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Extradural Sacral Spinal Concurrent Meningioma and Ganglioneuroma Arising from a Nerve Root: Rare Case Report

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Abstract: ***Concurrent occurrence of tumors of different pathological types are exceptionally rare at the same spinal level except for in genetic disorders, such as neurofibromatosis (NF) and von Hippel - Lindau disease (vHL). Concurrent occurrence of meningioma and Ganglioneuroma at the same spinal level has not been reported so far. Such a concurrent tumor arising from the sacral nerve root is exceptional. We report one such exceptionally rare concurrent lymphocyte rich meningioma and ganglioneuroma arising from S2 nerve root in a 38 year old female without any evidence of NF/vHL who presented with lower limb radiculopathy.***

Keywords: Coexisting tumor, Concurrent tumor, Meningioma, Ganglioneuroma, Multiple spinal tumors

1. Introduction

Multiple intracranial tumors are commonly reported; But occurrence of multiple intraspinal tumors are rare. Concurrent occurrence of tumors of different pathological types are exceptionally rare at the same spinal level. Only 9 cases have been reported to date, with meningioma found with schwannoma in 7 cases and with neurofibroma in 2 cases. [1 - 9]

Concurrent occurence of meningioma and Ganglioneuroma at the same spinal level has not been reported so far. We report one such rare occurrence in 38 year old female who presented with lower limb radiculopathy. This case is exceptionally unique becauseit is a spinal meningioma with the absence of the dural attachment, its origin from S2 nerve root and the lymphocyte rich variant histologically with a concurrent Ganglioneuroma in the same tumor.

2. Case report

A 38 year old female presented with low backache and pain in left leg. On examination, the sensation, power and reflexes of the left leg were nomal and there was tenderness at the level of L5, S1, S2 and S3. On CECT of sacro - iliac joint an expansile osteolytic lesion was noted in left ala of sacrum and S1/S2 vertebrae. On MRI of Lumbo - sacral spine a well - defined multi lobulated, mixed solid/cystic mass lesion was seen involving the S1 - S3 vertebral bodies and left ala of sacrum measuring 2.2 x 6 x 4 cm. (Figure 1) Enucleation of the tumor was done after expanding the sacrum. The lesion was arising from S2 nerve root and was seen entrapped with nerves producing mass effect on the sacral bone.

Grossly, a well circumscribed already cut open soft tissue mass (4 x 3 x 1.2 cm) was received. Outer surface of the mass was gray white, dull congested and cut surface revealed multicystic mass filled with friable material. (Figure 2) On microscopic examination, tumor was composed of dual component. The first component revealed meningioma comprising of sheets of round to oval to elongated/ spindled meningothelial cells with frequent nuclear grooves and intranuclear inclusions. A dense lympho - plasmacytic infiltrate was seen throughout the tumor. (Figure 3) Focal whorling, a single psammoma body and focal areas of necrosis were also noted. The second component was Ganglioneuromas comprising of sheets and fascicles of schwann cells in a myxoid background with scattered ganglion cells. (Figure 4) IHC revealed EMA, vimentin and PR positivity in meningioma component and S - 100, chromagranin A, synaptophysin positivity in Ganglioneuromas componenet. The tumor was diagnosed as Concurrent Meningioma (lymphocyte rich variant) WHO Grade I and ganglioneuroma arising from S2 nerve.

3. Discussion

Spinal meningiomas are mostly intradural in nature. Occasionally they may show some extradural extensions. Purely extradural spinal meningiomas (EDSMs) are extremely rare (2.5 - 3.5%). [10] Such extradural meningiomas are difficult to diagnose preoperatively. Less than ten cases of pure EDSM have been reported till date. All these reported cases were found in the thoracic and cervical spine. [11]

Extradural meningiomas are relatively rare and those arising from spinal root are exceptionally rare [12] and only a handful of cases have been reported so far. Jang WY [12] reported a case of extradural spinal meningioma (transitional variant) arising from the fourth thoracic spinal nerve root. Ng THK et al have described extradural meningioma arising from lumbar (L5) nerve root [13] in a young man.

