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Case Report

BONE METASTASIS OF MEDIASTINAL PARAGANGLIOMA: CASE REPORT

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ABSTRACT

Mediastinal paraganglioma is a rare and slow growing neurogenic tumor. Here, we describe a 54-year-old male with a non-functional anterior mediastinal paraganglioma. Complete resection is the standard treatment of a paraganglioma because of the tumor's potential malignancy and poor response to chemo- or radiotherapy. However, the highly vascular nature of the tumor and its characteristic anatomic location make complete resection difficult.

Keywords: Paragangliomas; diagnosis; mediastinum; surgical treatment

INTRODUCTION

Case report: A 54 year's old male, one month ago, there was no obvious cause of sacrococcygeal pain, no chest tightness, no urinary and fecal disorders. Recently, patient feel lower back ache. Computed tomography (CT) examination of sacral vertebra showed space-occupying lesions of sacral 4 (Fig. 1a, 1b). Sacral Magnetic resonance imaging (MRI): An irregular mass was seen in the sacral 4 vertebral body, with slightly high signal on T2WI and high signal on lipid-pressing T2WI (Fig. 1c, 1d). Preoperative physical examination revealed posterior mediastinal mass. Chest CT: There was an irregular mass in the right anterior mediastinum, which was lobulated and uneven in density. On contrast-enhanced scan, the mass in arterial phase was obviously unevenly enhanced, while the mass in venous phase was continuously enhanced (Figure 2); on the left side, the 8th rib was destroyed with soft tissue mass. On contrast-enhanced scan, the mass was obviously unevenly enhanced (Figure 3). Immunohistochemical staining: CgA (+), Syn (++), S-100 supporting cells (+), Vim (-), CD34 blood vessels (+), CK (-), Ki-67 positive index was about 10-20%. Puncture biopsy of posterior mediastinal and sacrococcygeal tumors guided by B-mode ultrasonography. The specimens showed mild atypia and nested distribution of tumor cells; with abundant sinusoidal fissures. The immunohistochemical studies suggested that the tumor was a paraganglioma.[1] However, the diagnosis of malignant paragangliomas is generally difficult.

DISCUSSION

Metastasis is the medical term for cancer and metastasis can spreads to a different part of the body. This case of mediastinal paraganglioma with multiple bone metastases is malignant. Paragangliomas are rare, usually non-cancerous growths in the paraganglia, groups of cells located near nerve cells bunches called ganglia.[2] A paraganglioma is considered cancerous (malignant) if it spreads to other parts of the body. This is a rare occurrence in paragangliomas. Cells from a cancerous paraganglioma can migrate to bone, lungs, lymph nodes, liver, or anywhere in body. Mediastinal paragangliomas are 2% of all case. Paragangliomas are rare extra-adrenal tumors of sympathetic or parasympathetic paraganglia origin [3] Mediastinal paragangliomas, slow-growing neoplasm's that comprise nearly 2% of all paragangliomas, can be divided on the basis of their location: tumors in the anterior mediastinum (arising from parasympathetic paraganglia) or those in the posterior mediastinum (arising from sympathetic chain)[4]. The treatment of paragangliomas involves complete tumor removal. In one report, the complete tumor removal rate was 76.9%, and only 20.0% of patients who were thought to have undergone complete tumor resection experienced late recurrence[5]. The survival rate associated with complete resection was 84.6%. On the other hand, the survival rate was only 50.0% for patients who underwent only a biopsy or partial resection and adjuvant treatment ($P < 0.01$)[6]. When the tumor is hypervascular and invades surrounding vascular structures, complete tumor resection may be challenging and result in massive bleeding. To prevent perioperative massive bleeding, preoperative embolization of the tumor-feeding vessels may be essential[7].Paragangliomas are extra-adrenal pheochromocytomas, rarely located in the mediastinum. If these tumors are functional, they can classically

