Paraganglioma of the Cauda Equina Presenting with Erectile and Sphincter Dysfunction

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Paragangliomas of the cauda equina are rare neuroepithelial tumors, usually manifesting clinically as sciatica. Here, we report a case of cauda equina paraganglioma with an unusual course in a 43-year-old man. His main complaints were erectile and sphincter dysfunction. The low back pain was initially ascribed to accidental injury. Magnetic resonance imaging revealed intradural tumor at the L2/L3 level. The patient underwent gross tumor resection, and the diagnosis of paraganglioma was based on neuropathologic examination. The symptoms completely resolved after tumor resection. [*J Chin Med Assoc* 2009;72(6):328–331]

Key Words: cauda equina, genitourinary involvement, paraganglioma, symptomatology, treatment

Introduction

Paraganglioma is a neuroepithelial tumor that arises from extra-adrenal paraganglia.[1–4] It is most commonly localized at sites of normal paraganglia, mainly intra-abdominally and in the head and neck region, but it may develop in different parts of the body, including the brain.[1–6] Paraganglioma of cauda equina (CEP) is a rare neoplasm, accounting for about 3.5% of tumors in this region.[1,2] Although their pathologic features are homogeneous, CEPs have variable symptoms and outcomes. The most common symptom of this tumor is sciatica; sensory or motor deficits and sphincter or erectile dysfunction are reported less frequently. CEPs are usually benign, but there may be recurrence several years after tumor resection. However, clinical outcome usually depends on the symptoms observed before operation. Urinary dysfunction is considered to suggest a poor prognosis.[1–3,6,7] Here, we report a case of CEP with uncommon symptoms and a very good outcome.

Case Report

A 43-year-old otherwise healthy man was admitted to our hospital due to impotence and pollakiuria. He complained also of numbness of the perineal region, but he ascribed this symptom to the injury he had 7 months previously when he had been struck on the back by a shovel while working in his garden and had suffered from low back pain for several days, radiating to both legs, alternately. The pain was not severe and tended to be alleviated spontaneously. He decided to visit the doctor only when urinary and erectile dysfunction occurred.

On admission, neurologic examination of the patient revealed bilateral straight-leg raising limited to 70° and Achilles tendon hyporeflexibility. No sensory loss was found. Magnetic resonance imaging (MRI) showed a 25-mm spherical-shaped lesion at the L2/L3 level, which was isointense with spinal cord on T1-weighted imaging and had high signal intensity on T2-weighted imaging. Contrast enhancement of the tumor was noted (Figure 1).

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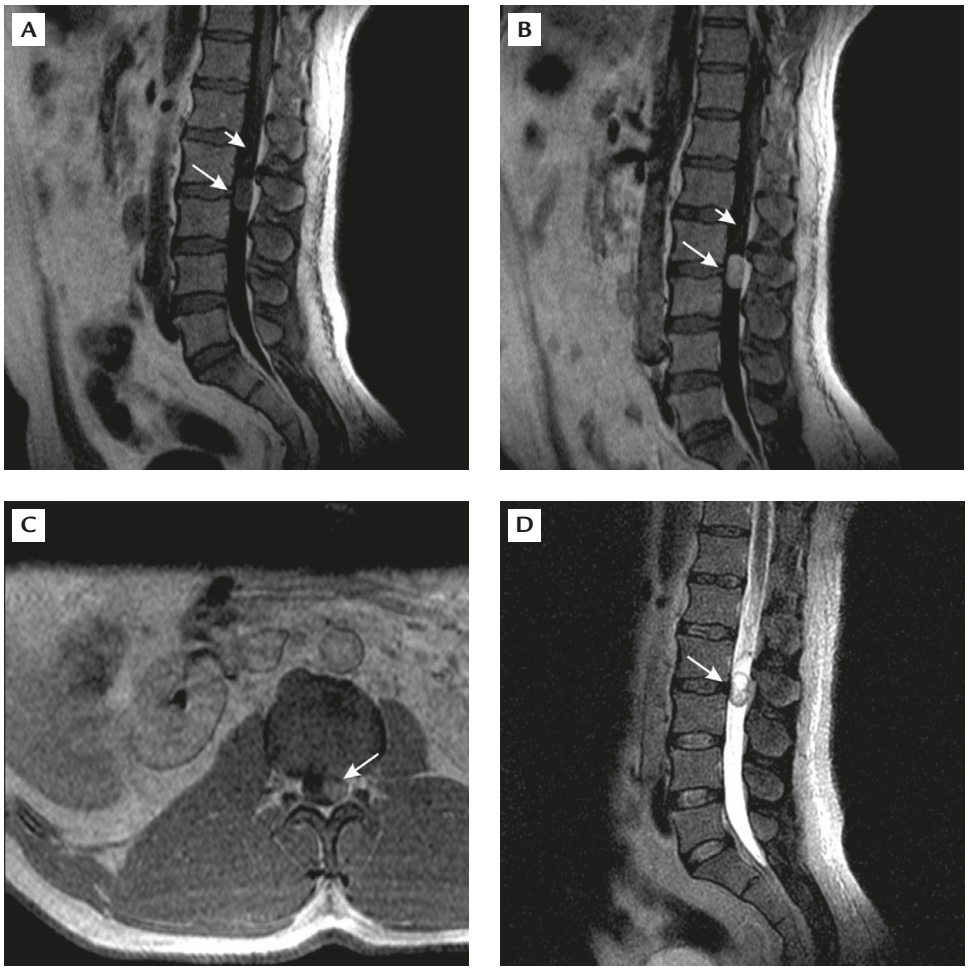