Cho KB et al described an unusual case of the spinal meningioma in the young woman with the absence of the dural attachment and the adherence to the fascicles of the sacral nerve root with the clear cell meningioma histologically. [14] The present case was similar to this case, but showed lymphocyte rich variant of meningioma and a concurrent Ganglioneuroma in the same tumor.

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Lymphoplasmacytic - rich meningioma (LPRM) is a very rare benign subtype. Occurs mostly in cerebral convexities, skull base, parasagittal tentorial area and cervical spinal canal. In 2019, Cardoso L et al reported first case of a patient with lumbar spine Lymphoplasmacytic - rich meningioma. [15] However such a tumor arising from spinal nerve root has not been reported so far. So our case is the first case of extradural Lymphocyte rich meningioma arising from S2 nerve root.

Concurrent occurrence of tumors of different pathological types are exceptionally rare at the same spinal level except for in genetic disorders, such as neurofibromatosis (NF) and von Hippel - Lindau disease (vHL). Only 9 cases have been reported to date, with meningioma found with schwannoma in 7 cases and with neurofibroma in 2 cases. [1 - 9]

Concurrent occurence of meningioma and Ganglioneuroma at the same spinal level, that too arising from sacral nerve root has not been reported so far. Our patient had no evidence of neurofibromatosis or von Hippel - Lindau disease. All these reported concurrent spinal tumors were distributed in the cervical spine and seven out of the eight lesions were found above the C3 level [1 - 6] Extradural components consisted of seven schwannomas and single neurofibroma. Notably, all intradural tumors were meningiomas. Our case was not only located at the S1 - S3 level, but also both components of the tumor (meningioma and Ganglioneuroma) were extradural in location.

The pathophysiology of different tumors concurrently arising in the same spinal level has remained unclear. Three possible theories have been proposed to explain concurrenceof different tumors in the same spinal level. [1 - 6] The first theory is that de novo tumor might be induced by microenvironmental factors influenced by the preexisting tumor [2, 4, 6]Further evaluation of the expression of tumorigenesis - related cytokines might play a role in elucidating the pathophysiology of concurrent tumors. The difference of the volume between two components may be partially explained by the first theory. According to the literature, all intradural meningiomas were significantly smaller than extradural schwannoma/neurofibromas. [1–6]This fact seems to support the hypothesis that preexisting large schwannoma/neurofibroma induces small de novo meningioma. The second theory is explained from the embryological perspective. Common mesenchymal progenitor cells may separately differentiate into schwannoma cells and meningioma cells in the same spinal level [2–5] The third hypothesis is that this rare entity develops only due to incidence. [2 - 4] Schwannomas and meningiomas are two of the most common tumors developing in the spinal cord, comprising 55% of all extramedullary tumors. [16] They may accidentally arise at the same spine level in very rare occasions [2, 4, 6]

In conclusion, a case of concurrent lymphocyte rich meningioma and Ganglioneuromas arising from S2 nerve root without any evidence of NF/vHL is treated with gross total excision.6 months postoperatively, no recurrence has been reported.

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Legends

Figure 1: MRI showing a well - defined multi lobulated, mixed solid/cystic mass lesion was seen involving the S1 - S3 vertebral bodies and left ala of sacrum
Figure 2: multicystic mass filled with friable material.
Figure 3: Photomicrograph showing lymphocyte rich meningioma (H&E, 20x)
Figure 4: Photomicrograph showing ganglioneuroma component (H&E, 40x)