present with sustained arterial hypertension, headaches and diaphoresis. [8] However, these are rare, representing only 2% of all catecholamine-secreting tumors. Most mediastinal paragangliomas are non-functional. These are mostly asymptomatic but can present with compressive local symptoms.[9] The incidence of malignancy in extra-adrenal paragangliomas has been reported between 21% and 76% [10] . After establishing the diagnosis of paraganglioma, imaging studies allow anatomic localization of the tumor. We used a CT-guided biopsy to confirm the diagnosis before operative planning. These tumors are not sensitive to chemotherapy or radiation, and complete surgical resection provides the only chance for cure. This allows long-term (10-year) survival in 84% of patients with complete and 50% with incomplete resection[6] . The hypervascular nature of these tumors is well known, and substantial hemorrhage has been reported during resection[11]

Follow up
A physical exam to check for indications such as high blood pressure
A history of symptoms, health habits, past illness and other treatments
24-hour urine test to measure levels of catecholamine (adrenaline and non-adrenaline) and substances affected by them
Blood catecholamine studies measure levels the hormones and affected substances in the blood
CT scan (computed tomography) and MRI (magnetic resonance imaging) to produce detailed views that can show a tumor

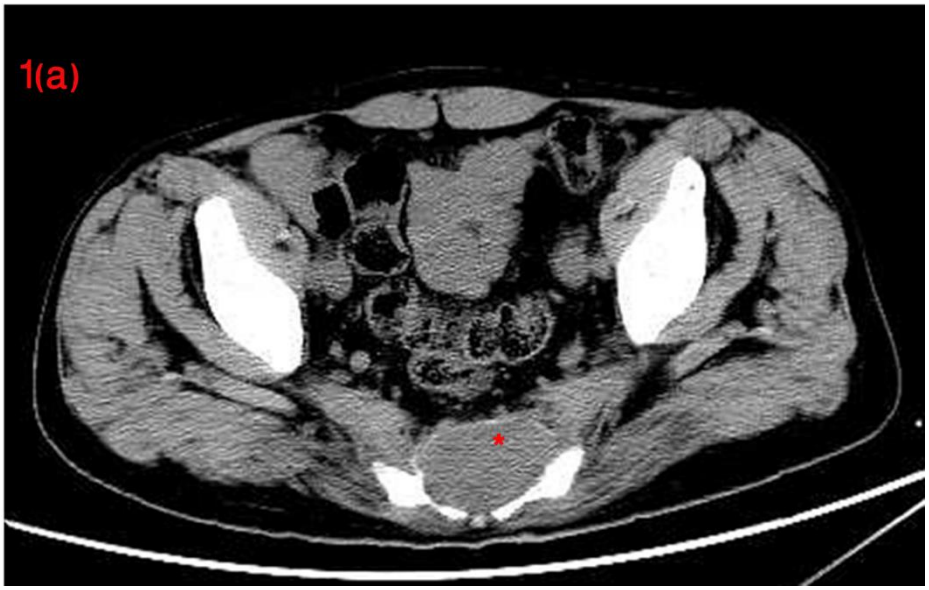
Table 1

Paragangliomas as one of three types:
(1)Localized paraganglioma meaning found in only one area.
(2) Regional paraganglioma, the tumor is a cancer that has spread to nearby tissues or lymph nodes.
(3) Metastic paraganglioma, when the cancer has spread to other organs, bone or lymph nodes far from the original tumor.

Table 2

Paraganglioma is a group of non-epithelial neuroendocrine tumors occurring in paraganglion. The origin of adrenal medulla is called pheochromocytoma, accounting for about 90%, and the occurrence of extraadrenal paraganglioma is about 10%. Paragangliomas outside the adrenal gland are more common in the abdomen than in the mediastinum. At present, it is believed that only the invasion of surrounding structures and/or distant metastasis of tumors is the reliable basis for judging malignancy.[12] This case of mediastinal paraganglioma with multiple bone metastases is malignant. The enhanced CT scan of this patient showed obvious enhancement in the early stage and continuous enhancement in the late stage, which accorded with the enhancement characteristics of paraganglioma. This is also the most characteristic imaging manifestation

