### **Case Report**

DOI: http://dx.doi.org/10.18203/2349-2902.isj20181148

# Primary intradural neuroendocrine tumour of spine: a rare pathology

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Received: 31 January 2018 Accepted: 06 March 2018

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#### **ABSTRACT**

Neuroendocrine tumour in central nervous system is a very rare pathology. We hereby report a primary intradural neuroendocrine tumour, second to the only other case reported in literature till now as per the best of our knowledge. The tumour was completely intradural, spanning across five segments in the lumbar spine and no primary tumour could be detected elsewhere in body. The patient underwent surgery followed by radiotherapy and showed symptomatic relief. This case report focusses on compiling the available literature on spinal carcinoids (both primary and metastatic) and discussing the relevant investigations and available treatment options for the same.

Keywords: Central nervous system, Intradural, Neuroendocrine tumour, Primary spinal carcinoid

#### **INTRODUCTION**

Neuroendocrine tumours (Carcinoid) are a group of tumours arising from neuroendocrine cells. These tumours commonly involve the gastrointestinal and the respiratory tracts. Uterus, ovaries, urinary bladder, salivary glands and testes are counted among the less common sites of involvement.<sup>1</sup>

Central nervous system (CNS) involvement is uncommon and an intradural spinal metastasis is extremely rare. The case discussed here didn't have any history of malignancy and upon investigations no primary tumour was elucidated. To date, only one other case of primary neuroendocrine tumour of the spine has been described in the literature.

#### **CASE REPORT**

A 53-year-old female presented to outpatient clinic with progressive weakness of left foot and radicular pain in the

left lower limb for past 1 year. The patient also had a history of lower back pain for the past 5 years which aggravated over the last 1 year. No prior history of any malignancy or tuberculosis was there. General physical examination was normal. Upon motor examination, there was overall left ankle weakness. The dorsiflexion power was scored 1/5 and the extensor halluces longus power was scored 0/5 according to Medical Research Council (MRC) scoring. Left ankle jerk was absent.

Sensory and motor functions on the right lower limb were normal. Magnetic resonance imaging of the lumbar spine showed a long intradural lesion extending from T12 to L5 which was isointense on T1 (Figure 1 A) and hyper intense on T2 weighted image and enhanced brilliantly on contrast image (Figure 1 B). Bony involvement of the spine was not noted. A provisional diagnosis of myxopapillaryependymoma was made. The patient was taken up for surgery and the tumour was found to be completely intradural, extending from the conus downward to L5 region.

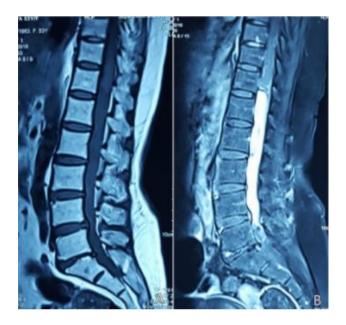


Figure 1: T1w sagittal MRI image (A) of lumbar spine showing isodense lesion extending from T12 to L5 with intense contrast enhancement (B).

The tumour was highly vascular and at places a definitive plane of demarcation from the surrounding nerve roots could not be defined, thus a subtotal resection was carried out

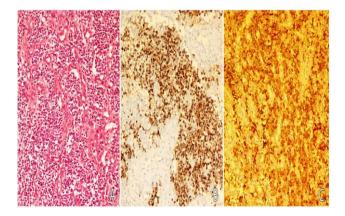


Figure 1: (A) Photomicrograph showing a cellular tumour arranged in nests and trabeculae, separated by thin fibrovascular channels (HE, x200), (B) The tumour cells show strong cytokeratin expression (immunohistochemistry, x200), (C) Diffuse CD56 expression by the tumour cells (immunohistochemistry, x200).

Biopsy of the excised tumour showed a moderately cellular tumour and tumour cells were arranged in nests, islands and ribbons, separated by delicate vascular channels (Figure 2 A). The cells have round to oval nuclei, coarse chromatin, inconspicuous nucleoli and moderate cytoplasm. Prominent vascular proliferation was noted. Mitotic activity was focally increased (3-4/10 high power fields). The tumour stained positive for Pan Cytokeratin (Figure 2 B), CD 56 (Figure 2 C) and faintly

positive for synaptophysin. CD 34 showed positivity in the proliferating vessels. However, it was negative for CD 45, GFAP, CK7, TTF1, EMA, GATA3 and CK20. Ki-67 labeling index was 5-6%.

Postoperatively, the patient improved symptomatically. Her radicular pain subsided but there was no improvement in her left lower limb weakness. Contrast enhanced CT scan of chest abdomen and neck didn't show any primary lesion in thymus, gastrointestinal and respiratory tracts.

