

Case Report

A case report of filum terminale paraganglioma invading the lumbar body

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Abstract: Paragangliomas within the spinal canal are mainly located in the filum terminale and the cauda equina region. Typical paragangliomas in the lumbar spinal canal show space-occupying of the intradural extramedullary in the lumbar spinal canal, with clear boundaries and obvious intensification, rarely attacking the lumbar body or the spinal dura. We treated one case of filum terminale paraganglioma that invaded the lumbar body and the spinal dura. The surgery has definitive therapeutic effect and the following is the clinical report.

Keywords: Paraganglioma, lumbar body, spinal dura

Introduction

Paraganglioma is a kind of neuroendocrine tumor that occurs in various parts of the body, mostly the head and the neck, and is mainly located in the carotid body and the jugular bulb [1]. Paragangliomas are rarely seen in the filum terminale or the cauda equina region, usually located in the intradural extramedullary, and occasionally found outside the dura, involving the conus medullaris or the nerve root of the cauda equina. There have been few reports of paragangliomas invading the filum terminale of the lumbar body or the cauda equina region so far [2, 3]. We treated one case of filum terminale paraganglioma invading the lumbar body and the spinal dura. Now the clinical report is given as follows and the literature is reviewed (Table 1).

Case report

A 39-year-old man who had back pain and numbness 20 years ago without obvious inducement. One month earlier his back pain worsened after hard work, and he felt pain in bilateral inguinal regions and numbness in bilateral lower extremities, especially the right one. Lumbar vertebrae MR scan and lumbar vertebrae three-dimensional CT showed hyper-

intense T1WI of a lump about the size of 6.9 cm * 4.3 cm * 2.8 cm, isointense and hyperintense T2WI, which reinforced clearly after intensified scan. Lumbar vertebral body 2, 3 and the surrounding bones were morphologically irregular, the corresponding horizontal spinal canal expanded, and the form was incomplete (Figure 1).

Preoperative diagnosis considered the high possibility of schwannoma. After complete preoperative examinations, space-occupying excision in lumbar spinal canal 2, 3 was performed under general anesthesia. Considering the obvious damage to lumbar vertebral body 2, 3 indicated by lumbar spinal MR and three-dimensional CT, the surgery was performed by the collaborations of neurosurgeons and spine surgeons under neurophysiological monitoring in order to maintain the postoperative stability of the lumbar vertebrae. Bone graft fusion and internal fixation was performed by the spine surgeons in advance, then the neurosurgeons performed intraspinal tumor resection. The surgery took the posterior midline approach from the back of the waist. The spine surgeons performed lumbar vertebrae side fixation and opened the vertebral plate L2, 3, 4 with piezosurgery. The tumor, with rich blood supply, was seen eroding the endorhachis, and reaching the

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Table 1. Reported cases of paragangliomas of the spine

Author (Year)	Age	Sex	Location	Operation	Follow up (time)
Fusun DO et al. (2009)	75	M	L3-L4	Total	Well
	70	F	L1-L2	Total	Well
	50	M	L3-S1	Total	Well
Mauricio SG et al. (2012)	26	M	T9-T11	Biopsy	NA
Sushrut MF et al. (2013)	60	F	L3-L4	Total	Well
R Bhatia LZ et al. (2013)	33	M	L5-S1	Total	Well (12 Mons)
Mehmer S et al. (2014)	36	M	L2	Total	Well (8 Mons)
	40	M	L4-L5	Total	Well (6 Mons)
Han SO et al. (2014)	76	F	L2-S1	Subtotal	Well
Rodolfo C et al. (2015)	33	F	L3	Total	Well (3 Yrs)
Chou SC et al. (2016)	58	M	L2-S1	Subtotal	NA
Erika D et al. (2016)	32	M	L3	Total	Well

L3 epidural space. The spinal dura was cut open under a microscope to confirm the upper and lower boundaries of the tumor, and to separate the surrounding cauda equine nerves. The tumor, resected by blocks, was gray red, tough, and closely combined with the filum terminale, from which the tumor was cut off and the stump electrocoagulated. The stump was repeatedly washed by saline, and confirmed of no clear bleeding. Since large areas of the spinal dura had been invaded and disappeared, it could not be tightly sutured, in case the imperfect place on the dural mater would be covered. One artificial dura mater was sutured with vertebrae attached with muscle. The vertebral plate was returned and fixed with a titanium strip. A drainage tube was left outside the spinal dura mater. The muscle, aponeurosis, subcutaneous and the skin were sutured. Intraoperative blood loss was about 1500 ml. 800 ml of red blood cells were transfused, as well as 200 ml of plasma.

