

Paraganglioma in the Spinal Cord Histologically Mimicking Ependymoma – a Case Report

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Abstract

Paraganglioma is a tumour lesion of neuroectodermal origin that occurs at various places in the human body, but is rarely observed in the spinal cord. Usually, it presents in the lumbar region (cauda equine and filum terminal) as a slow-growing painless tumour mass that causes local compression.

We present a 53-year-old man with chronic low back pain, difficulty in walking, and impotence. The magnetic resonance study showed a tumour lesion suspicious for ependymoma. Surgical excision was performed and the result from the intraoperative frozen section was also ependymoma due to the presence of pseudorosettes structures. After the surgery, a thorough histological examination of the specimen revealed an encapsulated formation composed of ovoid and spindle-shaped cells with eosinophilic to light cytoplasm, with rounded nuclei with finely dispersed chromatin. An arrangement of cells in the form of pseudorosettes – ependymoma-like features was found in a large area of the tumour. Immunohistochemically, it was proved that the tumour was paraganglioma, not ependymoma.

Paraganglioma is a rare tumour in the spinal cord and it should be distinguished from other tumours normally observed in this area like ependymomas by using immunohistochemical stains.

Keywords

differential diagnosis, histology, immunohistochemistry, spinal cord tumours

INTRODUCTION

Paraganglioma is a tumour lesion of neuroendocrine origin. It arises from the neural crest cells in the segmental or lateral autonomic ganglia.^[1]

It occurs in a variety of places in the human body: in the head, neck, abdominal and thorax cavity, but is rarely observed in the spinal cord. Usually, it presents in the lumbar region (cauda equine and filum terminal) as a slow growing painless tumour mass causing local compression.^[2] The prognosis is very good after surgical excision, although

local recurrence has been observed in 4% of the cases; metastases are rarely observed.^[3]

Ependymomas are tumours that could be found also in the spinal cord. The differential diagnosis between these two entities is important because paragangliomas are generally considered to be WHO grade 1 while ependymomas are WHO grade 2.^[4] Histology and especially immunohistochemistry are crucial for the correct diagnosis.

CASE REPORT

We present a case of a 53-year-old man with chronic low back pain radiating to his right inguinal area and difficulty in walking. The pain worsened and became intolerable about 1 week prior to admission to the hospital. A spine MRI revealed a tumour in the thoracolumbar region at the level of Th12-L1. The patient was admitted for neurosurgical treatment. He denied any alcohol use and cigarettes. His disease history was unremarkable. On physical examination, his vital signs were normal. The neurological examination showed a vertebral syndrome in the thoracolumbar region. He had pain along the dermatome of the L1 nerve root on the right, paraplegia, and impotence (erectile dysfunction). The MR images demonstrated a tumour mass in thoracolumbar region of Th12-L1 suspected of ependymoma.

The patient underwent neurosurgical treatment under general anesthesia. He was placed in a prone position and the operative level was determined radiographically using a C-arm. A linear midline skin incision followed by paravertebral muscles dissection and retraction and L1-Th12 laminectomy were performed. After an incision in the dura mater, the tumour was exposed as a greyish-reddish mass involving several nerve roots and compressing the medullary cone. A piece of tissue was sent to the Pathology Department for frozen section analysis. Microsurgically, under optical magnification and ultrasound assistance, total extirpation of the tumour was achieved, with decompression of the spinal cord and the corresponding nerve roots. The dura was sutured in water-tight fashion and the wound was closed in several layers. The postoperative period was uneventful as the patient was without radicular pain and with improved gait. No additional neurological deficit was observed.

Pathology

The frozen sections and cytology made during the operation showed a picture that resembles ependymoma (**Fig. 1**).

After the whole tumour lesion was received in the Pathology Department, a thorough examination of the material was performed. Grossly, the specimen was oval and encapsulated (4×2.1×1.7 cm in size), on a cut section it was with variegated appearance whitish-brownish and with reddish areas. The consistency was moderately dense.

Histologically, there was an encapsulated formation composed of ovoid and spindle-shaped cells with eosinophilic to light cytoplasm, with rounded nuclei with finely dispersed chromatin. The tumour cells formed nests and alveolar structures (**Fig. 2**). An arrangement of cells in the form of pseudorosettes – ependymoma-like features was found in a large area of the tumour. There were areas with perivascular hyalinization in the tumour. The tumour stroma was well vascularized. Immunohistochemically, there was a diffuse positive expression of chromogranin A in the tumour cells, negative expression of glial fibrillary acidic protein (GFAP) and positive expression of S-100 protein in the sustentacular cells (**Fig. 3**).

