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An Unexpected Diagnosis of Femoral Paraganglioma: A Case Report

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Abstract

Paragangliomas are rare neuroendocrine neoplasms, commonly located in the head and neck. They are sometimes first found in the bone, especially when there are contributing factors such as trauma, which can make clinical diagnosis more challenging. In this reported case, a patient presented with a painful mass in the right thigh caused by trauma. During the procedure for resecting the right femoral mass, the patient's blood pressure significantly increased upon touching the tumor. Subsequently, a biopsy of the right thigh femur and a whole-body contrast-enhanced computed tomography (CT) examination confirmed the diagnosis of a right femoral paraganglioma with metastasis to the right neck.

Keywords

Paraganglioma, Pheochromocytoma, Trauma, Distant Metastasis, Case Report

Introduction

Paraganglioma is a neuroendocrine tumor that originates outside the adrenal gland. Its incidence rate is 1 in 300,000 [1]. According to the latest WHO classification, the disease is no longer differentiated as benign or malignant, but is primarily categorized based on whether it has metastasized [2]. As a neuroendocrine tumor, some paragangliomas can secrete catecholamine substances, leading to increased blood pressure and heart rate. Therefore, for functional paragangliomas, which exhibit three signs: headache, palpitations, and sweating [3], prediction before surgery is possible, allowing for appropriate preoperative preparation. However, some functional paragangliomas lack the typical features mentioned above but are triggered during surgery, indicating the possibility of the disease. This not only increases the difficulty of preoperative diagnosis but also significantly raises perioperative risks, especially when confounding factors interfere, thus rendering the diagnosis of this disease more challenging. This case represents the first report of the incidental diagnosis of functional paraganglioma in the femur as a result of a right thigh mass after trauma.

Case Presentation

A 53-year-old female patient developed a lump in her right thigh following a collision injury to her right leg, accompanied by pain that could be alleviated by rest. The patient initially dismissed the symptom and did not seek any medical treatment. However, six months later, the pain in her right thigh returned, along with an increase in the size of the lump, and this time the pain persisted even after resting. An MRI examination conducted at a local hospital revealed a fracture in the upper part of her right femur and

associated soft tissue changes. Malignant tumor lesions such as osteosarcoma or metastatic tumors were considered.

Upon admission, the patient's physical examination revealed the following: Blood Pressure: 125/80 mmHg, Temperature: 36.2°C, Pulse: 80 beats per minute. A firm mass of approximately 3 centimeters (cm) x 2 cm was observed at the angle of the mandible on the right side of her neck. No abnormalities were found in the local skin. A substantial lump measuring around 12 cm x 10 cm was noticeable on the front part of the middle section of her right thigh. This lump was firm and accompanied by tenderness and pain upon percussion. The right lower limb showed significant atrophy, with intact skin. The right hip joint exhibited good flexion and extension activities, demonstrating muscle strength at level IV for both hip flexion and extension. Muscle tension was normal, while the rest of her limbs displayed regular muscle strength and tension. The circumference of the middle section of her right thigh was 2 cm smaller than that of her left thigh (**Fig-1A**).

This patient had a history of left lymph node tuberculosis, and she did not have a history of hypertension, palpitations, or excessive sweating. A PET/CT scan revealed hypermetabolic activity in lymph nodes in the neck and abdomen, bilateral adrenal glands, right maxilla, and the upper part of the right femur, indicating the presence of multiple malignant tumors, possibly lymphoma. A biopsy of the mass in the right cervical area suggested a potential tuberculosis infection. Following six months of anti-tuberculosis treatment, the mass in the right cervical area significantly reduced in size. However, there was no substantial change in the size of the mass in the right femur (**Fig-1B**).

As a result, a plan was made to proceed with the resection of the mass in the right femur. During the surgical procedure, the patient's blood pressure markedly increased upon touching the tumor, reaching a peak systolic pressure of 200 mmHg (**Fig-2**). Based on intraoperative exploration, it was suspected that the tumor might be a primary growth originating from the femur, possibly a pheochromocytoma or paraganglioma. Consequently, a local tumor resection biopsy was performed. Subsequent postoperative pathology confirmed the presence of pheochromocytoma/paraganglioma.

