Paranglioma of the Cauda Equina

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Summary: Paranglioma is an uncommon tumor in the cauda equina. Twenty-five cases have been reported in the literature including two cases in Japan. A further case is reported here that presents the largest tumor as the others. The dominant symptom was low-back pain, and mild neurologic deficit. After the extent and the level of the tumor were demonstrated by magnetic resonance imaging and computed tomography, we performed operative treatment. The histologic examination was made by microscopy, with the diagnosis being confirmed by electron microscopy. Two years after the operation, the clinical prognosis is good and no recurrence is recognized by magnetic resonance imaging.

Key words: paranglioma — cauda tumor — magnetic resonance imaging — low back pain — electron microscopy

Introduction

Paranglioma rarely occurs in the spinal canal. Paranglioma of the cauda equina has been reported in only 25 cases in the world literature since first reported by Lerman, et al., in 1972. In 1970, Miller reviewed the first case of these, but he confused it with secretory ependymoma. The characteristic histologic appearance is of a Zellballen pattern that consists of polygonal cells separated by a delicate fibrovascular stroma. However, a diagnosis is accurately confirmed only by electron microscopic examination. We present a confirmed case of paranglioma of the cauda equina in a 49-year-old woman who was treated operatively, that is unusual in the presented enormity of the tumor, and review the previously reported cases.

Case Report

A 49-year-old woman was admitted to

the hospital because of severe low-back pain. The patient had suffered from a low-back pain over the previous 10 years with there having been no apparent cause of the pain. She had been treated conservatively with diagnosing of sciatica over the previous 5 years. A physical examination demonstrated a limitation of the lumbar motion with pain and tenderness in the lumbo-sacral region. She had often complained of severe constipation, but had no bladder dysfunction. Sensation, motor and reflex functions were all normal. On anteroposterior lumbar radiography, the lamina of L5 and sacrum were destroyed with marginal sclerosis, and the interpedicular distance at the level of L5 was increased. On lateral view radiography and on tomography, the posterior margin of L5 and sacrum were concaved with sclerosis (Fig. 1). A myelogram showed a complete block of the upper margin in the L4 body. Magnetic resonance (MR) images showed a large mass from the upper margin of L4 to the S2 (Fig. 2). On computed

Fig. 1 Tomographs of A-P and lateral view. Lamina of L5 and sacrum are destroyed, in A-P view, and the posterior margin of the L5 and the sacrum are invaded by the tumor mass.

Fig. 2 MRI and CT findings. The extent of the tumor mass is clearly demonstrated by MRI. The spinal canal through L5 and S1 is expanded with marginal sclerosis by the tumor mass.

Fig. 3 Microscopic findings. Typical "Zellballen" pattern of paraganglioma (H and E. left $\times 40$ right $\times 100$).

Fig. 4 Electron microscopy. The tumor cell contained numerous membrane-bound dense core vesicles (arrow) ($\times 500$).

tomography, the spinal canal through L5 and the sacrum was extended by the mass, and had a lower density than the muscle, with the lamina becoming thin (Fig. 2). A technetium bone scintigram demonstrated the abnormal concentration on L4, L5 and the sacrum, but a Ga scintigram was normal. An ultrasound examination of the abdomen showed no tumor mass in the retroperitoneal space. A histologic examination of the tumor obtained from an open biopsy revealed paraganglioma.

Laminectomy was performed at L3-S2. On opening the dura, a dark reddish encapsulated tumor was seen with the greatest dimension measuring 13 cm. Bilateral L4, L5, and S1 roots were displaced laterally, and the tumor was found to be adherent to the other roots of the cauda equina. Using microsurgical techniques, the tumor was completely removed piece by piece after incising the filum terminale and involving a necessary resection of the left S2, 3, and 4 nerve roots which could not be dissected from the tumor itself. After the suture of the dura, a bone graft was performed into the concave space of the body, and a free fat graft was performed on the dura. At her 2-year follow-up examination, she had no low-back pain and no muscle weakness, though she showed a slight bladder dysfunction and hypalgesia in the left saddle region. MR imaging showed no recurrence of the tumor. From CT examination, the grafted bone and fat were still alive.

Pathological Examination

Microscopically, the round or polygonal epithelioid cells were arranged in a small nest around a delicate vasculature that could be clearly outlined with a reticulin preparation "Zellballen". The cytoplasm had a granular eosinophilic appearance. The centrally located nuclei were regular and had a finely clumped chromatin. The

tumor cells had no pleomorphism and few mitotic figures (Fig. 3).

Electron microscopy: Many cells contained numerous membrane-bound dense core neurosecretory granules in the cytoplasm. The electron microscopic examination confirmed a diagnosis of paraganglioma (Fig. 4).

Discussion

A paraganglioma usually arises in the paraganglia, with paraganglioma of the head or neck being the most frequently encountered. However, such tumors may occur in the mediastinum or retroperitoneum. To our knowledge, in the region of the cauda equina there have been 25 such cases (excluding our case) reported in the literature. Of these, 14 were male and 11 were female. Most of these cases occurred during the fourth and fifth decades of life, although the youngest patient was 13 years old and oldest was 70 years, with an overall mean of 47.1 years. The characteristic clinical symptoms of these were low-back pain with or without neurologic deficits and without the other symptoms characteristically seen in cases of other caudal tumors. Most of the 25 reported cases were of intradural tumors, arising from the cauda equina, conus medullaris, or filum terminale. Our case involved both the filum terminale and the cauda equina. The size of these tumors in the long axis were reported to range from 1.5 cm to 10 cm, though most of these were within 3.5 cm. Our case was 13 cm in the long axis, and was of the greatest size compared with the others. The paraganglioma has dense neurosecretory granules that implies the presence of biogenic amines. Symptoms related to catecholamines were not reported in cases of caudal paraganglioma. The catecholamine levels in urine and in serum in our case were within normal ranges. The biological

behavior of caudal paraganglioma has not been accurately assessed, since the reported cases have been few in number with limited follow-up data. In the paraganglioma of the neck, the incidence of metastasis was estimated to have been 6 to 9% by Lack et al, (1977) and the incidence of malignancy was reported as 50% by others at the Mayo Clinic. In one case of thoracic epidural paraganglioma, there had occurred recurrence that showed marked signs of malignancy, 8 months after the operation and the patients died 13 months later. However, a malignant paraganglioma in the caudal region has not been reported. Recurrence occurred in 2 cases (10%) of the 20 reported follow-up studies. Both of these had undergone incomplete non-curative extirpation in the initial surgery. Paraganglioma in the cauda equina are cured if complete extirpation can be accomplished at the time of the initial surgery. The prognosis of our case was noninvasively examined by MR imaging, and the nonrecurrence was confirmed at two years after the operation. A long-term postoperative follow-up is necessary because of the slow evolution of these tumors. MR imaging is the best examination for the post-operative follow-up.

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