Primary Lumbar Spine Neuroendocrine Tumour: A Case Report

Puong You Law[a], Xin Ying Lim[b], Noor Afidah Binti Abdullah[c], Nelson Kok Bing Yap[a]

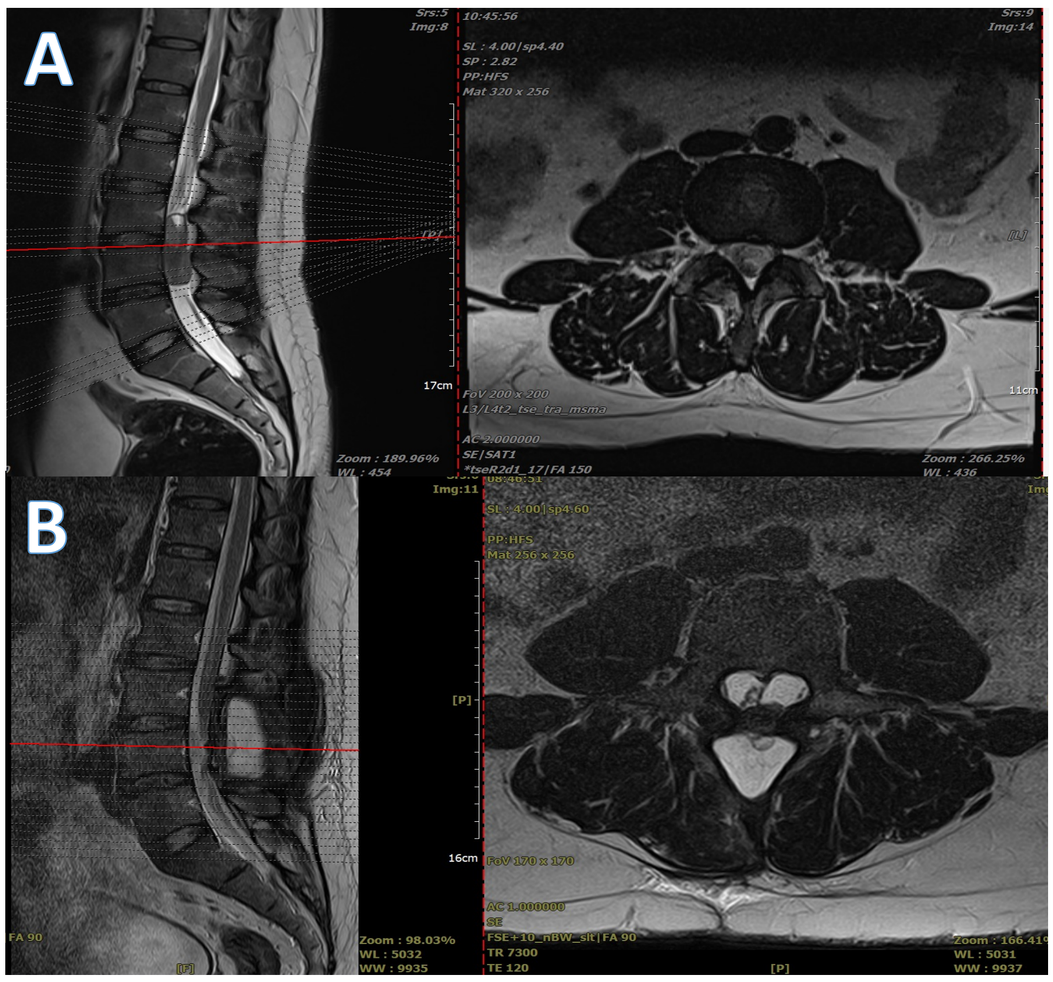
[a]Department of Neurosurgery, Sibu Hospital, Malaysia
[b]Department of Radiotherapy, Oncology and Palliative Care, Sarawak General Hospital, Malaysia
[c]Department of Pathology, Sarawak General Hospital, Malaysia