A 24-hour urine test for metanephrines and catecholamines yielded the following results: dopamine, 295.95 μg/24h (normal range: 65-100 μg); metanephrines, 12.56 μg/24h (normal range: 30-180 μg); norepinephrine, 58.44 μg/24h (normal range: 15-80 μg); epinephrine, 5.36 μg/24h (normal range: <21 μg). Plasma levels of metanephrines and

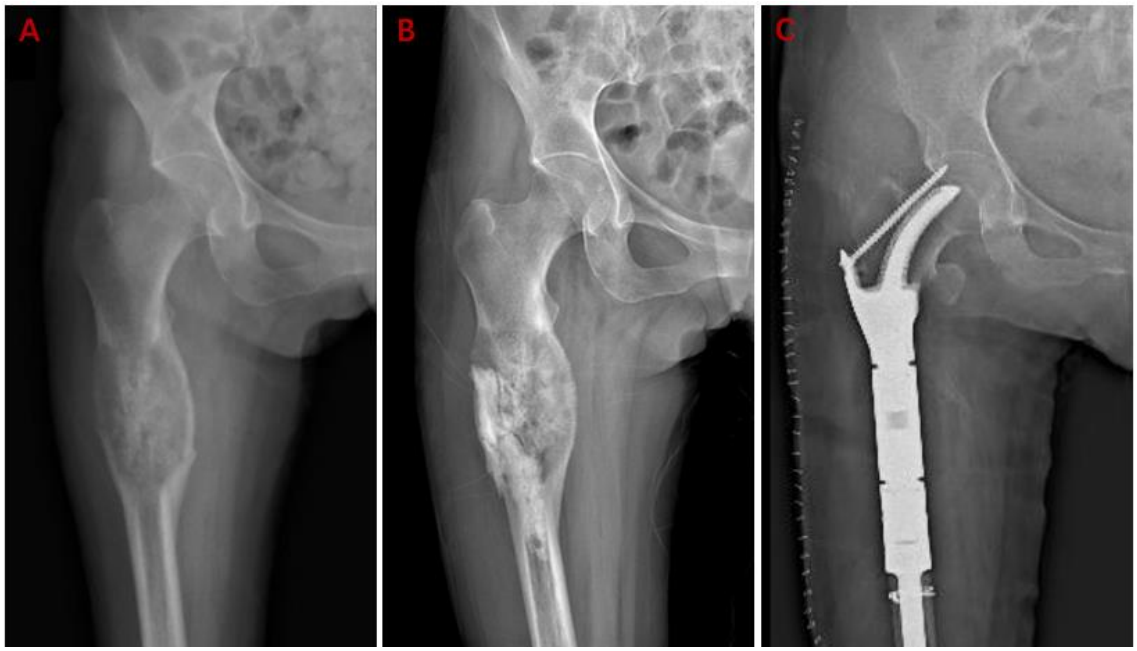

Fig-1: Treatment Process Diagram of Right Thigh Paraganglioma

catecholamines were as follows: dopamine showed a mild increase at 0.39 nmol/L (normal range: <0.31); metanephrines, 0.21 nmol/L (normal range: <0.71); norepinephrine, 3.09 nmol/L (normal range: <5.17); epinephrine, 0.31 nmol/L (normal range: <0.34). Abdominal contrast-enhanced CT imaging showed no tumor lesions in either adrenal gland.

Following thorough preoperative preparation, including hypotension and volume expansion treatment, the tumor in the right femur was successfully removed, and a three-dimensional artificial femoral head replacement surgery was performed (**Fig-1C**). Throughout the operation, there was a significant increase in blood pressure when the tumor was being separated, necessitating continuous administration of nitroglycerin and nicardipine to lower blood pressure. Once blood supply to the tumor was blocked, blood pressure began to decrease, and a small dose of norepinephrine was required to maintain normal blood pressure until the conclusion of the surgery (**Fig-2**). After the operation, the patient's blood pressure returned to normal.

