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Primary Phylum Terminale neuroendocrine tumour: An unusual case report

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ABSTRACT

Background: Cauda equina and filum terminale neuroendocrine tumours are rare neoplasms that develop in the cauda equina or filum terminale region of the spinal cord. Paragangliomas are grade 1 tumours (WHO grade I) that originate from neural crest cells. The diagnosis of paragangliomas is primarily based on histopathological findings, as there are usually no specific clinical or radiological indications. On radiological examination, they present as tumours similar to schwannomas and ependymomas.

Observation(s): A 59-year-old male patient presented at the neurosurgery clinic with a history of chronic low back pain. The patient underwent contrast-enhanced magnetic resonance imaging of the lumbar vertebrae. On the basis of the MRI imaging, a differential diagnosis of a typical filum terminale ependymoma or neuroma was considered. The patient underwent a decompressive laminectomy, a surgical procedure performed to relieve pressure on the spinal cord. The mass was excised en bloc and submitted for pathological examination. A total of 13 distinct immunohistochemical examinations were conducted for the purpose of differential diagnosis.

Discussion: The histopathological diagnosis of paragangliomas is challenging, with the primary differential diagnosis being ependymoma and metastatic paragangliomas to this region. A meticulous approach is essential when dealing with tumours exhibiting such morphology, as this allows for the exclusion of other tumours with analogous histopathological findings and the performance of a comprehensive panel to ascertain the nosological origin.

Conclusion: Surgical intervention represents the primary treatment option, and there is currently no definitive consensus on the use of chemoradiotherapy.

Keywords
paragangliomas, phylum terminale, neuroendocrine tumour, histopathological, spinal cord

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INTRODUCTION

Neuroendocrine tumour is a type of neoplasm that occurs in cells in the *endocrine system* and *nervous system.* There are many types and they have special secretory particles. They act like a group of tissues because they secrete biogenic amine and polypeptide hormones [1]. These are also referred to as paragangliomas (PGs). It is uncommon for there to be extra-adrenal PGs. Paragangliomas of the central nervous system (CNS) are rare, with the majority of cases occurring in the cauda equina region of the spinal cord [2].

Cauda equina and Phylum Terminale neuroendocrine tumours are rare neoplasms that develop in the cauda equina or filum terminale

region of the spinal cord. Previously, they were designated as paragangliomas in the World Health Organization (WHO) classification of central nervous system (CNS) tumours. The most common extraspinal localisations of PG in the CNS are the petrous ridge, pineal gland, and sella turcica [2-5]. Only a few hundred cases have been reported in the medical literature since it was first described in the 1970s [6]. However, it has been demonstrated that approximately 3% of spinal tumours fall into this group [7,8]. Primary neuroendocrine tumours are typically situated in the extradural region. Additionally, they may occur in intramedullary locations [9]. The clinical findings of PGs are typically nonspecific, manifesting as pressure on the spinal cord. The diagnosis of an PGs is primarily based on histopathological findings, as there are often no specific clinical or radiological indications.

The condition manifests in individuals across all age groups, with a notable prevalence observed in the fourth and sixth decades [6,10]. There is a higher incidence of the condition in males compared to females [5,6]. From a radiological perspective, it bears resemblance to tumours such as schwannoma and ependymoma [11].

It is characterised by well-defined borders and demonstrates gadolinium contrast enhancement. Variations in signal intensity may be observed.

Although paragliomas in the cauda equina have been previously described in the literature, our case represents a unique instance of such a tumour occurring in the filum terminale. We sought to present histopathological evidence to our readers.

Case Report

A 59-year-old male patient presented at the neurosurgery clinic with a chief complaint of low back pain that had persisted for an extended period. Additionally, he has recently begun experiencing urinary incontinence. The patient's physical examination revealed normal muscle strength in the lower extremities, absence of sensory deficit, and normal deep tendon reflexes. The Babinski sign was negative bilaterally. The patient's bowel functions were found to be normal. A contrast-enhanced magnetic resonance imaging of the lumbar vertebrae was performed on the patient. The lesion exhibited a nodular appearance, measuring approximately 10 mm in diameter, with well-defined margins. It demonstrated heterogeneous hyperintensity on T2-weighted (T2W) images and isointense signal on T1-weighted (T1W) images, which were comparable to those of the normal lumbar spinal cord. The lesion was observed to extend within the spinal canal at the level of the L3 vertebra superior (Fig. 1). Based on the MRI imaging, a differential diagnosis of a typical filum terminale ependymoma or neuroma was considered. There was no evidence of spinal canal or neural foramen expansion, cortical bone erosion, or scalloping of the posterior margins of the vertebral bodies.

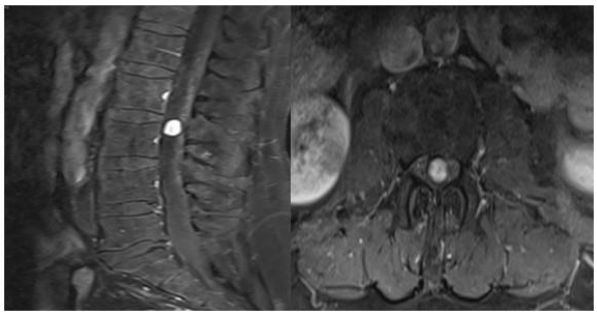

Figure 1. On magnetic resonance imaging, a hyperintense mass lesion with contrast enhancement in the spinal canal at the L3 vertebra level, on T1 sagittal and axial sections.