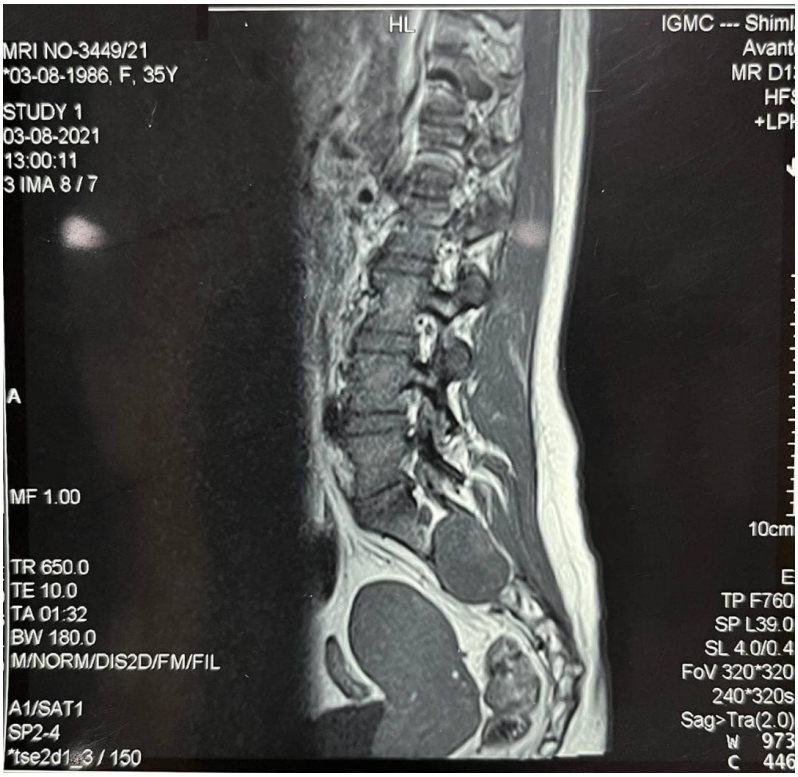

Figure 1

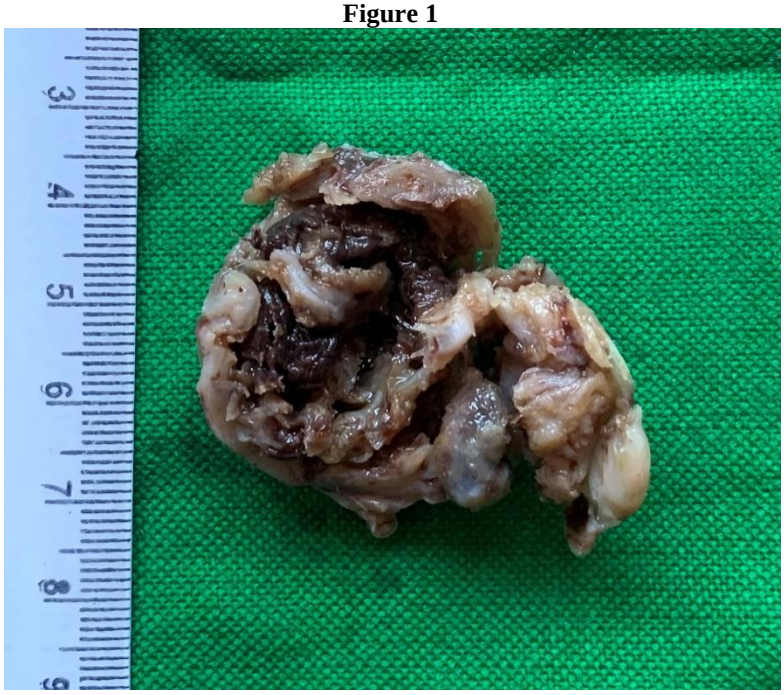

Figure 2

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