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Case Report

Combination of Cardiac and Carotid Glomus Tumour: a Rare Case Report

Feridoun Sabzi[1], Aghigh Heydari[1], Mohammad Rouzbahani[1], Reza Heidari Moghaddam[1], Atefeh Asadmobini[1]

[1] *Cardiovascular Research Center, Kermanshah University of Medical Sciences, Kermanshah, Iran*

Corresponding author: Atefeh Asadmobini, Atefeh Asadmobini, Shaheed Beheshti Ave, Cardiovascular Research Centre, Imam Ali Hospital, Kermanshah University of Medical Sciences, Kermanshah, Iran. Postal Code: 6715847145; Email: a.asadmobini@gmail.com; Tel.: 098 8338395970

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Abstract

The most common tumour of the heart is myxoma but paraganglioma (also called glomus tumour in extracardiac sites) in the cardiac position is the rarest of them. While this tumour accounts for 0.8% of all primary benign tumours, the combination of both neoplasms is an exceedingly rare occurrence. Herein, we present a case of combined carotid glomus tumour and left atrial paraganglioma tumour in which respiratory distress was the presenting symptom of cardiac type but carotid tumour was asymptomatic. The case underwent a two-step resection of the neck and cardiac mass with an uncomplicated postoperative course and in the 1year follow-up, no recurrence of tumour in both sites was found on physical exam and imaging studies.

Keywords

case report, glomus tumour, heart neoplasms, paraganglioma

INTRODUCTION

Myxoma is the most common tumour of the heart and paraganglioma in the cardiac position is the rarest of these tumours.[1] This tumour is known as glomus tumour in extra cardiac sites. While this tumour accounts for 0.8% of all primary benign tumours, a combination of both neoplasms is an exceedingly rare occurrence. Carotid and cardiac tumours are rare in the majority of cases but constitute most of the head and neck paragangliomas.[2] The neoplasm present as asymptomatic neck mass. Herein, we present a case of combined carotid glomus tumour and left atrial paraganglioma tumour in which respiratory distress was the presenting symptom of cardiac type but carotid tumour was asymptomatic.

CASE REPORT

A 43-year-old woman was referred to our center with a new onset of dyspnea (grade 2 Medical research council), which was temporary and exacerbated by walking. She did not report any similar respiratory problems in the past medical history. Physical examination showed normal temperature, a rate of 100 beats/min, and a respiratory rate of 19 breaths/min. No others finding were detected during cardiac exam, except for a cardiac diastolic murmur. She also complained of a mass in the left side of the neck that had progressive growth in the last 6 months. This neck mass was not painful and had a well-defined border but had a pulsatile nature in palpation. There was no familial history of paraganglioma. On physical examination, her neighbouring cranial nerves had a normal function. The left side of her neck had a 4×4 mass palpable in the region of the left carotid bulb. The mass was immobile and did not have a well-defined border. Color flow Doppler

revealed a large tumour of approximately 3×4 cm over the left carotid artery bifurcation. Physical findings of the chest were unremarkable and respiratory sound was normal as other findings in the abdomen and extremities. The electrocardiogram exhibited a normal sinus rhythm of 100 beats/min, which changed to temporary atrial fibrillation, while no ST-T wave changes were found. Specific cardiac enzymes were within normal values: troponin=0.05 (<0.1), CK-MB=4 (0-9 U/l), LDH=110 (100-200 U/l), myoglobin=70 (<85 ng/ml). The chest radiograph was normal. Transthoracic echocardiography (TTE) revealed a large mass in the left cardiac atrium ***(Fig. 1)****. This mass temporarily obstructed the left atrium and lead to dyspnea. Computed tomography of the neck showed an avascular mass in the bulb of the carotid. Laboratory exams such as complete blood count, blood urea nitrogen (BUN), creatinine, urine metabolites of adrenaline, thyroid and liver function tests were unremarkable. The patient underwent a two-step surgery to remove both masses. After median sternotomy and aortic and bicaval cannulation, ascending aorta was cross-clamped (clamp time=70 minutes) (CPB time=110 minutes), and cardioplegia infused to aortic root and with cardiac arrest. Left atrial access was obtained from the interatrial groove. To better access the mass, right and left atrium were simultaneously opened. The mass was detected to originate from the interatrial septum of the left atrium. It was not pedunculated and extended into mitral valve apparatus during cardiac systole. The tumour had many feeding arteries running along the interatrial septum. Complete removal was achieved with a safe margin. The left interatrial septal defect was repaired with the fresh pericardium. After a diagnosis of carotid body tumour and removal of the left atrial mass, the patient underwent surgical resection of the neck mass within the periadventitial layer with excision of one jugular lymph nodes specimen for pathology* ***(Figs 2A, 2B)****. The patient's postoperative course was normal and she was discharged 7 days postoperatively. The postoperative courses of both operations were unremarkable. In the follow-up with examination and imaging, recurrence of the tumour in both locations was not detected. The histologic exam of both cardiac and neck masses confirmed the diagnosis of benign glomus tumour as well as the negative of the lymph nodes* ***(Fig. 3)****. A frozen section examination of the resected tumour of the neck revealed a vascular tumour. Hematoxylin and eosin staining showed assembling of polygonal cells that contained oval nuclei, some with eosinophilic cytoplasm, all of which characteristics were consistent with glomus or paraganglioma tumour. Immuno and histochemical exam of the mass tested was positive for chromogranin, and synaptophysin and CD57 S-100 protein was found an adventitial cell layer surrounding the neoplastic cells. In the postoperative course, the serum levels of metanephrine and normetanephrine were unremarkable. On day 2 postoperatively, she was successfully weaned from the ventilator and discharged 7 days after operation. She was in good condition at 6 months after surgery, and the one-year follow-up imaging showed the intact postoperative structure of the neck and no recurrence of the tumour.*