of paraganglioma. There is no difference between benign and malignant paraganglioma in the degree of enhancement. Paragangliomas can occur in different parts of the body and should be differentiated from other diseases by their enhanced features. In mediastinum, it should be differentiated from giant lymph node hyperplasia, both of which are obviously enhanced, but the signal on T2WI of paraganglioma is higher than that on giant lymph node hyperplasia, and often accompanied by necrotic cystic degeneration, with uneven density/signal. In vertebral body, it should be differentiated from chordoma and giant cell tumors of bone; unlike paraganglioma, the mass of paraganglioma grows centrally at the vertebral body, and the mass of chordoma generally grows towards the anterior edge of the vertebral body; however, it is difficult to differentiate paraganglioma from giant cell tumors of bone based on imaging findings. Paraganglioma is mostly benign and rarely malignant. It is difficult to determine benign and malignant tumors by histology. In this report, immunohistochemical staining could be used to effectively analyze the shape of paraganglioma.[1] Hence, it should be determined according to its biological behavior. If the tumor has lymph node metastasis or/and distant metastasis, it can be considered as malignant paraganglioma. Complete removal of tumor by 1-stage resection is the first choice for the treatment of paraganglioma in the spinal canal. If the 1-stage operation can completely remove the tumor, the prognosis is good. [13] Surgical resection and endoscopic resection were the only two curative methods used historically in treating these tumors, and the permanent pathological examination was the gold standard to obtain a correct and precise diagnosis. There was only one report of a case that received adjuvant radiation[14]. In summary, surgical resection combined with radiotherapy can effectively alleviate clinical symptoms, and improve long-term survival rate.[15]



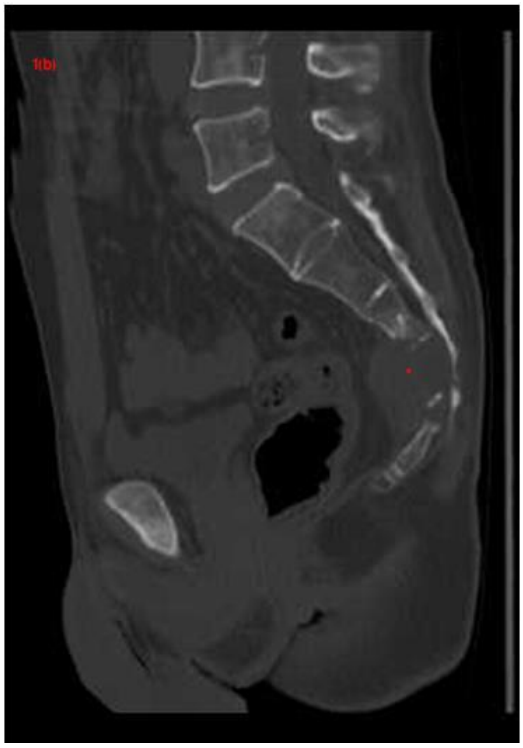
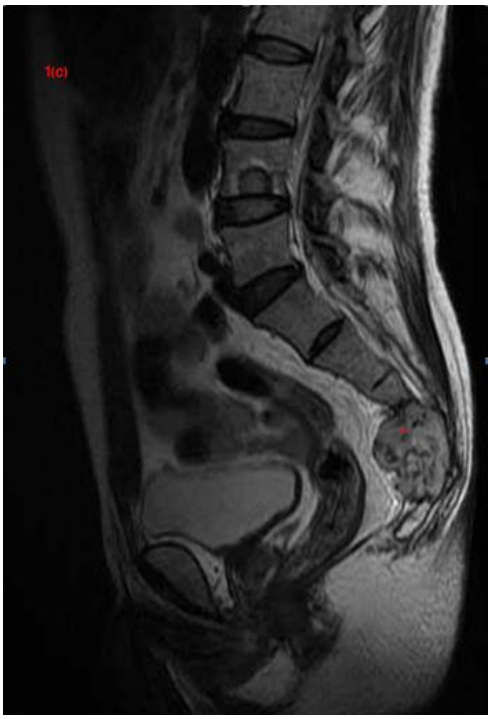


