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Paraganglioma of the filum terminale mimicking neurinoma: Case report

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Abstract

Background: Paraganglioma of the filum terminale is an uncommon tumor of cauda equina region. Lumbar radiculopathies are revelations that can get complicated from cauda equina syndrome. Magnetic resonance imaging (MRI) allows the diagnosis without distinguishing it from a neurofibroma. Only histopathological study can bring certainty in the diagnosis. The treatment of choice is a complete removal.

Case Description: We report a case of paraganglioma of the filum terminale in a 74-year-old female patient, admitted for cauda equina syndrome, which has been progressing for 2 years. MRI of medulla objectified an extramedullary lesion at L3–L4, diagnosed as neurinoma. A complete resection of the tumor was performed with a section of the filum terminale, which she was taking since birth. Histology confirmed the diagnosis of paraganglioma. The clinical course was characterized by a complete recovery of the deficit.

Conclusion: Through this case report, the authors discuss clinical and radiological aspects of this tumor, as well as the management in the light of published data.

Key Words: Cauda equina, neuroendocrine tumor, paraganglioma

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INTRODUCTION

Paragangliomas are rare neuroendocrine tumors, usually benign. They grow in paraganglia and derive from the migration of neural crest cells that constitute the sympathetic and parasympathetic nervous system. Paraganglioma of the filum terminale is an exceptional tumor, compressing 2.5–3.8% of all cauda tumors.[6,9,22] The clinical picture illustrates that cauda equina syndrome is sometimes associated with neuroendocrine symptoms. Magnetic resonance imaging (MRI) is the key of diagnosis; however, the diagnosis that distinguishes it from neurinoma is anatomopathological, since they are both similar in terms of radiological characteristics. The treatment of choice is a complete removal of the tumor. It allows a better management of the disease and reduces risks of recurrence to 1–4%.[18] Adjuvant therapy, given during partial resection, has not showed any benefit in terms of recurrences.

CASE REPORT

Our patient was a 74-year-old woman, without any particular pathological antecedents, presenting intermittent and badly systemized bilateral sciatica, along with weaknesses in both lower limbs, gradually progressing for 2 years. This symptomatology got complicated by sphincter disorders characterized by urinary incontinence and constipation.

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The clinical examination objectified incomplete paraplegia without sensory loss. Patellar and Achilles reflexes were reduced on both the lower limbs. MRI showed an intradural fusiform lesion at L3–L4, with an isosignal on T1 and T2, associated with a central nodular hypersignal on both sequences. The lesion took up the whole spinal canal, making a scallop shape on the vertebral body and laminating its posterior arch. The tumor homogenously enhanced following a gadolinium injection [Figure 1]. The diagnosis for neurinoma was highly suspected. Through an L3 and L4 laminectomy and an opening of the dura mater, the tumor appeared to be grayish red, closed, very hemorrhagic, and tied to the filum terminale. The lesion retracted nerve roots on the back and on the left side without any invasion. The tumor was attached to the filum terminal and his section has facilitated total removal. The patient fully recovered her motor function after a sphincter and motor function rehabilitation. Macroscopically, the anatomopathological examination showed an encapsulated proliferative tumor, lobulated and surrounded by tiny fiber tracts. It was vascularized with large plaques of hemorrhagic suffusion. Microscopically, we showed lobulated cells with eosinophilic cytoplasms, sharply demarcated, with dense and rounded nuclei, pointing in immunohistochemistry neuron-specific enolase (NSE) and chromogranin A (CgA) [Figure 2].

DISCUSSION

Paragangliomas are tumors that arise from neuroectodermal cells of the extra-adrenal sympathetic or parasympathetic nervous system. They are sporadic tumors; however, their origin has been discussed in many observations.[8] In about 1% of cases, the transmission is done through autosomal dominance.[13] Paragangliomas can be found along the paraganglioma cells in various sites of the body. They belong to a large category of neuroendocrine tumors, known as Amine precursor uptake and decarboxylation, based on their secretory capabilities; in this case, epinephrine, dopamine, serotonin, adrenocorticotropic hormone, and somatostatin.[18] Their localization on the filum terminale is very rare, accounting for 2.5–3.8% of cases.[18,22] A review of 215 cases, reported in a study by Gutenberg *et al*.[6] on paragangliomas cauda equina and paragangliomas of the filum terminale, showed that 60% of paragangliomas occur on males of 44 years of age on average, and from 9 to 74 years of age in extreme cases. From an anatomopathological point of view, paragangliomas are slow-growing tumors, classified as Grade 1 based on the World Health Organization classification.[7] The macroscopic examination of paraganglioma demonstrates an aspect of a tumor well-circumscribed by a capsule richly vascularized. These numerous vessels at the capsule can be responsible for hemorrhagic complications or diagnostic errors leading to a suspicion of arteriovenous malformations,[3] of which the bleeding constitutes a surgical difficulty, as it was seen in our patient. Microscopically, we see a proliferation of polygonal tumoral cells, well demarcated, lobulated, pointing in immunohistochemistry CgA (Novocastra, LK2H10, 1/50e), synaptophysin (Dako, Polyclonal, 1/100e), and NSE (Dako, BBS/NC/VT-H14, 1/800e).[12,17] The diffusion in the central nervous system and distant metastases is rare.[16,21] Paraganglioma of the filum terminale, in contrast to other locations, is less active; therefore, the clinical symptomatology is usually due to the tumoral mass.[4,22] The clinical expression includes lumbago in 50% and radiculalgia in 25% of cases. In <10% of cases, patients have had sensory or motor deficit; and in 3% of cases, constipation or urinary incontinence have been supplementary.[6] A total motor deficit is extremely rare.[1,6] Some authors report a higher frequency of sphincter and genital disturbance, compared to other tumors of cauda