Figure 1. Magnetic resonance imaging of the lumbosacral spine. (A) Sagittal T1-weighted image shows a large intradural tumor isointense to the spinal cord in the spinal canal at the L2/L3 level (long arrow) and the vessel supplying this lesion (short arrow). (B) Sagittal T1-weighted image following intravenous contrast administration. Both the lesion and the vessel are enhanced. (C) Transverse T1-weighted image demonstrates a well-defined spherical enhanced soft tissue filling the spinal canal from the left. (D) Sagittal T2-weighted image shows that the tumor signal is higher than normal spinal cord tissue.

A laminectomy from L2 to L3 was performed to remove a firm, well-encapsulated, pale tumor, located in the intradural space and attached to one of the nerve roots. There was 1 feeding artery entering the tumor along the involved nerve root. Microscopically, the tumor had typical neuropathologic features of paraganglioma, i.e. nests of polyhedral or oval cells, with ample cytoplasm, bordered by thin strips of focally hyalinized connective tissue and capillary vessels (Figures 2A and 2B). Staining for reticulin revealed a dense network encircling the clusters of cells known as "zellballen" (Figure 2C). A few mitoses were seen (Figure 2B), but the tumor did not show any other features that suggested malignant characteristic. Neoplastic cells were strongly positive for chromogranin and synaptophysin (Figure 2D), and some traces of positivity for epithelial membrane antigen in some cells were noted.

The postoperative course was uneventful, and all the symptoms, including urinary and erectile dysfunction, resolved. The patient has remained free of any of the former symptoms for 11 months.

Discussion

Paragangliomas rarely develop within the central nervous system as the existence of paraganglionic cells within the brain and spinal cord is unconfirmed. Some authors believe that paraganglionic cells can be found in the cauda equina and other parts of the central nervous system, whereas others suggest that paragangliomas of the cauda equina do not originate from the neural crest.[1–3,7] All CEPs share the pathologic features of other extra-adrenal paragangliomas. They are composed