DISCUSSION

Paraganglioma is a tumour lesion of neuroendocrine origin. It arises from the neural crest cells in the segmental or lateral autonomic ganglia.^[1]

It is rarely observed in the spinal cord and in most of the cases, spinal paraganglioma is found in the terminal filum; other localizations are the lumbar region, cauda equine, and conus medullaris.^[2]

Clinical symptoms could be variable the most common being low back pain and radiculopathy.^[5] In our case, the patient had also paraplegia and impotence. Sexual dysfunction was also reported in a few other studies.^[6,7]

On gross pathological examination, paragangliomas are encapsulated, oval masses with soft consistency and red brown cut surface.^[4] The histology usually reveals the presence of two types of cells – chief and sustentacular

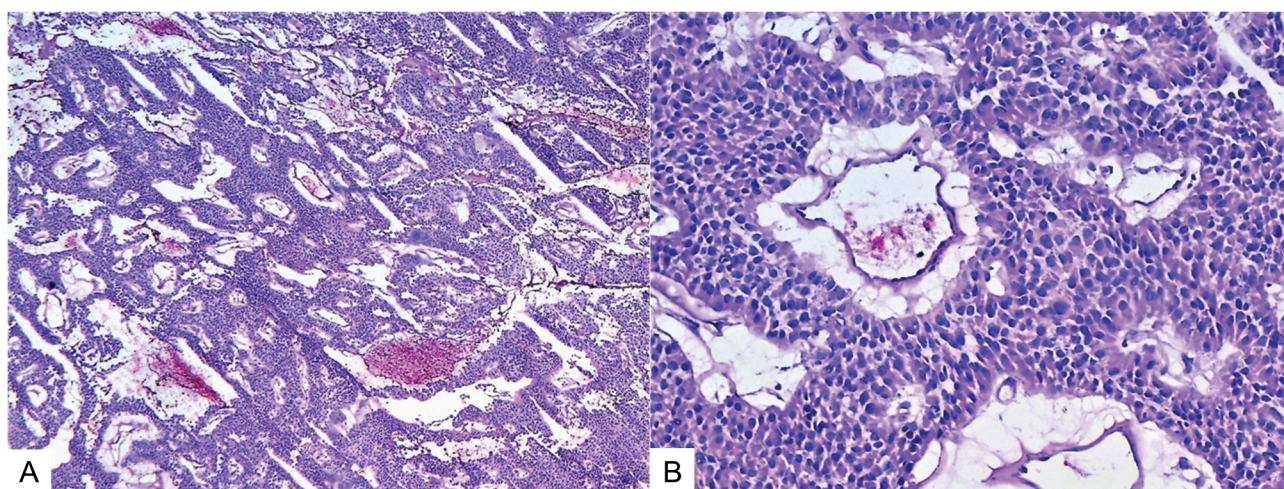


Figure 1. Intraoperative frozen sections of the tumour lesion showing ependymoma-like features – pseudo rosettes. **A.** Frozen section, original magnification ×40; **B.** Frozen section, original magnification ×200.

