A Multimodal Therapeutic Approach To Functional Paraganglioma Of The Thoracic Spine

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Abstract

Functional paragangliomas of the thoracic spine are uncommon malignancies that require a multidisciplinary therapeutic approach. These catecholamine-secreting tumors increase the risk of life-threatening complications such as hypertension, cardiac arrhythmias and massive bleeding during surgery due to an excessive vascular supply. Appropriate manipulation of functional paragangliomas reduces morbidity and mortality. In the article below we present a case of functional paraganglioma, which was resected via surgery after arterial embolization. Following the resection, the patient has remained disease-free for one year.

INTRODUCTION

Tumors arising from neural crest derivatives are generally found in the adrenal medulla, and commonly known as pheochromocytomas [1]. Those tumors that arise in extraadrenal locations are uncommon and known as paragangliomas [1,2]. Mediastinal paragangliomas are typically nonfunctional tumors [2]. However, functional paragangliomas are also uncommon and actively produce vasoactive amines [2]. Functional paragangliomas require a multidisciplinary therapeutic approach that includes appropriate control of hypertension, pre-operative embolization of the feeding arteries of the mass and surgical resection [2,3].

Extraadrenal functional paraganglioma account for 0.3% - 0.5% of patients being investigated for hypertension and those catecholamine-secreting tumors increases the risk of life-threatening hypertension and cardiac arrhythmias [3,4]. Preoperative tumor embolization decreases hemorrhaging from the tumor during surgical manipulation and decreases surgical morbidity and mortality [3]. In the report below we discuss a functional paraganglioma case of posterior mediastinum, which was treated with a multidisciplinary approach that included vascular radiology, thoracic surgery and neurosurgery.

CASE REPORT

A 32 year-old man was admitted to a state hospital with a complaint of left pleuritic pain. His past medical history was clear except for controllable hypertension, which was regulated with irbesartan combined with hydrochlorothiazide. His thoracic computerized tomography (CT) revealed a large left paravertebral soft tissue mass (80 x 76 x 56 mm) extending from the thoracic vertebrae T6 to T7 (Figure 1a). Mass view invading left half of T6 vertebral corpus and neural foramen at the left lung lower lobe was detected and a thoracic magnetic resonance imaging (MRI) was requested. Upon monitoring the paravertebral located mass at the posterior mediastinum in the thoracic MRI examination of the patient, he was referred to our hospital (Figure 1b). As high involvement (SUVmax: 24.1) was detected only in the mass in the positron emission computerized tomography (PET-CT) examination, a decision to conduct a tru-cut biopsy was made. The biopsy revealed the tumor to be neurogenic without a definitive histopathologic diagnosis.

Consequently, a decision of surgery was made in order to make a definitive diagnosis. In the exploration, the paravertebral sulcus located elastic mass was not adhesive to the lung and a vascularized surface of the mass was observed. By opening the parietal pleura, it was attempted to reduce the adhesiveness of the mass to the thoracic wall. During the manipulation on the mass, increase in arterial tension values (250 - 300 mmHg) were observed. Since the mass was severely vascularized and did not allow manipulation, the operation was completed, and it was decided that firstly arterial embolization and then surgical operation would be done.

Endovascular embolization was performed and multiple feeding vessels were embolized before the surgical procedure due to the highly vascular nature of the tumor. After the embolization, the patient was taken to the operating room again by combined thoracic surgeons and a neurosurgery team, and resectioning of the mass was performed successfully (Figure 2a). After the mass was excised, hemivertebrectomy was added to the operative procedure.

In the pathologic examination, neuroendocrine neoplasm characterized by a nested (Zellballen) pattern without necrosis which is typical for paraganglioma, was observed (Figure 2b,c). On immunohistochemical examination, the tumor cells were positive for CD 56, synaptophisin and neuron specific enolase; but negative for pancreatin and epithelial membran antigen (EMA). Postoperatively, the patient made an uneventful recovery without any neurological symptoms. At the one year postoperative follow-up examination, the patient was normotensive and had a disease-free survive.