Since the dura of the patient was severely attacked by tumor during the surgery, anhydrous suture was impossible. The patient was placed in the prone position after he awoke from general anesthesia. A drainage tube was left outside the vertebral plate for 8 days for continuous vacuum drainage; the daily volume of drainage was about 400 ml, and the drainage tube was removed on the 8th day. Then deep suture was given in the sinus tract. No cerebrospinal fluid incision leakage occurred. The patient was discharged on the 20th day and asked to stay in bed for 3 months. The patient's postoperative course was uneventful, and during the 2-year follow-up period, no evi-

dence of clinical or radiologic recurrence was found.

The pathological shows paraganglioma, and the immunohistochemistry of tumor shows: Vim (+), CD56 (+), Cga (+), Syn (+), GFAP (-), EMA (-), PR (-), S-100 (-), CD34 (vascular +), Ki67 (focal + 5%) (**Figure 2**).

Discussion

Exo-adrenal paraganglioma is more common in the carotid artery and the jugular bulb. It can also be found in the thyroid gland, eye socket, medi-

astinum, lung, retroperitoneum, uterus, bladder, spinal canal and so on. Paragangliomas within the spinal canal are mainly located in the filum terminale and the cauda equina region. Therefore, it is also called CEP (Cauda equine paraganglioma). It is more common in adults, and in men slightly over women, aged from 13 to 71. There have been reports of paraganglioma occurring in the thoracic vertebrae [3] and the cerebral curtain [4]. Since under normal circumstances, paraganglion does not exist in the brain or the spinal cord, so the origin of paraganglioma in the brain and the spinal cord remains unclear. Most CEP are sporadic cases, some related to familial diseases including multiple neuroendocrine tumors IIA, IIB and III, Von Hippel-lindau syndrome, neurofibromatosis, and Sturge-Weber syndrome.

Clinical manifestations of intraspinal paragangliomas are mostly non-specific, and are related to the location of tumors and the presence of endocrine functions. The most common clinical symptoms of paraganglioma in the cauda equine region is low back pain and sciatica, with or without lower extremity motor or sensory loss, and occasional bladder and bowel dysfunctions [5]. Jeffs [6] and others reviewed the literature and found that there have been only 3 case reports of intraspinal functional paraganglioma. Since it releases catecholamines, in addition to the aforementioned symptoms of spinal cord compression, there are also symptoms of high blood pressure, flushing, diaphoresis, tremors, weight loss, tachycardia, nausea, vomiting and others. Hemorrhagic shock can be ruled out, but we do not rule out the partial neuroendocrine functions of the tumor.

