Aortic paraganglioma requiring resection and replacement of the aortic root

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Abstract

This case report demonstrates the resection of an aortic root paraganglioma incidentally discovered in a 32-year-old male with replacement of the root for an oncologically complete resection.

Keywords: Paraganglioma; Cardiac tumor; Aortic root

1. Introduction

Cardiac paragangliomas are rare entities, constituting <5% of all cardiac tumors. These tumors typically occur in young individuals and tend to remain asymptomatic until discovered incidentally or grow to a size large enough to cause symptoms. Resection is often technically demanding due to their location. We present one such case of aortic root paraganglioma that required an aortic root replacement.

2. Clinical summary

A 32-year-old male was admitted to another hospital after a motorcycle accident with clavicular fracture. As part of the trauma workup, a CT-scan of the chest was performed that revealed a cardiac tumor that involved the aortic root (Fig. 2). A cardiac catheterization was performed which revealed a tumor blush from both the right and left coronary arteries (Fig. 1a,b). A percutaneous biopsy of the mass was performed that revealed a diagnosis of paraganglioma. The patient had significant intrapericardial hemorrhage from the biopsy that was managed with a pericardial drain and resolved without any further intervention. The patient was referred to our institution for further management. An echocardiogram was performed that revealed an aortic anulus of 24 mm and the appropriate sized aortic homograft was ordered. The choice of using a homograft was based on preoperative discussions with the patient regarding the use of postoperative anticoagulation. The patient had a very active outdoor life that he wanted to preserve, which would place him at a high risk for anticoagulation.

A median sternotomy was performed. Cardiopulmonary bypass was initiated with distal ascending aortic cannulation and bicalveal venous cannulation. Antegrade cardioplegia through the distal ascending aorta and retrograde cardioplegia via a catheter in the coronary sinus were given. A sump catheter was placed in the left atrium through a purse-string suture in the right superior pulmonary vein. The thymus and mediastinal lymph nodes were excised. The tumor was dissected away from the distal ascending aorta. One centimeter beyond the sinotubular junction the tumor was firmly adherent to the aortic wall, suggesting invasion, the aorta was transected at this point (Fig. 1c). The tumor extended around the base of the aorta, extending posterior to the junction of the right main pulmonary artery (PA) and the main pulmonary artery. In order to better expose this area, the PA was transected just proximal to its bifurcation. The tumor was then dissected off the PA and the left mainstem bronchus. This maneuver also facilitated the dissection of the left main coronary artery, which was mobilized as an ostial button. The right coronary ostium was also mobilized as a button. The aortic valve leaflets and the non-coronary sinus of valsalva was excised, which enabled the complete excision of the tumor with the attached aortic wall (Fig. 1d). A 24-mm aortic homograft was used to replace the aortic root with re-implantation of the coronary ostial buttons. The postoperative course was uneventful with the patient discharged to home on postoperative day 7.

3. Discussion

Paragangliomas are tumors arising from extra-adrenal chromaffin tissue. These have been classified based on histology, location and innervation into brachiomeric, intravagal aortico-sympathetic and visceral-autonomic groups. Aortic body paragangliomas are included in the brachio-
meric group [1]. This distinction is important as the brachio-
meric group typically has fewer metastases and presumably
better prognosis, justifying a more aggressive resection. While most patients with aortic body paragangliomas are
asymptomatic, some patients present with angina or myoc-
cardial infarction due to compromise of the coronary cir-
culations [2, 3]. The mainstay of therapy for these tumors
is complete surgical resection [4].

The case that we have presented here highlights several
issues in the management of these tumors. First, a high
index of suspicion is important for diagnosis as these tumors
are often misdiagnosed clinically as lymphomas or cardiac
sarcomas. Second, these tumors are very vascular and
percutaneous biopsy can lead to life threatening hemor-
rage. Therefore, open biopsy with a plan to resect is a
safer approach. Preoperative embolization to decrease the
vasularity of this tumor has also been described [5]. Third,
proper preoperative planning is crucial as resection can
sometimes be technically challenging. As we have described
elsewhere, resection may entail complex maneuvers,
including cardiac auto-transplantation [6]. In this case,
preparations were made for replacement of the aortic root,
including having available an appropriate sized aortic
homograft.

We conclude that cardiac paragangliomas of the aortic
root can be safely resected using well established surgical
techniques with appropriate surgical planning.

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