Background

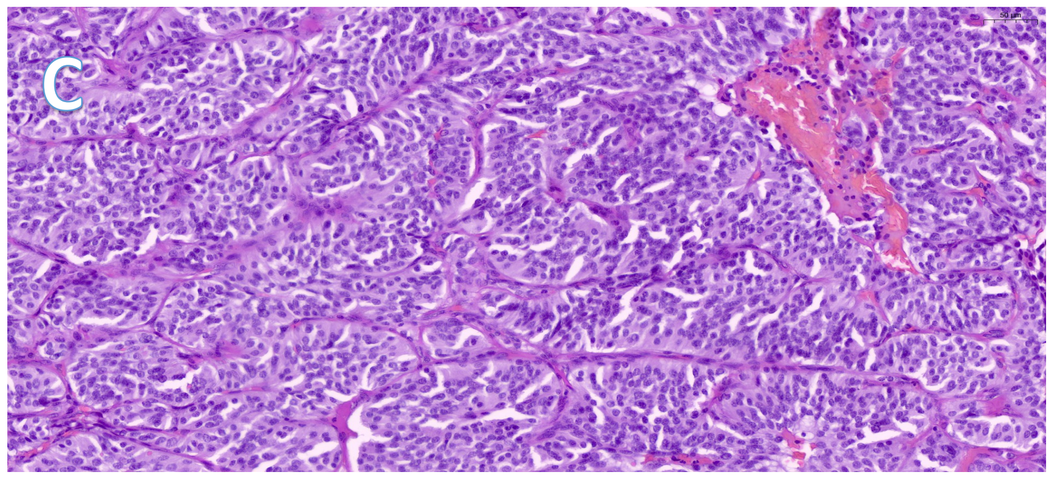
- **Neuroendocrine tumours** are **uncommon neoplasms** that often originate in the gastrointestinal or respiratory tract[1].
- They often metastasize to bone, liver, and pancreas.
- The central nervous system and notably the spinal cord are hardly involved.
- **Primary neuroendocrine tumours of the CNS are even rarer.**

Case Presentation

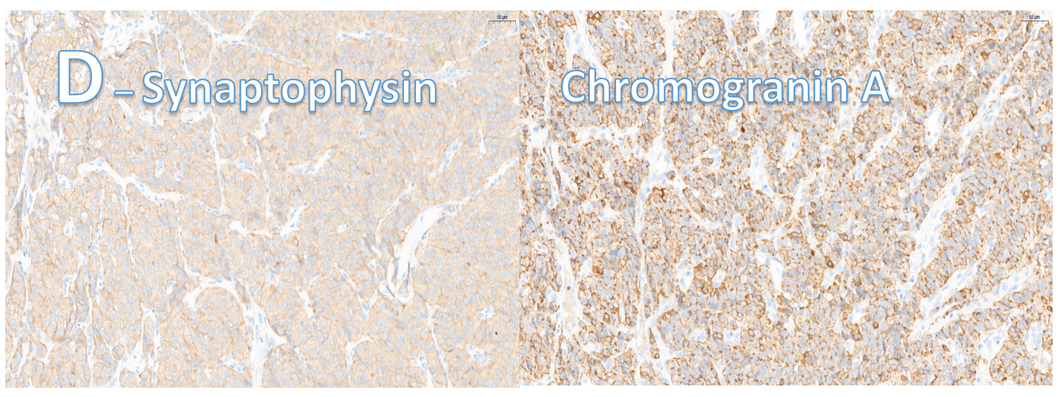
- A healthy 40-year-old Asian man presented with a **6-month** history of **progressive bilateral lower extremities paraesthesia, weakness**, and a negative history of cancer and trauma.
- A neurological examination revealed hypesthesia and mild weakness in his left L3 & L4 spinal nerve root distribution, as well as absent deep tendon jerk of the left ankle.
- Preoperative laboratory tests were normal.
- Magnetic resonance imaging (**MRI) (A)** of the spine showed an **intradural extramedullary (IDEM) tumour** at **the L3 and L4 spinal levels. (B)** showed the tumour was grossly removed via laminectomy and the presence of pseudomeningocele.