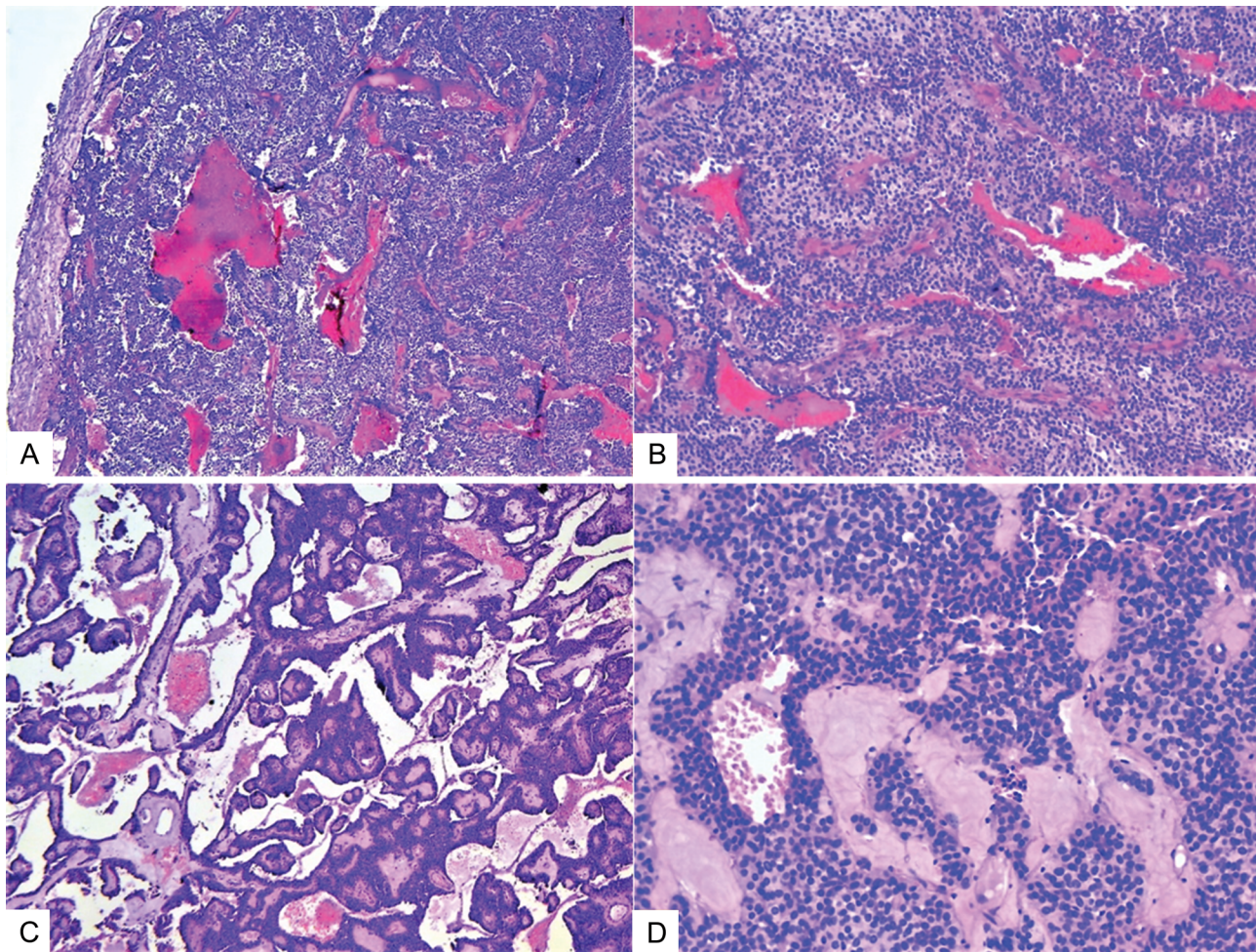


Figure 2. Histology of the tumour lesion. **A.** The lesion is capsulated and composed of tumour cells with well-developed vascular stroma; hematoxylin-eosin stain, original magnification $\times 40$; **B.** Tumour cells are located around blood vessels with ependymoma-like appearance; hematoxylin-eosin stain, original magnification $\times 100$; **C.** Hematoxylin-eosin stain, original magnification $\times 40$. **D.** Part of the tumour lesion with hyalinization; hematoxylin-eosin stain, original magnification $\times 200$.