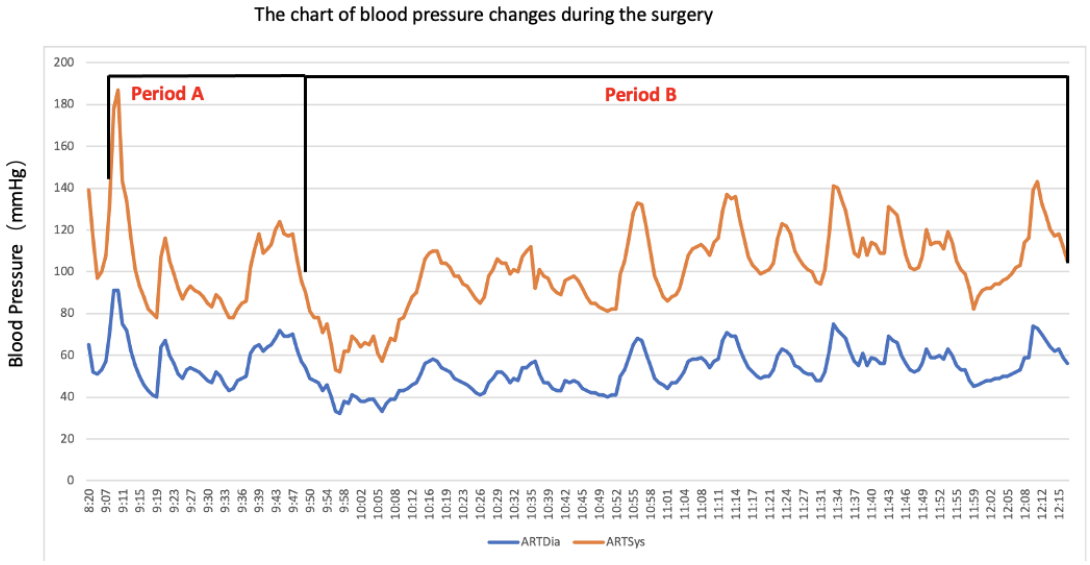

Fig-2: The Chart of Blood Pressure Changes During the Surgery of Right Thigh Paraganglioma

The operation time divided into two period time (period A and period B). Period A: the time from intubation to the tumor separated, the blood pressure increased significantly, and needed continuous injection of Nitroglycerin and nicardipine to reduce blood pressure. Period B: the tumor blood supply was blocked until the end of the operation, the blood pressure began to declined significantly, and a small dose of Norepinephrine was needed to maintain normal blood pressure.

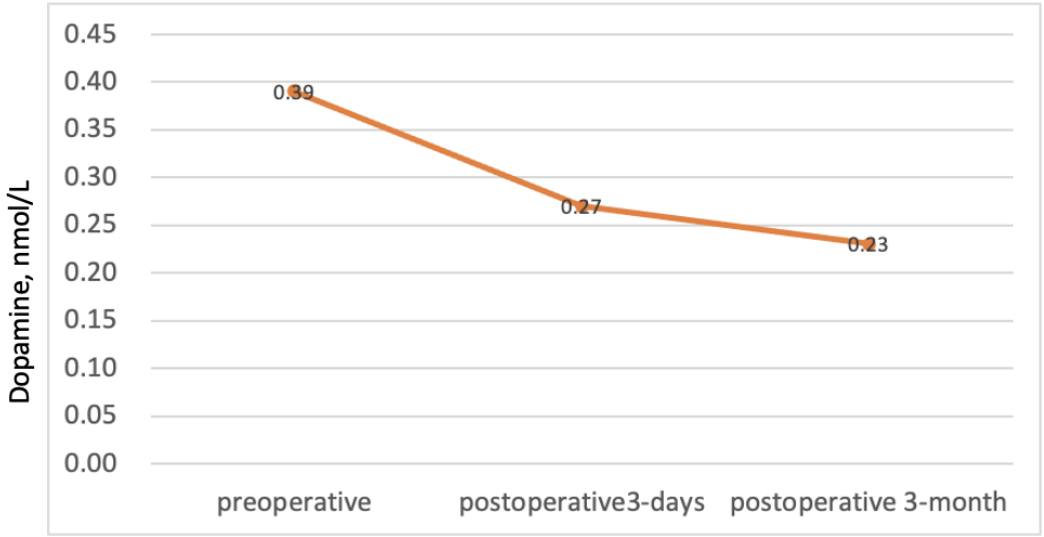

Fig-3: The Levels of Dopamine in Plasma Preoperative, Postoperative and Follow-Up 3-Months

Two days postoperatively, levels of metanephrines and catecholamines in the 24-hour urine sample were normal. Plasma levels of metanephrines and catecholamines were as follows: dopamine decreased to 0.27 nmol/L, while metanephrines, norepinephrine, and epinephrine remained within normal ranges. Follow-up examinations conducted three months after the surgery revealed that both plasma and urine levels of metanephrines and catecholamines had returned to normal (**Fig-3**).