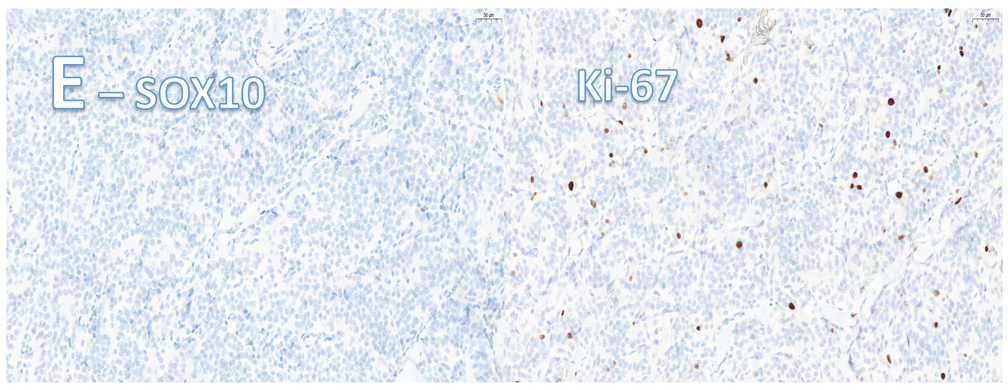
- The tumour was seen to have eroded through the dura and was pressing on the spinal nerves.
- Postoperatively, histopathological examination (**HPE**) and immunohistochemical (**IHC**) staining revealed the tumour to be a **low-grade neuroendocrine tumour.**
- **HPE (C) –** neoplastic cells arranged in nest and insular pattern. The cells had round nuclei and fine chromatin with eosinophilic cytoplasm.



- **IHC (D) –** Synaptophysin and Chromogranin A stains were positive



- **IHC (E) –** SOX10 was negative and Ki-67 proliferative index was low.



- Other body parts were screened and found to be normal.
- The patient's sensation and lower limb power improved.
- He was back to work 1 month after surgery.
- There was no recurrence at follow-up for 6 months. No chemo-radiotherapy was given.

Discussion

- Neuroendocrine tumours occur most frequently as **metastases** in the spine.
- Primary neuroendocrine tumour of the spine is a **very rare entity**.
- The differential diagnoses of IDEM of the spine includes **nerve sheath tumours, meningioma, and ganglioneuromas[2].**
- **Radiographically, it is hard to distinguish nerve sheath tumours from primary spine neuroendocrine tumours** when the lesion is well circumscribed and lobulated presenting as an IDEM.
- **Most commonly, they are diagnosed on histopathologic examination** that shows cells arranged in nest and island patterns with round nuclei and salt and pepper chromatin with eosinophilic cytoplasm[3].
- Tumour cells are **positive for synaptophysin and chromogranin.**
- Examination to rule out metastatic disease to the spine included a CECT chest and abdomen did not reveal any other lesion. However, the Dotatate scan is still pending.
- **The treatment of choice remains complete excision of the tumour** as in this case**,** allowing definitive healing of the primary neuroendocrine tumours of the spine[4].
- Radiation therapy is recommended for patients with metastatic neuroendocrine disease of the spine.

Conclusion

- **Neuroendocrine tumours of the central nervous system** are **extremely rare.**
- But it should remain in the **differential diagnosis** for patients experiencing extremity numbness and weakness and back pain with an **intradural extramedullary mass** and no other primary source of the tumour identified.
- Surgical resection may offer a definitive cure.

References

1. Audlin, J. R.; Khullar, G.; Deshaies, E. M.; Kurra, S.; Lavelle, W. F. Intradural Carcinoid Tumor Found in a Patient with No History of Cancer. *World Neurosurgery* **2016**, *96*, 607.e1-607.e6. https://doi.org/10.1016/j.wneu.2016.09.088.
2. Kim, Y.-M.; Lee, S.; Ryou, K.-S.; Kim, S.-H.; Park, T.-J. Lumbar Intradural Extramedullary Non-Metastatic Carcinoid Tumor: A Case Report. *Nerve* **2020**, *6* (2), 131–135. https://doi.org/10.21129/nerve.2020.6.2.131.
3. Zhang, X.; Zhang, Y.; Yan, X.; Bie, L. Primary Carcinoid Tumor of Medulla Spinalis: Case Report and Review of the Literature. *Eur J Med Res* **2014**, *19* (1), 71. https://doi.org/10.1186/s40001-014-0071-7.
4. Kumar, S.; Maheshwari, V.; Mukherjee, A.; Raman, D. Primary Neuroendocrine Tumor of the Lumbar Spine: Rare Tumor Mimicking Nerve Sheath Tumor of the Spine. *Asian J Neurosurg* **2019**, *14* (03), 894–896. https://doi.org/10.4103/ajns.AJNS_276_18.

Law Puong You
lawyou56@yahoo.com