Figure 1 a-b*:* CT shows bone destruction and mass formation of sacral 4 vertebrae



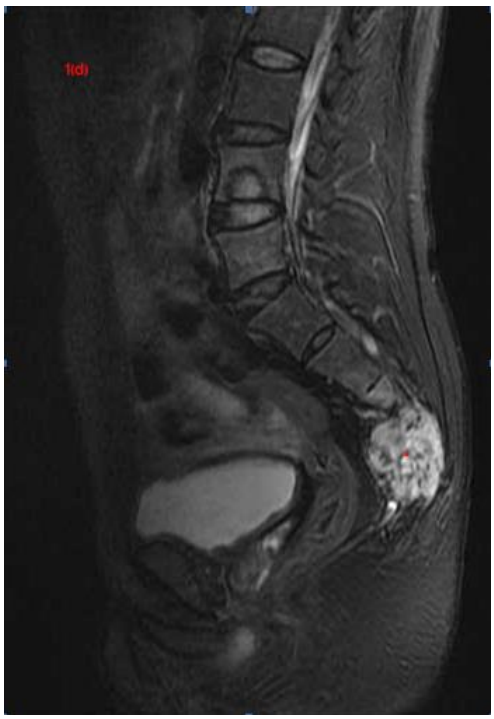


Figure 1c-d: Sacral 4 vertebral bodies, slightly hyperintense mass on T2WI and hyperintense lipid pressure on T2WI