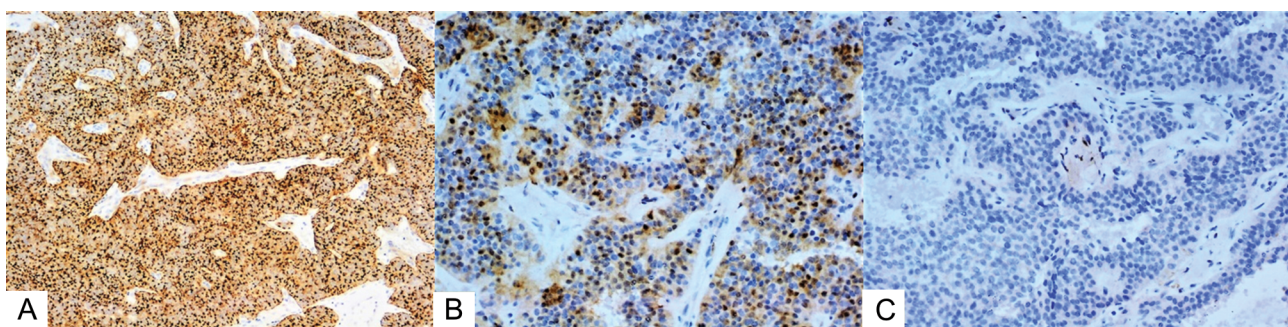


Figure 3. Immunohistochemistry. **A.** Positive expression of chromogranin A, original magnification $\times 100$; **B.** Positive expression of S-100 in sustentacular cells, original magnification $\times 200$; **C.** negative expression of GFAP, original magnification $\times 200$.

cells. The chief cells have oval nuclei with finely dispersed chromatin of the ‘salt and pepper’ type. The sustentacular cells are placed around the chief cells. The tumour parenchymal cells are arranged in nests or lobular structures (‘zellbalen’). The tumour stroma is composed of multiple thin-walled small blood vessels. Occasionally, the tu-

mour cells can form pseudorosettes (perivascular radial arrangement of the cells) which can mimic ependymomas.^[2] In our case, the tumour lesion also exhibited ependymoma-like features (**Fig. 2B**). Some authors also reported that paragangliomas can have ependymoma-like features – ependymal tubules.^[8,9]

Immunohistochemistry is essential in making the exact diagnosis. There is positive expression of S-100 in all paraganglioma tumours, especially in the sustentacular cells.^[2,9,10] There is also positive expression of chromogranin A and synaptophysin in the chief cells because they have neuroendocrine origin.^[11] It is important to note that the cells in paraganglioma are negative for cytokeratin which could be used to distinguish paragangliomas from other types of neuroendocrine tumours.^[12] Another antibody is crucial in the differential diagnosis between paraganglioma with ependymoma-like features and real ependymoma – GFAP. There is absence of the expression of GFAP in paragangliomas and positive expression in ependymomas.^[9,13,14]

As a clinical behaviour, paragangliomas are considered to be benign, well-demarcated from adjacent structures tumour lesions, usually located intradurally.^[2] The prognosis is very good after surgical excision, in 4% of cases there is a local recurrence and metastases are rarely observed.^[3] If full extraction of the tumour lesion is impossible, radiation therapy is recommended.^[15]

Ependymomas are tumours that are very common in the lumbar region and especially around cauda equine. Paragangliomas are generally considered to be WHO grade 1 while ependymomas are WHO grade 2.^[4] Hence, it is important to distinguish paraganglioma from ependymoma which is known to have a more aggressive behaviour.

CONCLUSIONS

Paragangliomas in the spinal cord must be distinguished from other tumours with similar localization such as ependymomas. The final diagnosis cannot be made intraoperatively. The use of immunohistochemistry is essential.

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Параганглиома в спинном мозге, гистологически имитирующая эпендимому – клинический случай

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Резюме

Параганглиома – опухолевое поражение нейроэктодермального происхождения, возникающее в различных местах тела человека, но редко наблюдающееся в спинном мозге. Обычно он проявляется в поясничной области (конский хвост и терминальная нить) в виде медленно растущей безболезненной опухолевой массы, вызывающей локальное сдавление.

Мы представляем случай 53-летнего мужчины с хронической болью в пояснице, трудностями при ходьбе и импотенцией. При магнитно-резонансном исследовании выявлено опухолевое поражение, с подозрением на эпендимому. Было выполнено хирургическое иссечение, и результатом интраоперационного замороженного среза также была эпендимома из-за наличия псевдорозеточных структур. После операции при тщательном гистологическом исследовании препарата выявлено инкапсулированное образование, состоящее из овоидных и веретенообразных клеток с эозинофильной или светлой цитоплазмой, с округлыми ядрами и мелкодисперсным хроматином. Расположение клеток в виде псевдорозеток – эпендимомоподобные признаки выявлены на большой площади опухоли. Иммуногистохимически было доказано, что опухоль является параганглиомой, а не эпендимомой. Параганглиома является редкой опухолью спинного мозга, и её следует отличать от других опухолей, обычно наблюдаемых в этой области, таких как эпендимомы, с помощью иммуногистохимического окрашивания.

Ключевые слова

дифференциальная диагностика, гистология, иммуногистохимия, опухоли спинного мозга