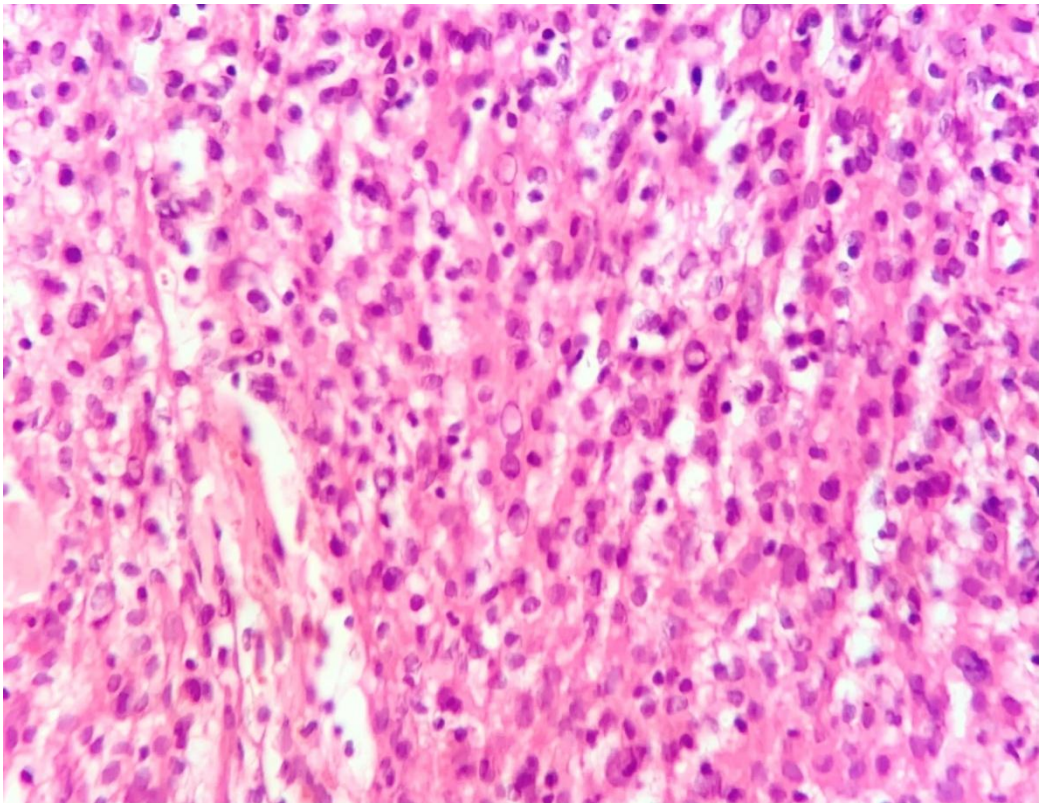

Figure 3

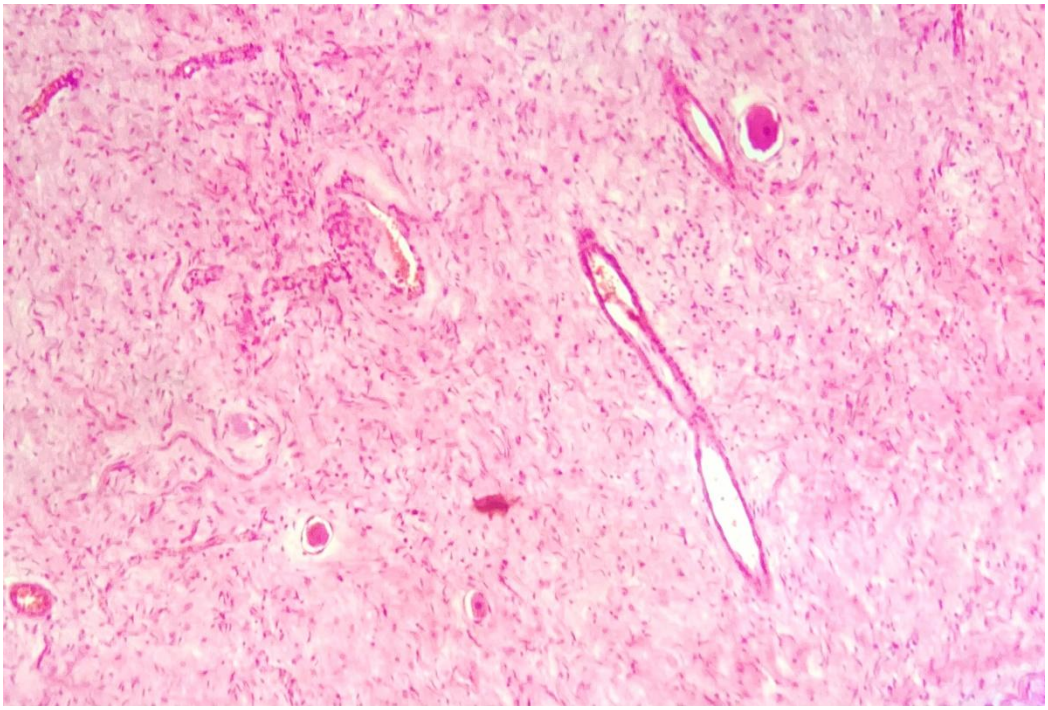

Figure 4

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