The patient underwent decompressive laminectomy spinal surgery. En bloc excision of the mass was performed and sent for pathological examination. Following the postoperative histopathological diagnosis, which was evaluated as paraganglioma, the patient was referred to the oncology clinic. No recurrence or residual mass was observed in the MRI examinations performed during the patient's first 6-month outpatient clinic follow-up.

Histopathological Examination

A tissue sample, measuring 1.5 x 1 x 0.5 cm in its largest diameter and of a beige-grey colour, was sent to the clinical pathology unit for examination. The immunohistochemical study was conducted in our laboratory using a fully automated Ventana Benchmark XT instrument (Arizona, USA). The tissues were fixed in buffered formaldehyde and an internal control for antibodies was employed in order to ensure the reliability of the results. The antigen was retrieved using an automated system. A total of 13 distinct immunohistochemical examinations were conducted for the purpose of differential diagnosis. Negative staining was observed for glial fibrillary acidic protein (GFAP), S100, SOX 10, vimentin, OLIG-

2, LCATTF-1 and epithelial membrane antigen (EMA). CK18 was diffusely positive, as were CD56 and synaptophysin and chromogranin, which showed diffuse staining (*Figure 2*). Evaluation of Ki67 indicated artefactual staining at 3–5%. Based on the histopathological examination of the specimen by a double-blinded pathologist with at least 10 years of experience, a diagnosis of neuroendocrine tumour, WHO grade I, was made. The surgical margins were clear.

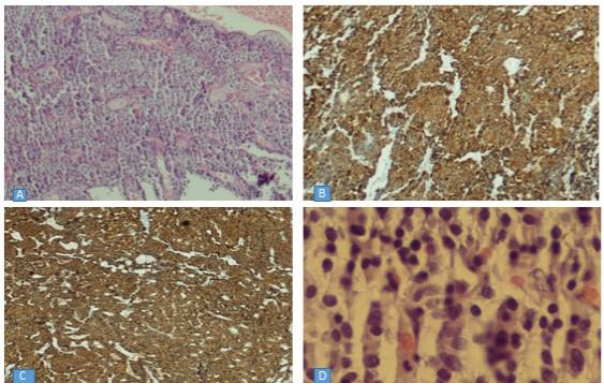

Figure 2. A; Structure consisting of slightly atypical, oval enlarged tumors with central hyperchromatic nuclei and salt-and-pepper chromatin characteristics, H&E, original magnification x100. **B;** Negative GFAP immunohistochemistry, original magnification x100 **C-D;** Intense cytoplasmic staining of tumor cells with the neuroendocrine markers chromogranin (C) and synaptophysin (D), original magnification x100.

Discussion

A phylum terminale neuroendocrine tumour, also referred to as a paraganglioma, is a rare tumour. It is a tumour of the nervous system that is frequently designated according to its location, with the cauda equina region being a particularly common site. It is a rare tumour and often difficult to diagnose. Patients affected by it are typically between 40 and 60 years old, although cases have been described for all age groups in recent years [6, 10]. It is more common in males. Our patient was a 59-year-old male.

Paragangliomas are grade 1 tumors (WHO Grade I) that develop from neural crest cells [11]. They may be secretory, but the vast majority are not. Secretory tumors produce catecholamines. They are often benign and grow slowly. The rate of malignant transformation has been reported to be 2.4% to 14% [12]. The prognosis is excellent if completely removed [2].

The classic histological features of PG consist of a "zellballen" or nesting pattern with trabeculae and cords of cells within a thin fibrovascular stroma. It has been demonstrated that almost 50% of cauda equina PGs contain mature ganglion cells, as well as transitional cells situated between the head and the aforementioned ganglion cells [2]. The most commonly employed method for differential diagnosis is immunohistochemical staining for chromogranin, nonspecific enolase (NSE), and synaptophysin, which are positive for PG. Conversely, glial fibrillary acid protein (GFAP) is negative in neoplastic cells. In the case we presented, negative staining was observed for GFAP, S100, SOX 10, vimentin, OLIG-2, LCATTF-1 and epithelial membrane antigen (EMA). CK18, CD56 and synaptophysin and chromogranin were diffusely positive.

In the context of radiological diagnosis, magnetic resonance imaging (MRI) is the imaging study of choice. MRI typically presents as a well-circumscribed, hyperintense, and sometimes partially cystic mass that is hypo- or isointense to the spinal cord on T1-weighted images and contrast-enhanced on T2-weighted images. In our case, the preliminary diagnosis based on MRI imaging was a filum terminale ependymoma or neuroma. There was no evidence of spinal canal or neural foramen widening, cortical bone erosion, or scalloping of the posterior margins of the vertebral bodies. The definitive diagnosis was made by histopathological examination, and the patient had no known oncological history.

The patient underwent a L2-L3 total decompression laminectomy, during which the tumour that was attached to the filum terminale was removed en bloc. The patient was under outpatient clinical control and a spinal and cranial MRI scan was performed in the first month after surgery. No primary lesions that may have been at rest or metastasised were detected. It is established that patients with CNS involvement may be treated with postoperative chemotherapy and radiotherapy. However, there is no specific consensus for spinal tumours [13]. No recurrent mass was found in the patient's 6-month follow-up.

Conclusion

PGs are uncommon and typically benign tumours. Diagnosing them based on radiological findings can

be challenging. The recommended treatment is complete surgical resection. The use of chemotherapy and radiotherapy is a topic of ongoing debate. Early diagnosis in paraganglioma cases has been shown to improve the quality of life of patients.

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