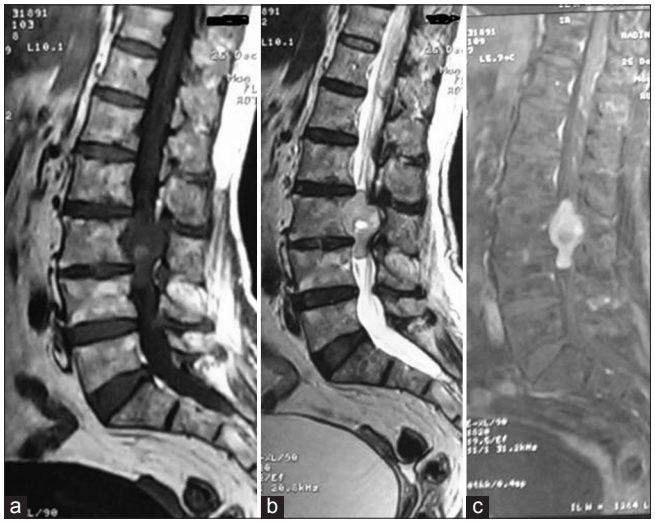

Figure 1: T1-weighted (a) T2-weighted (b) and after contrast (c) sagittal magnetic resonance images showing an intramedullary tumor in the conus medullaris with homogenously enhanced after a gadolinium injection and scallop shape on the vertebral body (×10)

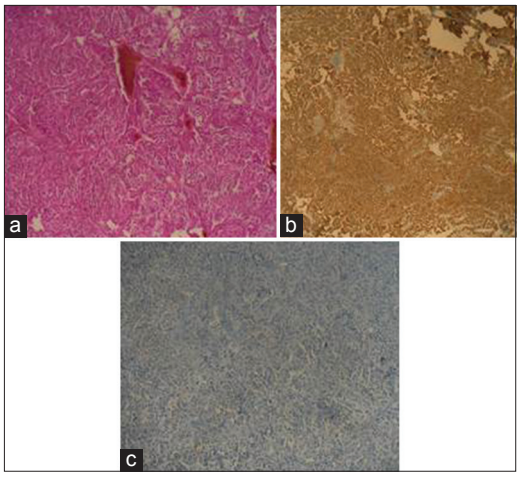

Figure 2: (a) Tumor proliferation image by H and E coloring, (b) chromogranin A demarcates cytoplasm of ganglion cells, (c) negativity synaptophysin immunoreactivity of ganglion cells (×10)

equine.[5,12] MRI plays an important role in diagnosis, but nonspecific.[13,19] It provides rich semiological information, without giving certainty, which is obtained through an anatomopathological study. The tumor is usually isointense or hypointense on T1-weighted and hyperintense on T2-weighted, with a great enhancement after a gadolinium injection.[15] This aspect, on MRI, can be similar to intraspinal neurinomas with comparable radiological characteristics. Other intradural lesions as myxopapillary, ependymomas, and meningiomas can also bring difficulties in the diagnosis.[1,10] We find an evocative aspect in favor of paragangliomas, serpiginous intratumoral vessels, and vascular pedicles of alimentation, which are not found in neurinomas and ependymomas.[10] I-mIBG scintigraphy is not preferable in terms of showing paragangliomas of the filum terminale since they are not or not very secretory in contrast to other extra-adrenal or adrenal locations, where they have an important diagnostic value.[16] The angiography of the medulla, on the other hand, allows the exposure of the most richly vascularized pedicle of the lesion, which allows us to distinguish paraganglioma from neurinoma even more easily.[2,20] Surgery is the treatment of choice. The objective is a complete surgical removal to reduce the risks of recurrence. The tendency of these tumors to bleed and to infiltrate the carrying nerve root makes the total resection very difficult.[11] On the other hand, the insertion of the tumor on the filum terminale makes the surgical resection easier after the section of the filum, which guarantees a cure. The preoperative electric stimulation, in the case of lesions invading the root, is important. It shows us the substitution or not of the motor function by another root, which will determine our surgical attitude and the neurological fate of the patient.[14] Resistances to adjuvant radiotherapy have been observed during partial removal.[3] However, distant and locoregional disseminations are rare.[18,21] Thus, clinical and radiological surveillance of patients who have undergone a partial resection is necessary. In the case of a total removal, some authors recommend a long-term follow-up due to a possibility of recurrence, which is rare about 1–4%.[8,9,18]

CONCLUSION

Paraganglioma of the filum terminale is a rare tumor, often disclosed by lumbago and sciatica that are sometimes complicated by cauda equina syndrome. MRI with gadolinium injection allows setting surgical strategies, but does not bring certainty in the diagnosis, which is obtained through an anatomopathology. The total resection of the lesion, sectioning the filum terminale, guarantees a total cure and prevents recurrences in most of the cases. Nevertheless, a long-term clinical and radiological follow-up is necessary.

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Conflicts of interest
There are no conflicts of interest.

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