Huge refractory retroperitoneal nerve sheath tumor treated with radiofrequency ablation: a case report with literature review

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Nerve sheath tumors include both benign and malignant schwannomas and neurofibromas. They occur frequently in patients of 20–45 years old. Nerve sheath tumors are mostly found in the roots of cranial nerves (mainly the acoustic nerve) and spinal nerves. They also occur in the head, neck, face, stomach, mediastinum, intracran, spinal canal, and so on. Peripheral nerve sheath tumors are relatively rare, which are seen in soft tissues of the limbs and neck, but seldom in retroperitoneal and pelvic nerve [1-4]. Nerve sheath tumor constitutes about 4% of all retroperitoneal tumors [5]. Between 1978 and 2008, 32 patients with retroperitoneal nerve sheath tumor were treated by surgical resection at Sun Yat-Sen University Cancer Center, including 18 cases of benign tumors and 14 cases of malignant tumors. We reported here a case of huge refractory retroperitoneal nerve sheath tumor treated with CT-guided radiofrequency ablation (RFA). The patient had tumor recurrence 1 year after combined treatment, and had local failure to radiotherapy and arterial chemoembolization. CT-guided RFA was then successfully performed in December 2005. No recurrence was detected during the follow-up till the end of 2009.

Case report

A 50-year-old woman suffered from repeating and deteriorating indistinct pain in the upper left abdomen and bearing down sensation in May 2004. CT examination at local hospital revealed retroperitoneal massive space-occupying lesion. CT scan at another hospital revealed two connected retroperitoneal masses in the left abdomen with size of 18 cm × 16 cm and 17 cm × 10 cm, respectively, which was diagnosed as “retroperitoneal tumor, detail undefined” and treated with exploratory laparotomy and partial tumor resection under general anesthesia. A 2 cm × 2.5 cm mass surrounding the abdominal blood vessels could not be fully removed. Pathological examination showed it was a nerve sheath tumor; immunohistochemical examination showed it was Vim (+), Syn NF (+), SMA (-), HF (-), CD117 (-), CD34 Lyo (-). The symptoms were alleviated after operation. The patient felt uncomfortable in her abdomen again 1 year later. CT scan revealed the recurrence of nerve sheath tumor with its diameter increased to 17 cm. The patient received 2 times of arterial chemoembolization (detailed medication unknown) and local radiotherapy (dose unknown) in another hospital. After the second arterial chemoembolization, the patient felt weakness and moderate pain in her left lower limb and walked unsteadily. She had to take orally weak-opioid analgesics. PET-CT scan at our hospital in December 2005 showed tumor residue with a SUV value reached 4.6–7.6 and the size increased to 17.4 cm × 11.5 cm, which invaded the erectors spinea (Figure 1). Physical examination showed active position, no obvious abnormalities in the heart and lungs, a palpable huge mass in middle-right part of the abdomen, and some moderate indistinct pain in May 2006.

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Figure 1  CT image of the relapsed malignant peripheral nerve sheath tumor
The huge malignant peripheral nerve sheath tumor relapsed after surgical resection. Tumor occupies the retroperitoneum, invades the lumbar vertebra, and pushes the left kidney onwards.
the lower abdomen, medium texture, unclear margins, slight tenderness, no shifting dullness, no percussion pain in renal region, normal borborygmus, normal myodynamia of the right lower limb, level 4 myodynamia of the left lower limb, and no pathologic reflex.

According to the results of PET-CT scan, CT-guided RFA was performed on this patient for 3 times in our department in December 2005, January and June 2006 (Figure 2). The tumor was ablated using Radionics Cool-tip R-F Ablation System powered up to 200 W. The electrode probe has a 3-cm exposed tip. Output power adjusted automatically according to the changes of tissue impedance during ablating process. In addition, the system detected real-time temperature in circumambient tissues. One ablation cycle was 12 min. Temperature in ablated region ascended to 70–110°C evenly by pulsed current to assure tissue necrosis in the ablated region. A single ablation site was 3.5 cm × 4.2 cm. Upon completion of tumor FRA, the probe track was cauterized to decrease the potential risk of track bleeding.

The retroperitoneal nerve sheath tumor in this patient was completely inactivated after a 4-hour continuous large-scale RFA and 2 complementary ablations. The patient received physical and imaging examinations every 3–6 months. In 2009, no recurrence was detected by PET-CT and enhanced CT examinations (Figure 3).

**Discussion**

Nerve sheath tumors clinically manifest as slowly growing painless tumors, which is usually asymptomatic. With the growth of the tumors, patients experience neurological dysfunction as well as pressure and pain in adjacent organs. Retroperitoneal tumor can be easily detected by CT and MRI scans. Because these tumors mostly contain blood vessels, fat, hemorrhagic and necrosis tissues, enhancement degree of the tumor is closely
related to its origin. For a huge mass, it is difficult to determine its origin\(^{[10]}\). Nerve sheath tumors are usually benign and 5\%–18\% of them are related to neurofibromatosis (von Recklinghausen disease)\(^{[8]}\). Malignant nerve sheath tumors are rare sarcoma that arise in deep soft tissue and are usually adjacent to nerve cords. About 4\% of nerve sheath tumors transform to malignant disease\(^{[8]}\) with high risk of lung metastasis in advanced stage. The pathologic features of nerve sheath tumors include single lesion, intact capsule, hard texture and connection with adjacent nerves\(^{[9]}\). Nerve sheath tumors can be categorized into type Antoni A and type Antoni B under microscopy\(^{[10]}\). Dense arrangement of abundant spindle cells can be seen in pathologic examination, and occasionally polymorphous cells with many mitosis arranged in spiral-shape are also observed. Nerve sheath tumors are further diagnosed by positive histoimmunochemical staining of S-100 protein, vimentin and Myelin basic protein\(^{[6,11]}\).