DISCUSSION

Pheochromocytomas, first described by Frankel in 1886, can be seen in any location within the sympathetic nervous system, but most commonly in the medullar part of the adrenal gland [1,3]. Extra-adrenal pheochromocytomas of the neural crest are known as paragangliomas and account for approximately 15% of all pheochromocytomas [5,6]. Most extra-adrenal paragangliomas are located in the abdomen; therefore mediastinum is a less commonly involved site [7].

Paragangliomas are generally diagnosed in the second and third decades of life, without sex predominance [3]. Functional paragangliomas typically secrete catecholamines; as a result, patients generally present with symptoms of hypertension, palpitation, headache, sweating, tremor and weight loss [3]. They are generally discovered during the surveillance of hypertension and other symptoms whereas non-functional paragangliomas are usually asymptomatic and found incidentally [7]. Our patient had a medical history of hypertension, which was regulated with anti-hypertensive drugs but without any screening modality or diagnostic test for adrenal medullar pathology.

Paragangliomas are extremely well vascularized tumors [2]. If the diagnosis is confirmed preoperatively, angiography for evaluating the tumor vascular supply and embolization for preventing intraoperative hypertension and reducing perioperative catastrophic bleeding is advocated [1,2,7]. Both Nawaz et al. and Kwan et al. [1,3] reported the importance of pre-operative embolization of the paraganglioma in managing catastrophic blood loss in their case reports. Our patient's tru-cut biopsy revealed a neurogenic tumor but did not offer a correct diagnosis for the paraganglioma. Hence, bleeding during surgery was unpredictable but the vascular embolization was performed successfully.

Screening modalities play a major role in both the diagnosis of and decision for surgery. The location of the tumor can usually be identified with CT and MRI, however, MRI is considered to be the superior imaging modality in detecting extra-adrenal paragangliomas [1,3,4]. The MRI features of paragangliomas include well-demarcated masses with low/intermediate signal intensity on T1-weighted scans and intermediate/high signal intensity on T2-weighted scans [6].

The management of spinal paragangliomas requires a collaborative multimodality endeavor including internal medicine (appropriate tension regulation), vascular radiology (embolization of the feeding arteries), thoracic surgery and neurosurgery disciplines [1]. The posterolateral thoracotomy approach seems better for both limiting bleeding and reconstruction of the vertebral column. If bleeding becomes difficult to control, the tumor excision should be postponed and angiographic embolization to the vascular supply of the tumor should be performed. As tumors originate from neural tissues, a neurosurgery team plays a major role in the surgical excision. The possibility of intraspinal extension of neurogenic tumors requires a combined approach for safety during the resection. Because thoracic manipulation of the tumor may cause bleeding within the tumor; hemorrhagic expansion of the tumor inside the spinal canal can result in cord compression and paralysis and this possibility heightens importance of neurosurgery.

Histological and immunohistochemical findings of paragangliomas demonstrate distinctive features. Dense vascularization and microcystic areas, typically consisting of neuroendocrine cells, which are uniformly clustered in nests known as the Zellballen formation, are surrounded by delicate fibrovascular stroma [4,5]. Additionally, paragangliomas stain positively for chromogranin A, synaptophysin, and S-100 protein and tumor specific antigen.

Paragangliomas show a variable prognosis depending on their anatomical location [2]. According to Nawaz et al. [1] recurrence after total resection or metastatic dissemination of the tumor (both cerebrospinal or hematogenous) is possible. Thus, close follow-up of patients is most appropriate.

CONCLUSION

The treatment of functional paragangliomas requires careful pre-operative assessment and a collaborative multidisciplinary approach that includes endocrinologists, vascular radiology, thoracic surgeons and neurosurgeons. Pre-operative embolization decreases intra-operative blood loss and increases surgical success. Adequate treatment requires total surgical removal. Therefore, the possibility of a functional paraganglioma should be kept on mind as a differential diagnosis especially for patients with hypertension.

Figure 1

(a) Thorax CT and (b) MRI revealed a large left paravertebral mass extending from thoracic vertebrae T6 to T7.



(a) In the exploration paravertebral sulcus located smooth

surface elastic mass was observed, (b) Macroscopic examination of the mass after the resection, (c) Microscopic examination of the paraganglioma with Zellballen pattern without necrozis.



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