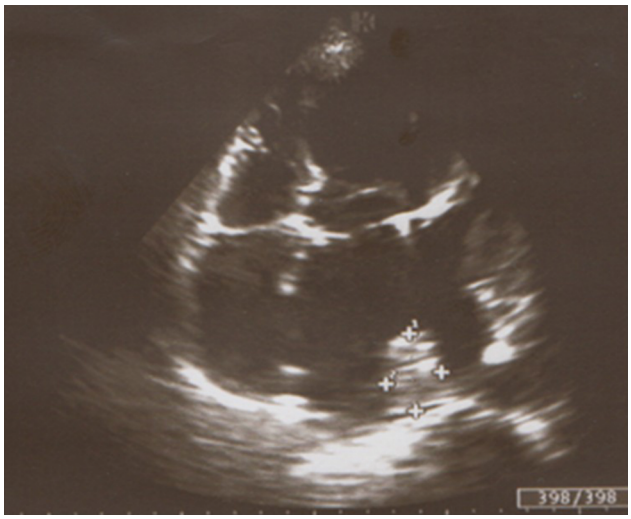
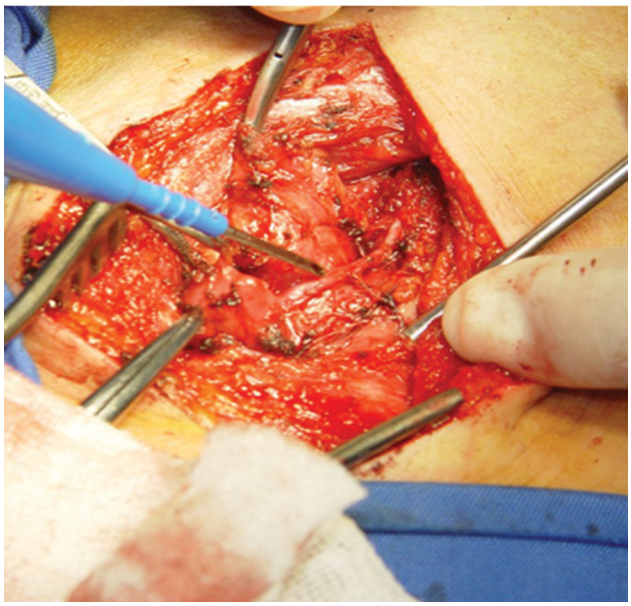
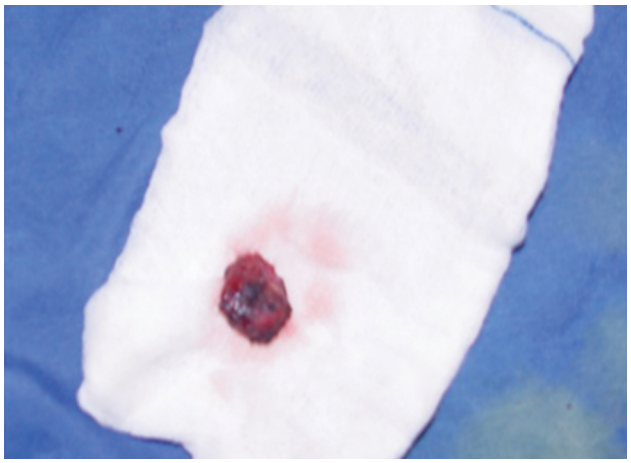


Figure 1. Transthoracic echocardiography shows left atrial mass.



A



B

Figure 2. **A**. shows surgical resection of left carotid bulb glomus; **B**. shows gross view of resected glomus mass.