Regular follow-up assessments were carried out after the surgery. One-year post-surgery, a mass was detected in the right cervical region. Surgical resection confirmed a paraganglioma, raising the possibility of metastasis from the right femur. As of now, the patient's overall condition remains favorable, and no new metastatic lesions have been identified.

Discussion

Paraganglioma is an exceedingly rare neuroendocrine tumor that originates from the chromaffin body outside the adrenal gland [1]. Its incidence rate is exceptionally low, and the occurrence of paraganglioma within bone is even rarer. In the course of our literature review, we came across only two case reports of femoral metastasis subsequent to Pheochromocytoma resection [4,5].

Functional Pheochromocytoma/paraganglioma is characterized by the development of progressive hypertension when stimulated. Pheochromocytoma /paraganglioma often arises without any evident triggers, and most patients are identified during hypertension screening or routine physical examinations. In this particular case, when presented with a right thigh mass subsequent to trauma, the initial considerations would likely encompass possibilities like hematoma or pseudoaneurysm. However, the unexpected diagnosis of a bone tumor was made through imaging modalities such as CT. Intriguingly, the act of touching the tumor during surgical planning led to a gradual rise in blood pressure. This observation greatly raised suspicion of functional Pheochromocytoma/paraganglioma. Following a comprehensive screening process, the diagnosis of paraganglioma was confirmed.

It is pertinent to discuss the differentiation between Pheochromocytoma and paraganglioma. While the former originates within the adrenal gland, the latter emerges from tissues situated external to the adrenal gland. Hence, in this specific case, our focus was on scrutinizing the patient's adrenal glands, revealing the absence of any space-occupying lesions and thus leading to a diagnosis of paraganglioma. Some patients afflicted with Pheochromocytoma/paraganglioma exhibit the classic hallmarks of hypertension, tremors, and elevated blood pressure. However, the patient under consideration did not manifest these typical signs, rendering differentiation challenging.

The prognosis for Pheochromocytoma /paraganglioma is generally favorable, with the primary determinant of poor prognosis being the presence of distant metastasis [6]. Consequently, regular postoperative follow-up plays a crucial role in preventing and managing the recurrence and distant spread of paraganglioma. Specific follow-up approaches encompass monitoring levels of fractionated metanephrines and catecholamines in plasma and urine, employing imaging techniques such as CT or T2-weighted magnetic resonance imaging (MRI), conducting limb X-ray photography, PET scans, and other relevant imaging examinations [7].

In the case of this patient, consistent postoperative follow-up was diligently carried out, leading to the identification of a cervical metastatic tumor one year after the initial surgery. Subsequent surgical resection has resulted in an absence of recurrence or metastasis during the follow-up period.

Conclusion

The implication drawn from this case highlights the possibility that bone tumors could potentially be paragangliomas. As such, it becomes crucial to distinguish this possibility during clinical diagnosis, especially for patients exhibiting symptoms of malignant hypertension, paroxysmal headaches, excessive sweating, tachycardia, and notably, progressive hypertension triggered by manipulation of the tumor during surgery.

In order to effectively address such scenarios, a

comprehensive approach is advised. This encompasses a thorough assessment of fractionated metanephrines and catecholamine levels in both plasma and urine, along with the utilization of imaging techniques such as CT, MRI, or PET scans. This meticulous preoperative evaluation not only aids in achieving an accurate diagnosis but also facilitates comprehensive preoperative preparation. This preparation, in turn, serves to reduce the potential occurrence of perioperative adverse events, thereby contributing to improved patient prognosis.

Conflict of Interest

The authors have read and approved the final version of the manuscript. The authors have no conflicts of interest to declare.

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