Cauda equina paraganglioma: Clinical presentation and radiologic aspects

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Abstract

Paraganglioma is a benign neuro-endocrine neoplasm rarely localized on the lumbar spine. A 50-year-old male who presented for paraparesis and urinary leakage. Lumbar spine MRI showed a lesion compressing the dural sac from L1 to L5. The patient was operated, and the pathologic examination concluded to a cauda equine paraganglioma

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Introduction:

Paraganglioma is a benign neuro-endocrine neoplasm mainly found in the head, the neck, the mediastinum and the retroperitoneum. Localization in the cauda equina is very rare for this tumor. It is frequently misinterpreted with neurinoma and epndymoma in this site (1,2). We report a case of cauda equina paraganglioma.

Observation:

A 51-years-old male with no pathological background presented after the onset of a sudden numbness in his lower extremities associated to urogenital disorders. Physical examination revealed a cauda equine syndrome. Deep-tendon reflexes were abolished in both lower limbs with bilateral foot dorsiflexion weakness (2/5). Hypoesthesia of the lower left extremity as well as saddle anaesthesia was also found. Magnetic resonance imaging (MRI) of the lumbar spine was performed. It showed an intradural well-circumscribed mass extended from L1 to L3. The mass was isointense on T1 weighted images, hyper intense on T2 weighted images with homogenous enhancement after Gadolinium injection (figure 1). Urgent lumbar laminectomy and total resection of a highly vascularized haemorrhagic intradural tumour was performed (Figure 2). No tight adherence were noticed with nerve roots, and tumour resection was macroscopically complete. Pathologic examination of the specimen (Figure 3) concluded to a paraganglioma. The patient had a good postoperative

recovery as well as an improvement of neurological signs. A 2 years follow up did not show neither clinical nor radiological signs of recurrence.

Discussion:

Paragangliomas are tumors whose embryological origins arise from autonomic nervous system (3). Normally, paraganglionic cells migrate along the neural tube. Thus, paragangliomas are a result of dysfunction of embryonic paraganglia, whether related to cell migration or their non-regression (2). Paragangliomas could be found in adrenal and extra-adrenal tissues. Extra-adrenal tumors can be categorized into sympathetic and parasympathetic types (4). The sympathetic paragangliomas have usually the possibility to be hormone-secretant, mostly cathecolamines (Dopamine, Noradrenaline and Adrenaline) (5). Parasympathetic paragangliomas tend to be non-secretory. These forms could result from a local differentiation from tissues that do not derive from the neural crest. Furthermore, the participation of the ependymal cells in their development cannot be excluded, which explains the non-secretory type (1).

In the central nervous system, nearly 80% of paragangliomas occur in the head and neck. They are typically parasympathetic and arise from the glomus jugularis or the carotid body. At level of cauda equina or filum terminale, they are very rare representing 2–4% of cases (2).

The highest prevalence for cauda equine paragangliomas commonly sets around the fourth and fifth decades of life. A male predominance is usually noticed. It is admitted that they are sporadic neoplasms, but nearly 1% of cases are autosomal dominant (5).

Most of the patients present with lumbar or radicular pain. Nevertheless, other clinical features may be found: motor or sensitive deficit, and genital or sphincter disorder. Symptoms of sympathetic secretion related to catecholamine are uncommon (2). There is no significant correlation between the tumour dimension and the duration of clinical symptomatology onset. In the literature, there would be only two cases in which an acute neurologic deficit was found, related to an intratumoral hemorrhage (1).

MRI is the gold standard for both the diagnosis and the follow-up of paragangliomas of cauda equina or filum terminale. Typically, they appear isointense to the conus medullaris on T1-weighted images (WI) and hyperintense on T2-WI, with homogenenous or heterogeneous enhancement after gadolinium injection which is is related to the important vascular supply for the tumor (3). However, these tumors do not have any pathognomonic features. This is why they are frequently misdiagnosed, as they lead to confusion with other tumors as schwannomas, ependymomas, meningioma, teratoma, and hemangioma (2). A serpiginous flow void from vessels seen on the upper pole of the lesion, and signs of intra or peritumoral bleeding can be present.

Histologically, paragangliomas are graded as WHO Grade I tumors. They are slow-growing benign tumors comprised of two cell types, spindle shaped sustentacular cells and chief cells. S100 staining is positive in sustentacular cells, but it is not specific as a possible positivity can be found in ependymomas. Ependymal cells are GFAP positive whereas GFAP staining is negative in neoplastic cells of paragangliomas (4). Mature ganglion cells are contained in nearly half of the paragangliomas of cauda equina.

Total surgical excision is presented to be the best option to achieve greater chance of cure and to reduce recurrences (5). Several authors queried this option because the relatively benign natural history can make from expectation a reasonable option in asymptomatic cases. Peroperative, paragangliomas are most often intradural and extramedullary. They appear well encapsulated, purple, friable and often haemorrhagic. The main technical difficulty is dense adhesion to nerve roots, which can make surgical excision hard without impairing the roots (2). The outcome is overall excellent when complete excision of the mass is performed. Nevertheless, some authors recommend a long-term follow-up due to a possibility of recurrence (1–4%) (2). When only subtotal resection is performed, 10% of the tumours recurred within one year following surgery. In the case of incomplete resection, radiotherapy is recommended with a significant effect on recurrence (3).

Conclusions:

Lumbar spine paragangliomas are very rare and benign extra-adrenal neuroendocrine tumors. Complete surgical resection achieves both cure and symptomatic relief. Excellent prognosis can be achieved without any adjuvant therapy. Care should be taken to distinguish paragangliomas from others with similar radiologic features, but different prognosis and long term outcome.

Key Clinical Message:

Paraganglioma is a benign neuro-endocrine neoplasm rarely localized on the lumbar spine. A 50-year-old male who presented for paraparesis and urinary leakage. Lumbar spine MRI showed a lesion compressing the dural sac from L1 to L5. The patient was operated, and the pathologic examination concluded to a cauda equine paraganglioma

Figures:

Figure 1: (a) Sagittal T1 weighted images MRI shows posterior and anterior epidural adipose tissue compressing the dural sac extending from L1 to L5 segment with high signal intensity. (b) Sagittal T2 weighted images MRI featured multilevel high intensity epidural mass lesion compressing the dural sac.

Figure 2: Photo showing the tumour after removal

Figure 3: Pathologic examination showing a tumoral proliferation made of polygonal cells well arranged in lobules, associated to vascularized stroma

Conflicts of interests:

Author and co-authors declare having no conflicts of interests.

Author contribution statement:

- Ghassen Gader wrote the article
- Mouna Rkhami made the bibliographic research and provided iconography
- Ihsèn Zammel and Mohamed Badri corrected the article

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