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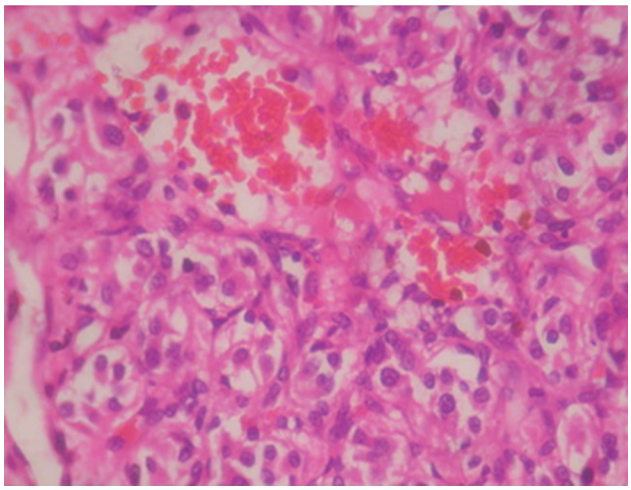

Figure 3. Photomicrograph showing nests of uniform bland polygonal cells with no mitotic activity, and vascular septa (H&E stain, ×200).

DISCUSSION

Most heart neoplasms are secondary to metastasis from an extra cardiac origin, and these metastatic tumours are approximately 20-30 times more common than benign primary tumours.[3] Neurilemoma, paraganglioma or chemoductoma or glomus is a different name for a tumour with a neural crest origin as a rarest tumour both in cardiac or neck position. The most common site of origin of a neural crest tumour is the adrenal glands. About 15% of the neural crest is detected in extra adrenal glands. The common sites for such mass include the periaortic sites and the sympathetic neural network of retroperitoneum.[4] Anterior mediastinal neural crest tumours could be located either intracardially or located in the extracardiac position. The most common sites of intracardiac neural crest is in the left atrium, interatrial septum followed by left ventricle cavity, anterior surface of the left and right ventricle, and right ventricular outflow tract. Our case has a combination of carotid glomus tumour and left atrial paraganglioma tumour. Cardiac tumour has a respiratory distress presenting but carotid tumour is asymptomatic. To the best of our knowledge, no previous cases of combined left atrial and carotid neural crest have been reported in the English medical literature.[5-8] Cardiac clinical sign and symptoms are variable and depend on the position of the neoplasm in relation to cardiac chambers and structures. Intrachamber neoplasm may present with positive signs and symptoms of valvular pathology, dyspnea, or faint. In addition, symptoms may be related to epinephrine secreted by the mass. Tumours that involve the carotid artery can present as hypersecretory syndrome, or sign of compression of neck nerves or in malignant form, causing an invasion to sheath of nerve. Rarely, neoplasm compress the carotids vessels and present with features of strokes.[7,8] Asymptomatic presentation of carotid neural crest tumour is rare, and diagnosed commonly with the aid current imaging, including computed tomography, magnetic resonance imaging, and MIBG (iodine-131-meta-iodobenzylguanidine) scanning. Treatment of an intracardiac mass is by open-heart resection and if malignant, it could be a complicated operation its outcome depending on the position and relation of the tumour to the surrounding structures.

CONCLUSIONS

This case report shows a rare asymptomatic presentation of neck neural crest tumour combined with symptomatic left atrial mass as dyspnea, followed by uncomplicated surgical removal. Presumed causes of the dyspnea and chest pain include temporary obstruction of the mitral valve or stretching of the left atrial wall by the mass. Our patient's postoperative outcome was uncomplicated, and her mass was nonsecreting, and serum metanephrines was normal.

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Competing Interests

The authors have declared that no competing interests exist.

Author contributions

All authors contributed equally to the development of the original idea, and abstracted, prepared, and revised the manuscript.

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Сочетание кардиальной и каротидной гломусной опухоли: отчёт о редком случае

Феридун Сабзи[1], Агиг Хейдари[1], Мохамад Рузбахани[1], Реза Хейдари Могадам[1], Атефех Асадмобини[1]

[1] *Центр сердечно-сосудистых исследований, Керманшахский университет медицинских наук, Керманшах, Иран*

Адрес для корреспонденции: Атефех Асадмобини, Центр сердечно-сосудистых исследований, Керманшахский университет медицинских наук, ул. „Шахеед Бехещи“, Больница „Имам Али“, 6715847145 Керманшах, Иран; E-mail: a.asadmobini@gmail.com; Тел.: 098 8338395970

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Резюме

Наиболее распространённой опухолью сердца является миксома, но наиболее редкой из них является параганглиома (называемая также гломусной опухолью в экстракардиальных локализациях) в области сердца. Хотя эта опухоль составляет 0.8% всех первичных доброкачественных опухолей, сочетание обоих новообразований встречается крайне редко. Здесь мы представляем случай комбинированной опухоли каротидного гломуса и опухоли параганглиомы левого предсердия, при которой респираторный дистресс был симптомом кардиального типа, но опухоль сонной артерии была бессимптомной. В данном случае выполнена двухэтапная резекция шеи и сердечной массы с неосложнённым послеоперационным течением, и в течение 1 года при физикальном осмотре и визуализационных исследованиях не было обнаружено рецидива опухоли в обеих локализациях.

Ключевые слова

клинический случай, гломусная опухоль, новообразования сердца, параганглиома