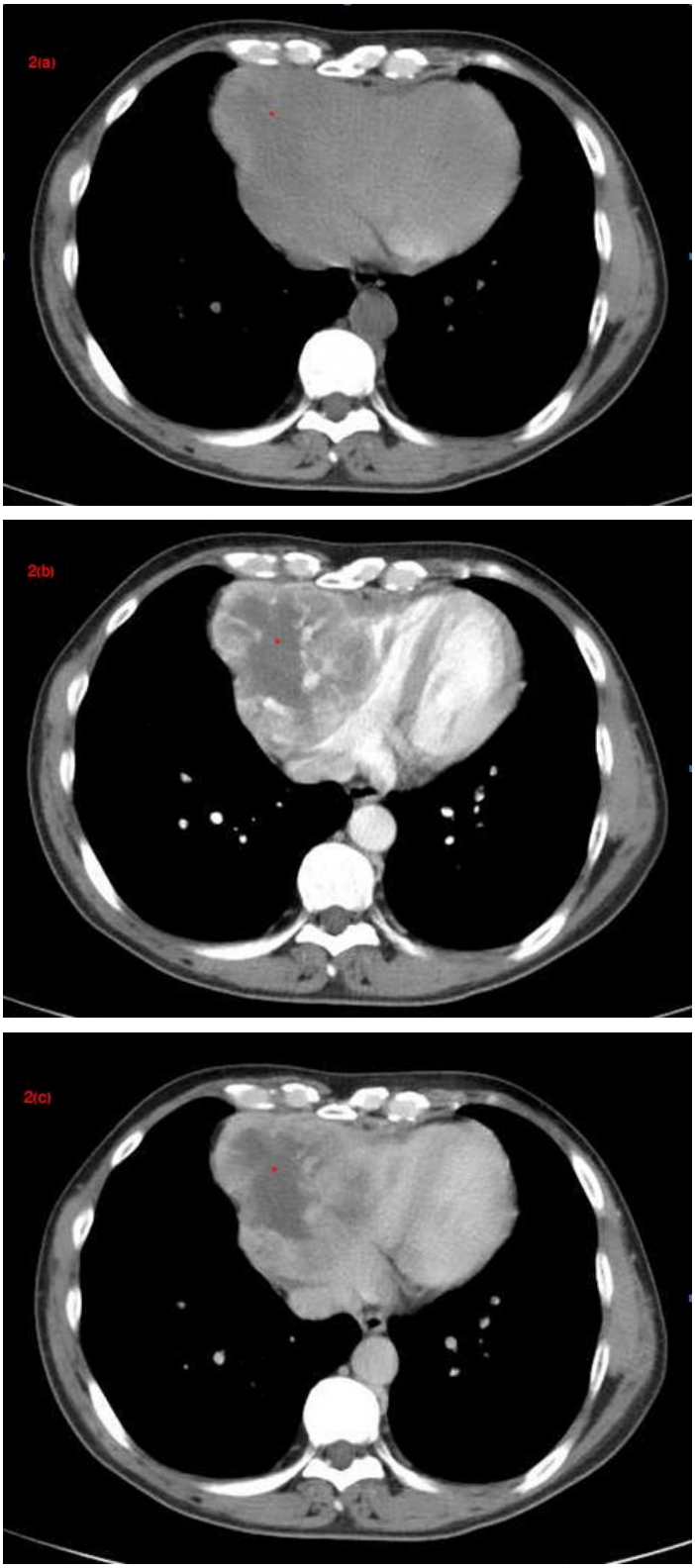


Figure 2a-c: CT plain scan showed irregular mass on the right side of anterior mediastinum with uneven density, obvious uneven enhancement in arterial phase and continuous enhancement in venous phase.

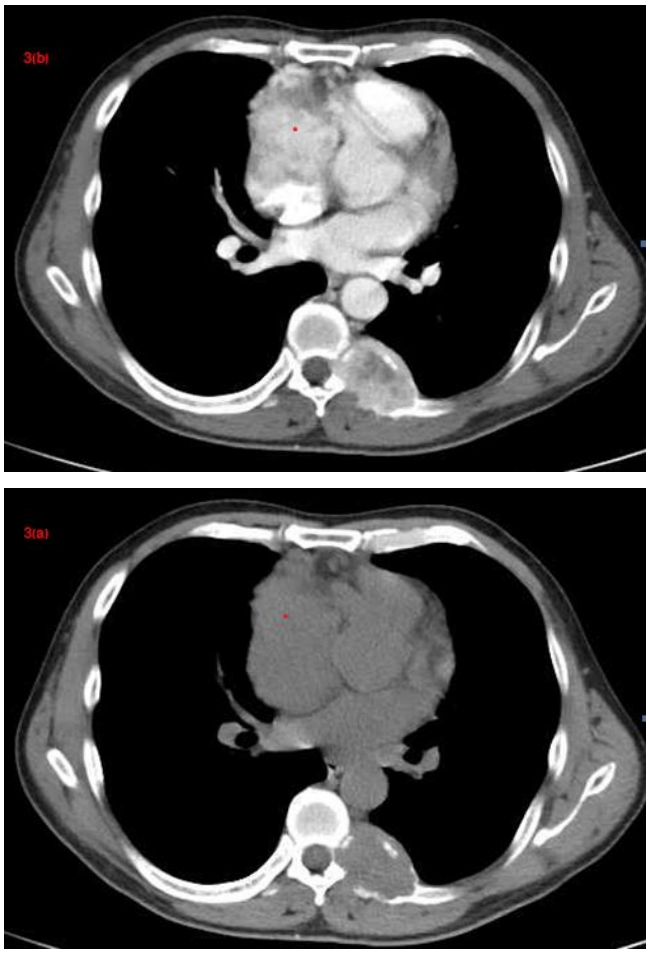


Figure 3a-b: CT plain scan showed destruction of the left eighth rib bone and formation of soft tissue mass. On contrast-enhanced scan, the mass showed obvious heterogeneous enhancement.

CONCLUSION

Paragangliomas, rare and often asymptomatic tumors, are of difficult diagnosis and should be considered malignant tumors, due to the potential aggressive behaviour of cases with high mitotic index and the frequent possibility of recurrence and metastases. Surgical resection is the treatment of choice and careful intraoperative manipulation is recommended, due to the high vascularity of these tumors, to prevent complications. After complete excision, long-term prognosis is generally good. However, even after surgical removal, a close, periodical and life-long follow-up is mandatory.

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Conflicts of Interest:

The authors declare no conflicts of interest

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