CASE REPORT

Rare Concurrence of Two Different Intraspinal Tumors at an Unusual Site: Ependymoma and Schwannoma

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Abstract

Ependymoma has been typically described as an intramedullary tumor which is derived from ependymal cells. Intradural extramedullary spinal ependymomas are extremely rare. Hereby, we present a case report of a patient was diagnosed with an intradural extramedullary spinal tumor extending from D8-D9 level. Patient underwent gross total resection of the tumor.The tumour was diagnosed as a collision tumor, ie Ependymoma with Schwannoma like areas on histopathological examination, later confirmed by immunohistochemistry.

Keywords

Spinal Tumors, Intradural, Extramedullary,Ependymoma,Schwannoma

Introduction

Ependymomas arising outside the lesion of the conus medullaris, cauda equina, and terminal filum or developing from ectopic ependymal cells are highly unusual. Reported cranial to spinal tumor ratio for ependymomas is 4:1. Intradural extramedullary ependymomas are extremely rare and predominate in women in their fifth decade of life. [1] Spinal schwannomas account for about 25% of intradural spinal cord tumors in adults. Incidence of spinal schwannomas varies from 0.3-0.4 cases/100,000 persons per year tumors.[2] Most cases of two histologically different tumors have been reported in the head and rarely in spine. The presence of multiple histologically different spinal tumors in the absence of Neurofibromatosis -2(NF-2) is extremely rare.

Case reoprt

A 32-year-old male presented with pain in the lower back followed by numbness in left leg since 20 days. His clinical and neurological examination was within normal limits with no signs of neurofibromatosis.

On contrast-enhanced MRI spine, a well-defined oval shaped altered signal intensity mass measuring 1.1x1.4x2.7cm was noted in an intradural extramedullary location extending from D8-D9 intervertebral disc level. Abnormal T2WI hyperintensity was suggestive of myelopathy with disc degeneration and posterocentral annular tear from L5-S1.

On blood examination, elevated liver enzymes and blood sugar along with neutrophilia were seen. D8-9 Laminectomy and excision was performed and sent for histopathological examination.

On gross examination, multiple grey white tissue bits altogether measuring 2x1.8x0.7cm were received. Histopathology showed cells arranged in ependymal

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Manuscript Received: 30.08.2022; **Revision Accepted**: 1.11.2022;

Published Online First: 10 July, 2023

Open Access at: https://journal.jkscience.org

Cite this article as: Rana S, Wavir NM, Mohroo R, Khetrapal S, Jetley S, Jairajpuri ZS.. Rare Concurrence of Two Different Intraspinal Tumors at an Unusual Site: Ependymoma and Schwannoma. JK Science 2023;25(3):180-82.

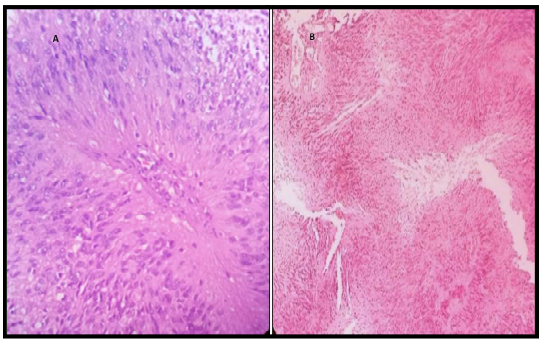

Fig 1 A: H & E section showing Perivascular Pseudorosette with elongated filamentous processes at 40X, B: H&E section showing both Schwannoma like areas with highly cellular Anotni A areas and Pseudorosettes at 10X

tubules and perivascular pseudorosettes.(*Fig 1A*) Lining cells were polygonal with moderate eosinophilic cytoplasm and vesicular nuclei. Elongated filamentous processes were seen extending to the vessel wall, which were thickened and hyalinised. Background showed extensive myxoid and fibrillary areas interspersed with highly cellular Antoni A areas.(*Fig 1B*)

A diagnosis of a tumor of neural origin with a possibility of ependymoma with schwannoma like areas was considered. Immunohistochemistry with GFAP and S100 were done for confirmation. (*Fig 2 & 3*)

The patient's neurologic condition improved after surgery and his stay was uneventful. Neither radiotherapy nor chemotherapy was required. Follow-up of the patient was done by MRI after two months which showed no mass lesion.

