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Case Report

Fever as the Main Presenting Symptom of a Carotid Body Tumor

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Carotid body tumor (paraganglioma) is a rare tumor which presents as a mass in the lateral part of the neck. It is typically a slow-growing and nonsymptomatic mass at the beginning. Fever is not an usual symptom of this tumor. Here, we report a 78- year-old woman presenting with this tumor as a cause of fever of unknown origin.

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Introduction

Carotid body tumors (CBTs) (paragangliomas) are rare and originate from the neural crest. Paragangliomas may be seen in numerous sites. When the primary site is the adrenal gland, the tumor is called pheochromocytoma; in the head and neck area, paragangliomas are mainly found in the carotid body, jugular, and vagal glomus, among other sites. The manifestations of the tumor are variable; in most cases, it presents as a nonsymptomatic slow-growing mass, compressing the surrounding anatomic structures.[1] Malignant paragangliomas are very rare. The annual incidence in retrospective studies is 1/10,000,000.[2] Fever is one of the most infrequent symptoms of CBTs. Herein; we present a 78-year-old woman with fever as the main presenting symptom of a CBT.

Case Report

This 78-year-old woman developed attacks of fever, lethargy, and vertigo 20 days before admission. The fever had a peak of 38.8°C and a nadir of 37.5°C. She had a true vertigo more in sitting or standing up position. The patient had no accompanying symptoms such as cough, expectoration, headaches, and abdominal or musculoskeletal symptoms. Apart from a history of toxoplasmosis seven years before, for which she received prompt treatment, she had no history of any medical condition. Physical examination revealed an ill, feverish patient with a mobile, well-defined, hard, and nontender 3×4 cm mass in the right side of her neck under the sternocleidomastoid muscle with no accompanying lymphadenopathy. A mild to moderate bruit was heard on auscultation of the mass. The patient experienced an attack of severe bradycardia and decreased level of consciousness, when changing position from recumbent to sitting. No sign of Horner's syndrome was detected.

Examination of other organ systems was unremarkable.

Laboratory data

Thorough evaluations for fever of unknown origin were done. She had a leukocyte count of 11800/mm^3 with a left shift but no bandemia or eosinophilia. Her erythrocyte sedimentation rate (ESR) was 95 mm after one hour and C-reactive protein (CRP) was 2+ positive. Urinalysis and blood smear examinations were normal. Blood and urine cultures, Wright, Coombs, Vidal and PPD tests were all negative. Bone marrow biopsy was insignificant. Thyroid and adrenal function tests

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were within normal ranges. Serum protein electrophoresis and electrolytes were normal.

Chest X-ray revealed diffuse interstitial changes in both lung fields. These findings were also confirmed in the subsequent lung computed tomography (CT) scan. No pathologic change was evident in the brain magnetic resonance imaging (MRI) or abdominal ultrasonography. Echocardiography, Holter monitoring and coronary angiography, and abdominal and pelvic CT scan were all normal. Cervical MRI and digital subtraction angiography confirmed the presence of CBT (Figure 1). No evidence of otitis was found in tympanometry.

Open lung biopsy of pulmonary infiltration revealed diffused pulmonary fibrosis without any evidence of malignancy.

Treatment

Since our patient had multiple attacks of severe bradycardia accompanied with loss of consciousness, she underwent classic resection of the CBT. During the operation, a mass measuring 4×5 cm in the right carotid bifurcation was removed. No accompanying lymphadenopathy was detected during the operation. The pathology report was consistent with CBT (Figure 2).

The immediate postoperative period was uneventful and the surgical drain was removed on the third postoperative day. By the second postoperative day, the fever ceased, ESR and CRP decreased to normal, and the patient no longer had attacks of bradycardia or vertigo. The patient was followed for six months during which no fever was reported and ESR was normal.

Discussion

We described a patient with CBT. The earliest report of a carotid body paraganglioma was published in 1891.[1] CBTs occur commonly in adults aged 45 – 60 years and are unusual in children.[3–4] Women are susceptible more than men to some extent. Sporadic forms of CBT are more common. Familial forms account for 10% in most series, with bilateral tumors seen in 30% of

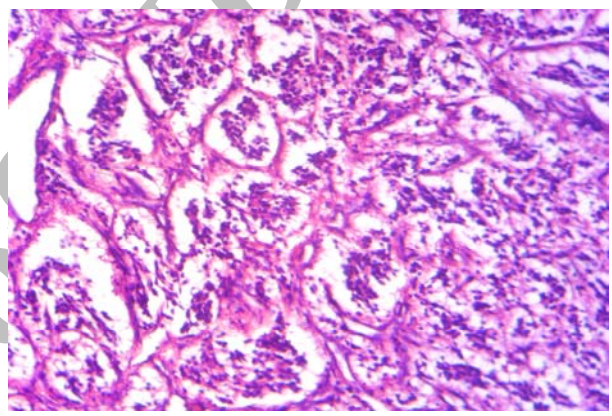
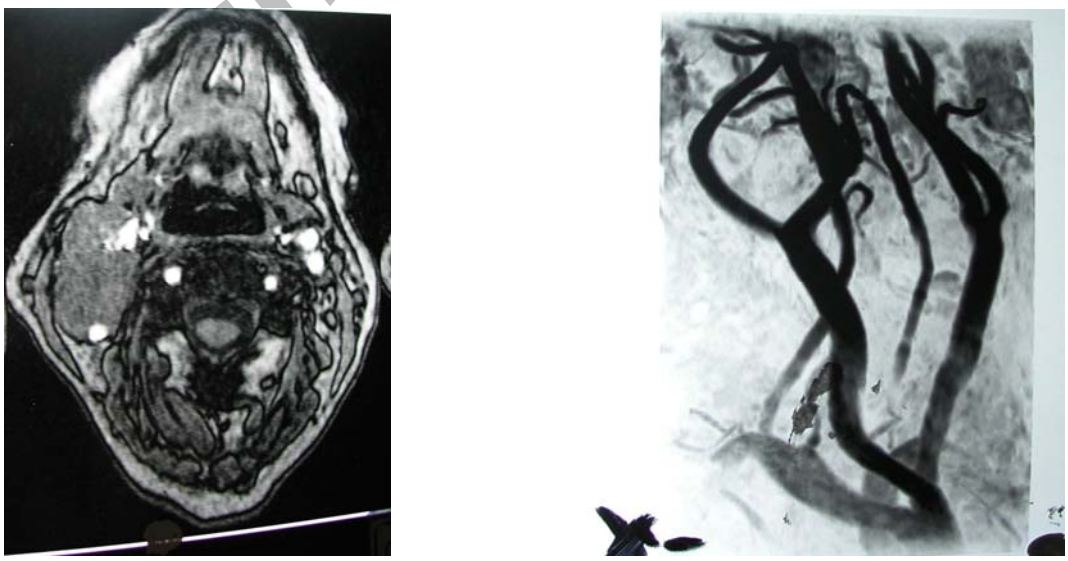