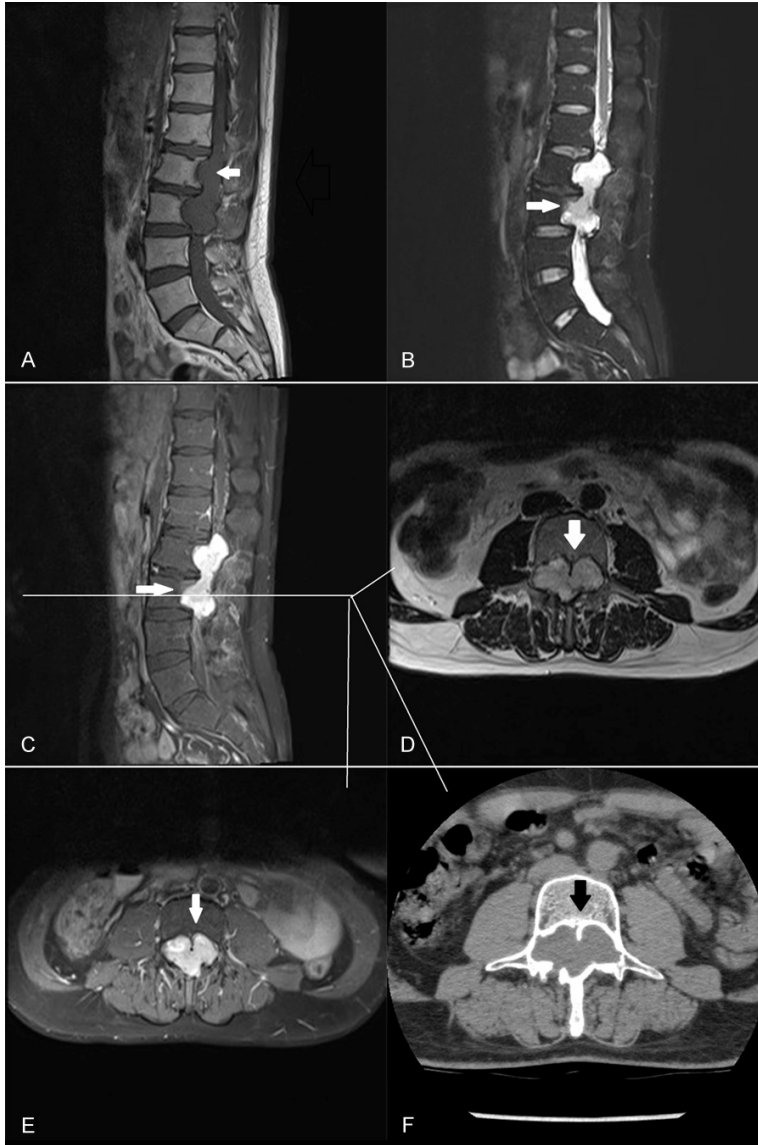


Figure 1. Lumbar spine MR scan and lumbar vertebrae three-dimensional CT showed visible hyperintense T1WI of a lump about the size of 6.9 cm * 4.3 cm * 2.8 cm (Whithe arrow in A, D; D was the level of L3), isointense and hyperintense T2WI (Whithe arrow in B), which reinforced clearly after intensified scan (Whithe arrow in C, E; E was the level of L3). Body of the third lumbar vertebral body 2, 3 and the surrounding bones were morphologically irregular in CT scan (F), the corresponding horizontal spinal canal expanded, and the form was incomplete (Black arrow).

MRI is the preferred intraspinal lesions. Three-dimensional CT of the vertebral body can display with more detail the bone condition of the vertebral body. Typical CEP shows hypointense or isointense on T1WI, hyperintense on T2WI, and distorted, strip-type low signal is visible, which is the flowing void effect produced by the slow flow of blood, ie, a salt-and-pepper appearance. Meanwhile a low signal band is often visible around on T2WI image, which might be the chronic hemosiderin deposition

caused by bleeding tumor; the tumor is significantly strengthened after enhanced scanning, indicating a rich blood supply, and consistency with the pathological changes, which is the basis of the diagnosis of highly vascular tumors. The diagnosis of paraganglioma should be distinguished from schwannoma, neurofibroma and spinal meningioma, and also from some neoplasms rich in blood vessels, such as hemangioblastoma and ependymoma [7].

Before the surgery, the MRI T2WI of the patient showed flakes of high signal shadow, which were considered to be hemosiderin deposition caused by tumor bleeding, consistent with the imaging of paragangliomas. In addition, this patient was preoperatively misdiagnosed as schwannoma, mainly because the tumor extruded from the intervertebral foramen, and showed a dumbbell-like change. The imaging of this tumor indicated obvious invasion of the L2 and L3 vertebral body, which was clinically rare. Obvious invading lesions of local spinal canal include schwannoma, ependymoma and paraganglioma [2]. For such tumors, bone graft fusion and internal fixation is preferably performed along with tumor resection, in order to maintain the stability of the vertebral body.

Paragangliomas are mostly benign tumors. Complete surgical resection is the primary treatment. In this case the posterior midline surgical approach was employed. The surgical method is the same with traditional intraspinal surgery. Since the patient had vertebral body damage, combined spinal surgery was performed along with interbody fusion with internal fixation. Complete postoperative suture of the dura was difficult after it had been invaded by tumors. Based on our experience, non

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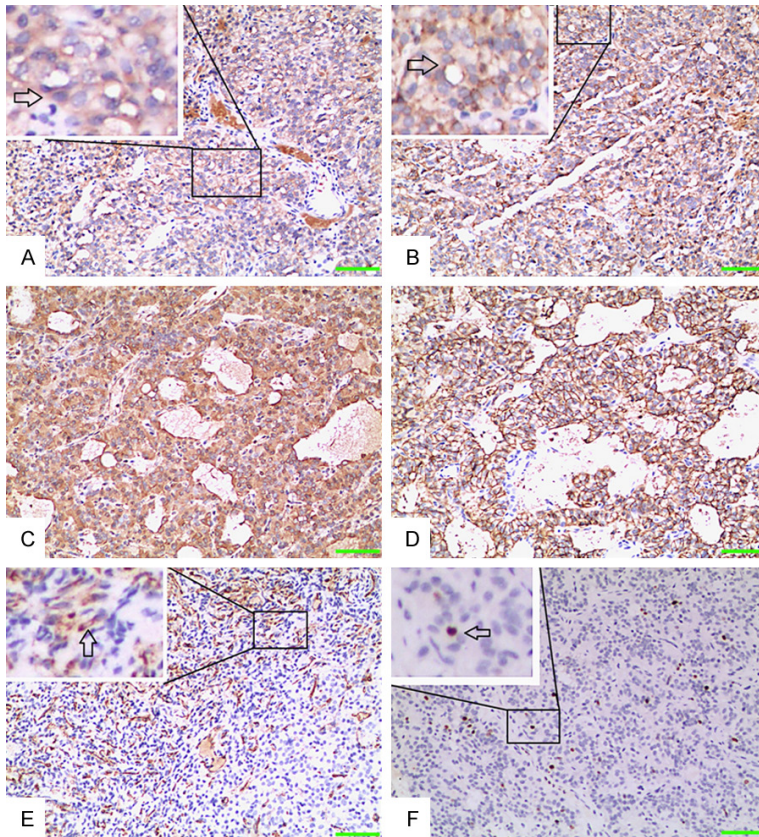