Discussion:

Ependymomas are the most common intramedullary tumours which occur predominantly in adults with reported cranial:spinal tumour ratio being 4:1 (range from 3:1 to 20:1) depending on histological subtype. [3]

Ependymomas arising outside the region of the conus medullaris, cauda equina, filum terminale or developing from ectopic ependymal cells are highly unusual. Intradural extramedullary spinal ependymomas are extremely rare, predominantly seen in women in their fifth decade of life. An extensive literature survey yielded

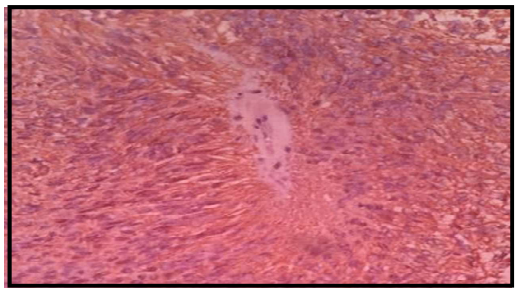

Fig 2, GFAP positivity predominantly around the pseudoresette at 40X.

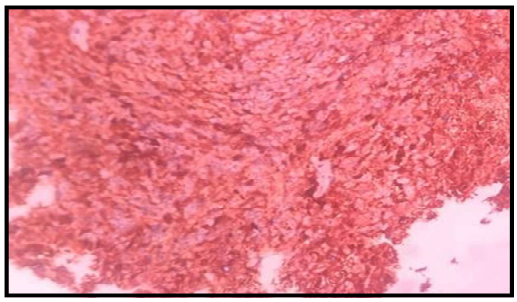

Fig 3. S-100 showing strong and diffuse positivity in Schwannoma like areas at 40X.

18 such cases.[3] This lesion is believed to arise from ectopic ependymal cell rests. Some of the ependymal cells may remain on the inside during neural tube closure. [4]

Although ependymomas are most often benign [5] there have been three previous reports of malignant transformation occurring in these lesions.[6,7] Some of these cells may undergo a malignant change, infiltrating the spinal pia mater, and the posterior spinal cord and may often lead to recurrence. Tumor recurrence does not occur after the first operation if a part of the adhesion lesion does not contain ependymoma tumor cells, or if those cells have not undergone malignant change. Few ependymoma cells remaining after the first operation might undergo anaplastic change. [8]

The presence of two histologically distinct spinal tumors in the present case suggested the possibility that this patient might have NF-2. However, neither the clinical findings nor MRI of the head provided a clue suggestive of the presence of NF-2 in our patient. Since 1975, there have been 11 case reports on 2 histologically different

spinal tumors in the same patient without evidence of NF. In clinical practice,it is possible to encounter the coexistence of two histopathologically distinct and rare tumors of the spinal cord in the same patient without NF-2. Concomitant presence of schwannoma and ependymoma, which are derived from different cell types, is extremely rare in the absence of NF-2, as in our patient. The possible explanation is mainly dependent on the hypothesis provided by Heuschling et al that the ependymoma may have developed in response to the microtrauma and irritation produced by the schwannoma.[9]

Schwannoma is the most common nerve sheath tumor. The incidence of spinal schwannoma is 0.3-0.5cases/ 100,000 individuals annually.Its prevalence is similar in males and females, and it is usually diagnosed during the fourth and fifth decades of life . Schwannomas commonly occur in the lumbar and cervical regions and originate from Schwann cell progenitors. [2]

Schwannomas are benign tumors that are typically round, well demarcated, and encapsulated. Multiple schwannomas in a patient are referred to as schwannomatosis, usually indicative of an underlying tumor predisposition syndrome, such as neurofibromatosis. [10] The co-existence of histologically different spinal tumors in the absence of NF-2 is extremely rare with most cases reported in the head region, rarely in spine.

Although the conventional hematoxylin and eosin (H and E) staining is vital for the histological diagnosis of such lesions, the immunohistochemistry (IHC) has become an integral part for the diagnosis in cases of neuropathology. Ependymomas tend to have diffuse positive staining for GFAP and vimentin with variable S-100 staining, whereas schwannomas commonly have strong, diffuse S-100 staining, and are typically negative for glial fibrillary acidic protein (GFAP). Our case revealed strong immunoreactivity for GFAP (*Fig 2*) with intermittent S-100 positivity. Some areas that were negative for GFAP, showed diffuse positivity for S-100. (*Fig 3*) Thus, confirming that the tumor was a collision tumor of ependymoma with schwannoma like areas. The best treatment is gross total resection, as was carried out in this case; adjunctive radiotherapy is necessary in malignant change cases.

Conclusion

The occurrence of two or more histologically different spinal cord lesions in the same patient without evidence of NF is extremely rare. Our case is an atypical and rare presentation in terms of clinical onset, lesion location and sex distribution. The best treatment for an extramedullary ependymoma is gross total resection, as was carried out in our case.

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