cells showing S-100 positivity (S-100, X100)

DISCUSSION

Sir Victory Horsley first gave the surgical description of spinal tumor in 1888. He reported an intradural extramedullary meningioma. Though Penfield described an intramedullary schwannoma in 1932, Kermohan was recognised as first neurosurgeon who reported on intramedullary schwannoma in 1952.

In the literature, 50 cases of intramedullary schwannoma without having association with neurofibromatosis have been reported. Among spinal schwannomas, intramedullary schwannomas constitutes 1.1% of cases and 0.3% of intraspinal neoplasms [4].

These tumors show male predominance with male to female ratio of 3:1. Age of occurrence of intramedullary schwannomas is 4th to 6th decade. These tumors are usually solitary affecting various regions of spinal cord with increased frequency in cervical spinal cord (63%). Other sites in spinal cord with decreasing frequency are thoracic spinal cord (26%) and lumbar spinal cord (11%). As these tumors are very slow growing, the average interval between appearance of first symptom and diagnosis is from 6 months to 20 years [5].

In the last 50 years several theories has been proposed regarding the origin and pathogenesis of intramedullary schwannomas. Origin of intramedullary

schwannoma was thought to be derived from schwannoma cells of the nerve fibres of spinal arteries. Ramamurthi *et al.*, proposed that intramedullary schwannoma cells arise from ectopic Schwann cells of embryonal neural tube. Mac Cormick and Wood in 1964, proposed that schwann cells in aberrant intramedullary nervous fibres are the origin for the development of intramedullary spinal schwannoma. However most accepted theory was proposed in 1971 by Russell and Rubenstein. According to their hypothesis, spinal schwannomas originate from the transformation of pial neuroectodermal cells into schwannoma cells which then undergo neoplastic change [6].

The most common clinical presentation is the pyramidal syndrome, sphincter dysfunction and sensitivity complaints. Few cases present with motor sensitive altarnal deficit with amiotrophy.

Imaging of these tumors on T1 weighted sequences are hypointense or isotense and on T2-weighted sequences are hyperintense. Heterogeneous enhancement is noted. Grossly schwannomas are soft in consistency with yellow brown discolouration. They may be lobulated and can have cystic change. Histologically these tumors are composed of two types of growth patterns termed as Antoni A and Antoni B. Antoni A areas are hypercellular showing spindle shaped cells in the collagenous background. Distinctive nuclear palisading in which parallel rows of nuclei are arranged with intervening eosinophilic zone termed as verocay bodies. Antoni B areas are loose and often have mucinous or myxoid background with stellate cells. Immunohistochemically tumor cells are positive for S-100 protein.

Schwannomas should be differentiated from meningiomas as meningiomas may show focal nuclear palisading. Immunohistochemistry with S-100 is helpful for differentiation.

Surgical resection is the treatment of choice. Intramedullary schwannomas may have infiltrative pattern which makes total tumor resection impossible.

Radiotherapy is suggested by some authors in such residual lesions [6].

CONCLUSION

Intramedullary schwannomas are slow growing rare tumors with benign course and should be considered as one of the possibility for the intramedullary spinal lesions in middle aged individuals. MRI may be helpful for diagnosis however clear distinction cannot be made between the intramedullary spinal lesions. Total surgical resection is treatment of choice but infiltrative lesions cannot be resected completely for which radiotherapy has been suggested.

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