Figure 2. Well-defined nests of cuboidal cells (Zellballen) are separated by highly vascularized fibrous septa (H&E, x40).



Figures 1. Cervical MRI (left) and digital subtraction angiography (right), confirming the presence of a carotid body tumor.

patients. Multiple tumors are reported in 25 – 33% of patients with a family history of paragangliomas. [5]

As mentioned above, a paraganglioma may arise from a number of locations, usually associated with mesodermal branchial arches. Histopathologically, it is similar to the adrenal gland neoplasm, pheochromocytoma.[6] Paragangliomas are generally found in the carotid body, beside the nodose ganglion of the vagus nerve (glomus vagale), along the jugular ganglion of the vagus nerve (glomus jugulare), and about Arnold and Jacobson's nerve in the middle ear (glomus tympanicum). Carotid body and glomus vagale tumors typically present as neck masses, whereas the other forms of glomus tumors present as expansile masses within the skull base.

Occasionally, they originate within the abdomen, usually in the retroperitoneal space.[7]

Paraganglionic tissue is composed of two types of cells: type 1 or chief neuroectodermal cells, bearing catecholamine granules; and type 2 or sustentacular cells. There is significant cellular atypia within most tumors, and by histologic criteria, the majority of CBTs can be classified as malignant. The malignant potential of the CBT has been described in some series reports, and its incidence varies from extremely low up to 50% (mean: 10%).[8,9] Our patient had no confirmed invasion or metastasis before or during the surgery, or even during more than six months of follow-up.

Primarily, CBTs emerge in the anterior triangle of the neck as gradually growing, asymptomatic palpable lumps. These tumors tend to enclose the external and internal carotid arteries without any significant compression even with no treatment. The sympathetic chain and internal jugular vein may also be involved by larger tumors. Pain, hoarseness, dysphagia, Horner's syndrome, tongue paresis, and vertigo may be present due to the compression effect of the tumor on the carotid artery.[4] Functional CBTs are rare and can generate paroxysmal hypertension, mimicking a pheochromocytoma because of catecholamine release.[3,6] The classic triad of headache, diaphoresis, and palpitations, and other symptoms of a hyperadrenergic state such as hypertension, cardiac, gastrointestinal and metabolic manifestations are typical of a pheochromocytoma, being less apparent in a paraganglioma and rarely present in a malignant paraganglioma.[1,6] On physical examination, this tumor is laterally mobile but vertically fixed because of its attachment to the carotid bifurcation.[7] The mass may be pulsating with a bruit.

Fever is not a typical presentation of this tumor. Apart from only one report of fever accompanying CBT by Ramsdell et al.[10] in 1971, to the best of our knowledge, no other similar case has been reported. The patient was febrile. This observation suggests CBT as a cause of fever of unknown origin. The cause of fever may be due to the release of catecholamines from chief neuroectodermal cells in paragangliomas or tumor necroses. The fact that by removing the tumor fever subsided, emphasizes that the tumor was the cause of fever.

The diagnosis of a paraganglioma is based on imaging methods along with biochemical assays in cases that a functioning tumor secrets catecholamines. Angiography is an essential examination for the diagnosis of vascular commitment caused by the tumor. MRI is the gold-standard method for detection of CBT. It has a great accuracy in demonstrating the size and density of the tumor and its relation to the adjacent structures. The salt and pepper appearance characterizes paragangliomas on MRI.[11] Ultrasonography, though has limitations, is an important tool for the diagnosis of retroperitoneal and pelvic paragangliomas, but has less sensitivity for tumors located in other sites. Plain X-rays and CT scan are valuable diagnostic methods in detection of bone and vertebral metastases.

The most frequent expression of the malignant paraganglioma is the local recurrence even after total resection of the tumor.[12] The typical locations are cervical and mediastinal lymph nodes, bone, lung, liver, and heart. There are no risk factors regarding gender, race, or age. The mean time of recurrence after the primary resection is almost six years, so a long-term follow-up is mandatory for all patients.[13]

The success in treating benign and malignant paragangliomas is based on the early diagnosis and complete resection of the tumor using an adequate catecholamine blockage under good anesthesia.[14] For primary masses, the best surgical approach is complete resection with sufficient safety margins and examining the neighboring organs and vessels. The resection of the isolated secondary masses shows a better prognosis, as demonstrated on pulmonary metastasis of tumors of the head, trunk, and extremities.[15]

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