Figure 2. The immunohistochemistry of neoplasma showed light brown by Syn (A, arrow showed the distribution of Syn in plasma), Cga (B, arrow showed the distribution of CgA in plasma), Vim (C), CD56 (D), CD34 (E, arrow showed the distribution of CD34 in vascular endothelial cell), Ki67 (F, focal + 5%, arrow showed the distribution of Ki67 in nuclear).

stitched artificial dura mater should be laid out on the surface of the residual tumor cavity. The length and width of the artificial dura mater should be greater than the dural defect so that it can be placed under the normal dura. Then another artificial dura mater was sutured with the muscles attached with vertebrae. Since the paraspinal muscle was severely damaged during the opening process, it was more difficult to suture there. So during the process of opening the vertebrae, the paraspinal muscles should be preserved as much as possible to facilitate suturing when taking into account tumor invasion of the dura mater. The vertebral plate was returned by titanium bars and titanium screws after the suture. The muscle, fascia, subcutaneous, and the skin were sutured layer-by-layer. After the surgery, drainage tube was left outside the vertebral plate for conventional vacuum drainage, the drain should attached to bulb suction and exit from a separated stab incision; the volume of drainage was controlled between

350-400 ml, and the chance of leakage of cerebrospinal fluid from the skin incision was reduced. After the surgery, anti-positive bacteria drugs were used to prevent infection. The drainage tube was removed after one week, and the sinus tract was sutured tightly. The presence of cerebrospinal fluid incision leakage was checked after the removal and the surgical dressing was kept dry. The re-examination three months after the surgery found no residual tumors in the patient, and no other special treatment was performed. Whether further postoperative radiation therapy is necessary is currently controversial. Most scholars believe that this belongs to the benign tumors. No further radiotherapy is necessary after the resection. The whole tumor cannot be resect alone or radiotherapy is not allowed for patients in general status. However, some scholars believe that the problem can be solved with simple radiotherapy [8]. The

intraspinal paraganglioma is rich in blood supply and blood sinus itself. Intraoperative bleeding is ferocious especially when the tumor invades the vertebral body, often disabling complete resection of the tumor or the termination of the surgery. For tumors considered to be paraganglioma preoperatively, we recommend the preoperatively viable tumor feeding artery embolization to reduce blood loss. Intraoperative transient hypotension occurred in the patient after resection of the tumor for about 3 minutes. We cannot rule out the possibility of the tumor having partial endocrine functions.

Diagnosis of paraganglioma mainly relies on pathological diagnosis. Observed by naked eye, the tumor became oval, slightly lobulated, and elastic, with a smooth surface, often closely attached to the vessel wall. The section is from gray red to reddish brown, very rich in blood vessels; sometimes it even looks like hemangioma. Under the light microscope, the image of

endocrine tumor is visible. It is constituted by nests of epithelioid cells and separated by rich fibrovascular stroma dilated into a vascular sinus shape. Around the nests are supporting cells, while nerve fibers are often difficult to see. The tumor cells are arranged like spumescence or nests, separated by rich fibrovascular stroma dilated into a sinus shape. The argyrophil fiber presents a distinctive reticular structure after being dyed. Immunohistochemical specific proteins play a decisive role in the diagnosis of tumor cells, among which NSE, Syn, and CgA are strongly positive, and NSE and CgA are most valuable. The diagnosis can be definite as long as one of the three is positive. CgA has a bigger value than NSE. Tumor cells typically do not express CK, EMA, and GFAP [9]. The Syn and CgA of this case of patient were positively expressed, which confirmed the diagnosis of paraganglioma.

Intraspinal paragangliomas are mostly benign, and rarely malignant. Sometimes it's even difficult for pathological tissue to determine whether the tumor is benign or malignant. The diagnosis should also consider its biological behaviors. If distant metastasis and (or) lymph node metastasis occur with the tumor, it can be considered malignant paraganglioma. Therefore, long-term follow-up visits are of great significance for determining the prognosis. Thorough and complete resection of the tumor is the key to preventing recurrence, improving survival rate and reducing disability rate. If the tumor cannot be resected completely, postoperative radiotherapy can relieve the clinical symptoms and improve the long-term survival rate of patients. Most scholars believe that chemical treatment is of little significance [10].

Conclusion

Cauda equine paragangliomas are rare benign tumors, and which invade into the lumbar body is even rare. Optimal treatment includes surgical resection when feasible. In case of subtotal tumor removal, radiation is other treatment modalities.

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Disclosure of conflict of interest

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