Even a whole-body Gallium-68 octreotide (DOTA-NOC) PET scan was carried out 2 months postoperatively which did not reveal any region of excessive uptake anywhere in the body. Only a slight elevation in the biomarker chromogranin A levels was observed, suggestive of some residual disease. So, the patient was subjected to postoperative radiotherapy.

#### **DISCUSSION**

Neuroendocrine tumours are relatively indolent tumours but uncommonly they may show aggressive course and present with metastatic disease.

Metastatic disease has been noted at presentation in 22% cases of malignant neuroendocrine tumours. Even in patients of advanced stages of neuroendocrine metastasis, involvement of the CNS is very rare, occurring in <2% of the metastases. 1,2,3,4

The current case showed no previous history of primary neuroendocrine tumour and contrast enhanced computerized tomography chest, abdomen and neck didn't show any lesion. For further confirmation, an octreotide scan was done. This scan has been shown to have sensitivity for neuroendocrine tumours between 75% and 100%. In our case, the scan didn't show any accumulation along the gastrointestinal tract, the liver, spleen, pancreas, thymus or the genitourinary tract, ruling out any primary tumour site.

Such findings are suggestive that our case represents a primary intradural neuroendocrine tumour. Only one such case has been reported till date. Of all the reported cases of neuroendocrine tumours of the spine, both primary and metastatic, our case represents the largest and most extensive intradural neuroendocrine involvement (spanning over more than 5 vertebral segments). Surgical resection has been shown to be an effective treatment. Procedures for spinal decompression and resection of the intradural tumour have been shown to relieve associated pain and limb weakness seen with neuroendocrine tumours of the spinal cord, as in our patient. 1,2,6

Neuroendocrine tumours are highly vascular and frequently these tumours have a poor plane of demarcation from the nerve roots. Hence, mostly a

subtotal excision is feasible. Post-operative radiotherapy has been shown to be useful in both preventing reccurence of the tumour and reducing the extent of the residual disease. The role of chemotherapy in these tumours is not clear. A high index of clinical suspicion needs to be kept for neuroendocrine metastases to the spinal cord as it is a rare differential diagnosis for spinal

masses detected on CT or MRI. Octreoscan (DOTA-DOC) is a useful investigation with a very high sensitivity and specificity for neuroendocrine tumours. <sup>10</sup> Chromogranin A level is an important laboratory investigation that is shown to be useful, as it is the most common tumour marker for neuroendocrine tumours. <sup>11</sup>

Table 1: Showing primary and secondary neuroendocrine tumours of central nervous system described in literature.

Report	Age/sex	Origin	No. of segments involved	Surgery (extent of excision)	Postoperative radiotherapy	Follow-up
Intradural neuroendocrine metastasis						
Ironside et al <sup>12</sup>	44/F	Lumbar spine	3 (12-14)	-	-	-
Rao et al <sup>6</sup>	53/M	Thymus	2 (12-13)	Complete excision	Not given	Primary symptom relieved
Hopster et al <sup>7</sup>	75/F	Lumber teratoma	3 (13-14;t1)	Decompression	Not given	Functional recovery; dissemination to CNS
Blondet et al <sup>13</sup>	21/M	Lung	2 (13-14)	Complete excision	Not given	Symptomatic relief; died 2 years post-surgery
Nagel et al <sup>8</sup>	57/M	Thymus	2 (14-15)	Subtotal	Not given	Mild recovery; died 1 year post-surgery
Karatas et al <sup>14</sup>	55/M	Lung	1 (14)	Complete excision	Not given	Symptomatic relief post- surgery
Camarero C et al <sup>9</sup>	80/M	Pancreas	2 (c8-t1)	Surgical excision	Given	Died 2 months post- surgery
Primary intradural neuroendocrine tumour						
Audlin et al <sup>1</sup>	58/M	-	2 (11-12)	Subtotal	Given	Sensory symptoms improved, no change in motor strength, 1.5-year follow-up record
Current case	53/F	-	6 (t12-15)	Subtotal	Given	Symptomatic relief, mainly sensory; motor findings persist, 1-year post-surgery

#### **CONCLUSION**

To conclude, primary neuroendocrine tumour of the spine is very rare pathology. We have tried to compile the thorough literature available, so that our current knowledge about this entity can be enhanced.

Funding: No funding sources Conflict of interest: None declared

Ethical approval: The study was approved by the

Institutional Ethics Committee

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Cite this article as: Gupta VK, Latawa A, Kalra G, Radotra BD. Primary intradural neuroendocrine tumour of spine: a rare pathology. Int Surg J 2018;5:1567-70.