Since nerve sheath tumors have intact capsule, patients could be cured after complete removal of tumors. Some researchers believe that even simple enucleation of tumor is tending to cure\(^{[7]}\). Recently, a case of retroperitoneal nerve sheath tumor treated by laparoscopic resection has been reported\(^{[12]}\). However, some other researchers believe that nerve sheath tumors may be huge and adhered to surrounding organs. This anatomical complexity results in great challenge of surgical treatment for nerve sheath tumors. In addition, some nerve sheath tumors are malignant in origin. It is necessary to excise en bloc any adjacent margins or organs around tumors if the malignance of the tumors cannot be excluded before operation or by frozen section during operation. This is based on the observation that compared to wide excision of malignant nerve sheath tumor results in a recurrence rate of 11.7\% margin excision results in a recurrence rate of 72\%\(^{[13,14]}\). Hence, complete surgical resection should be emphasized at present.

The case reported here was pathologically diagnosed as malignant peripheral nerve sheath tumor, with follow-up of over 5 years after surgical resection. Because part of the tumor was adherent to main vessels, some active tumor mass were not completely removed and led to a residual tumor which enlarged to 17 cm in diameter after one year. Theoretically, this tumor can be re-excised, but re-excision can not preclude the recurrence of tumor since the possibility of incomplete removal of the tumor mass increases as the tumor get larger.

High-dose chemotherapy includes ifosfamide, anthracyclines, combined anthracyclines, vincristine, and so on. Applying chemotherapy to suppress tumors before surgery can help the surgical excision and postoperative chemotherapy, but the objective response rate of high-dose chemotherapy is about 12\%\(^{[11]}\), and it should be applied according to individual clinical situation. Postoperative radiotherapy can also reduce local recurrence rate, but has no obvious effect on long-term survival. Combination with radiotherapy and chemotherapy can be considered in case of lung metastasis\(^{[15,16]}\).

Interventional arterial chemoembolization can be used to identify blood vessels that supply tumors. Sensitive chemotherapy drugs and selective intra-arterial embolization can then be used to embolize tumor supply vessels. However, the existence of multi-vessel blood supply including traffic branches is highly possible in huge tumors. Therefore, interventional arterial chemoembolization is only suitable for palliative treatment. Figure 2 shows high metabolic activity in the tumor after embolization, which confirms the above argument. In this reported case, the supplying blood vessel was originated from lumbar artery and palliative embolization therapy is one choice. However, the lumbar arteries may have same branches as supply vessels of the spine, or have anastomotic branches with the anterior spinal artery. If DSA imaging examination fails to clearly show these anastomotic branches, embolizing the anterior spinal artery can lead to motor dysfunction of the lower limbs. Indeed, our patient experienced weakness in the left lower limb after the second chemoembolization, which was possibly due to spinal motor nerve damage. It is less likely related with tumor compression since embolization produces ischemia in the tumor and thus shrinks it in a certain degree.

RFA has been used to treat malignancies for 20 years in the world. Our center is the first one that applies RFA in China for 10 years. The principle of RFA is to use high-frequency current, thereby the temperature of ablation region (usually 4–5 cm in diameter) increases quickly to 70–110°C. This high temperature physically kills tumor cells in ablation region, which mimics the surgical excision. With the guidance of CT, MRI, or B-ultrasound, RFA completely destroys tumors in a micro-invasive way. RFA has been most successfully applied in liver tumors, and is also applied in lung tumor\(^{[17]}\), bone tumor\(^{[19]}\), adrenal tumor\(^{[18]}\), renal tumor\(^{[20]}\) and other solid tumors. Our case is the first one that achieved salvage ‘cure’ using CT-guided RFA. The keys of the success are summarized below: (1) The physician needs to be familiar with retroperitoneal anatomy and imaging anatomy to avoid the unwanted impact of ablation electrode on important functional tissues and organs. (2) The physician needs abundant operating experience and skills to master physical attribute of ablation electrode, so that to achieve precise control of ablation. (3) RFA is mostly performed in a region of 3.5–4.5 cm in diameter. The physician needs a good sense of three-dimensional space to assure that there is no blind area in tumor ablation. (4) The physician should follow the principle of ‘from shallow to deep’ to avoid tumor planting. (5) For large tumors, RFA should be performed under general anesthesia to ensure adequate treatment time and to achieve the purpose of ‘cure’. (6) Due to the repeatability of RFA, additional treatment is necessary. (7) Upon completion of tumor RFA, the probe track is cauterized to decrease the potential risk of track bleeding. (8) CT scan has great advantages compared with ultrasound to locate large tumors, because CT-guided ablation can completely avoid gas artifacts arisen from the first ultrasound-guided ablation, which will interfere with the succeeding position of ablation electrode.

The design of ablation electrode is more scientific than ever. The electrode now is as small as 17G in diameter, which causes similar body injury as compared with needle biopsy. No charring occurs in ablation electrodes even heated up to 100°C due to its impedance or water cooling system. This feature assures the...
completeness of local ablation. In addition, great advance in guidance device such as open MRI will greatly extend physicians’ ‘eyes’ and achieve a more desirable monitoring role"[13,19].

References