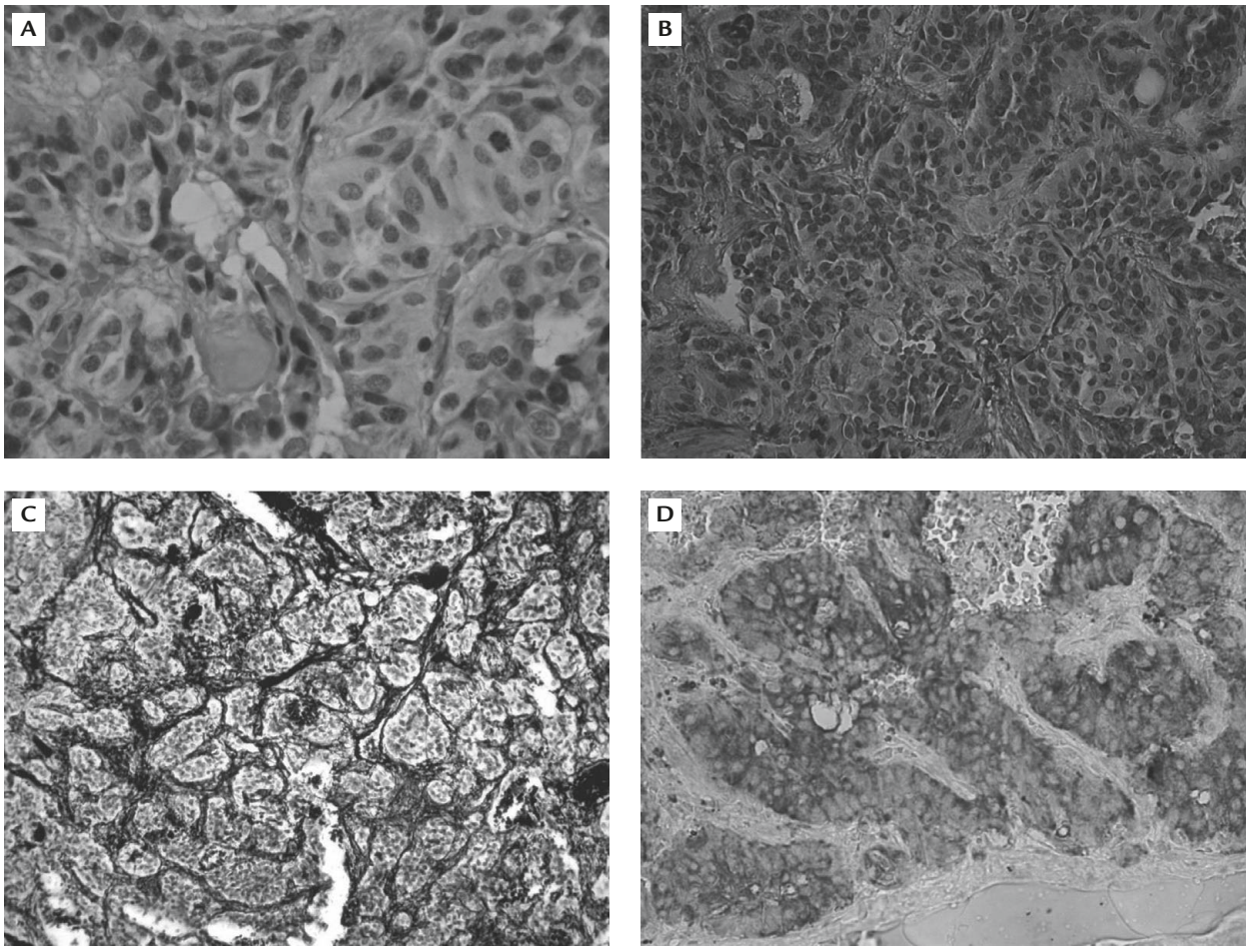

Figure 2. Paraganglioma of the cauda equina. (A) Typical pattern of cells forming nests or lobules ("zellballen") separated by thin vessels and connective tissue (hematoxylin & eosin, 200×). (B) Higher magnification shows relative uniformity of cells, which have rich eosinophilic cytoplasm and round or oval nuclei with delicate chromatin and sometimes barely discernible nucleoli. Mitotic figures are also seen (hematoxylin & eosin, 400×). (C) Reticulin staining emphasizes the lobulated structure of the tumor (Perdrau method for reticulin, 100×). (D) Strong positivity for synaptophysin (200×).

of nests of oval or polyhedral cells with granular cytoplasm surrounded by a fibrovascular network. The tumors are vascularized. Tumor cells were reported to express neuron-specific enolase, serotonin, chromogranin A, synaptophysin and sometimes S-100 protein.[1–3,6] Paragangliomas do not secrete pressor amines, so biochemical preoperative diagnosis is not available. The clinical symptoms of CEP are similar to those of other cauda equina tumors. Moreover, CEPs differ in many features; they can be benign or malignant, and may metastasize. They may also be multiple; however, single tumors are more common. The cases of CEPs reported in the literature differ also in symptomatology. In our patient, there was no classic sciatica, and the low back pain was not the main symptom as the patient ascribed the pain to a back injury. His main complaints were impotence, urinary disturbance, and perineal numbness. These symptoms are rare in CEP.[1,2,4–7]

Neuroradiologic examinations, including MRI, do not yield any characteristic findings for CEP. In this case, based on MRI, we could not exclude schwannoma, ependymoma, astrocytoma, lipoma, epidermoid tumor or metastasis.[1,6–8] Intraoperatively, it was also impossible to establish the diagnosis. The tumor in our patient was rather pale and indistinguishable from neurinoma; however, a darker red color has also been reported.[1,7,8] Cauda equina paraganglioma is usually attached to a nerve root,[1,2] but it can also grow in connection to filum terminale.[1,8] In our case, it was attached to a nerve root.

Gross tumor resection is the treatment of choice in CEP. Consecutive radiotherapy is restricted to malignant or recurring CEPs.[1,3,6,7] The prognosis in CEP is usually good if there is no urinary disturbance. In this patient, total gross removal of the tumor was achieved, with a complete regression of symptoms and neurologic abnormalities. The patient remains under follow-up

as the recurrence of CEP, even after several years, has been reported.

The reported case suggests that patients with genitourinary complaints should be thoroughly examined by a neurologist. If any neurologic symptoms are found, neuroimaging study should be considered.

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