Human Cardiac Explantation and Autotransplantation: Application in a Patient with a Large Cardiac Pheochromocytoma

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A rare paraganglioma of the chromaffin type, located intrapericardially, was removed from a 42-year-old man by autotransplantation of the heart. Although the results were unfavorable in this case, the technique of autotransplantation deserves consideration in resecting large tumors that encroach upon the heart. To our knowledge, this technique has not been previously used for surgical excision of cardiac tumors.

PRIMARY CARDIAC tumors are infrequent and, until the advent of cardiopulmonary bypass (CPB), they were mostly nonremovable. Tumor resection with the use of CPB was first reported by Crafoord in 1954.[1] From 1961 until 1983, 20 primary cardiac tumors, exclusive of myxomas, were operated upon at the Texas Heart Institute.[2] At this institute during the same period, a total of 52,500 cardiac operations were performed using cardiopulmonary bypass, emphasizing the rarity of these tumors. Ten tumors were seen in children, and all were benign. Of the ten seen in adults, five were benign. Complete excision in benign lesions usually results in permanent cure and relief of symptoms. Although not uniformly possible, complete excision of maligant lesions should alleviate symptoms and offer a chance for cure.

Paraganglioma of the chromaffin type (pheochromocytoma) is rare in the intrapericardial location. Only three such tumors were reported in the world literature by Saad and Frazier in 1983.[3] A patient with a left atrial pheochromocytoma confirmed by biopsy was referred to the Texas Heart Institute for a more aggressive surgical approach after the tumor could not be removed by conventional techniques in an operation elsewhere.

CASE REPORT

A 42-year-old man underwent excision of a left carotid paraganglioma and saphenous vein interposition graft reconstruction in Italy in 1972. This tumor had presented as an asymptomatic mass, and no sequelae followed its removal. Because of arrhythmias (ventricular and supraventricular) and a systolic murmur at the apex, the patient was followed by a cardiologist. Retrospective examination of serial M-mode echocardiograms (done since 1976) showed evidence of a left atrial mass occupying a large part of the atrium. This was only recognized in 1984. Operation was then undertaken in Italy, with the clinical diagnosis of left atrial myxoma. Using cardiopulmonary bypass, the left atrium was explored through the interatrial groove. Because of the extensive size of the tumor and involvement of the posterior left atrial wall, only biopsy was

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performed. Examination of the biopsy showed a paraganglioma. Profuse bleeding complicated the procedure.

The patient was subsequently referred to the Texas Heart Institute for evaluation and consideration of radical surgical removal of the tumor. Physical examination revealed an apical systolic murmur, suggesting mitral regurgitation. An echocardiogram showed a mass occupying a great part of the left atrial cavity. Cardiac catheterization demonstrated a large left atrial mass and moderate mitral regurgitation with an atrial V wave of 36 mm Hg. After full explanation of the problem with the

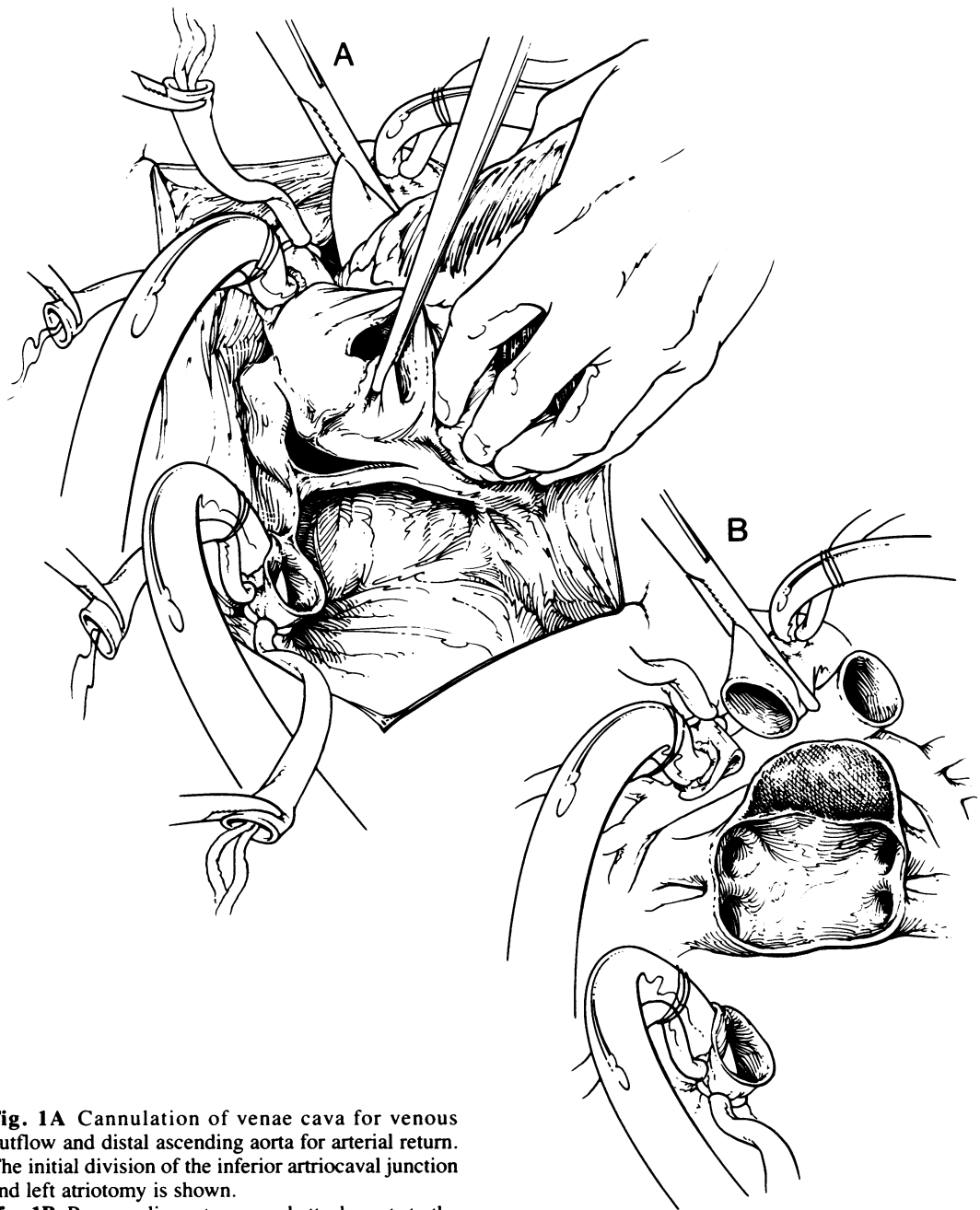

Fig. 1A Cannulation of venae cava for venous outflow and distal ascending aorta for arterial return. The initial division of the inferior artriocaval junction and left atriotomy is shown.
Fig. 1B Paraganglioma tumor and attachments to the left atrium and transverse pericardial sinus.

patient and with his full consent, operation was performed on February 4, 1985.

PROCEDURE

The previous median sternotomy was entered. Direct cannulation of both cavae and the distal ascending aorta, as previously described for cardiac transplantation, was performed.[4] Cardiopulmonary bypass was begun with a bubble oxygenator and nonblood prime. Systemic hypothermia (20° C) and cold crystalloid potassium cardioplegia were utilized for myocardial protection after aortic cross-clamping. Initially, the apex of the heart was elevated, the inferior right atrium divided beyond the inferior vena cava cannula, and the left atrium entered behind the heart (Fig. 1A). This allowed excellent exposure of the entire

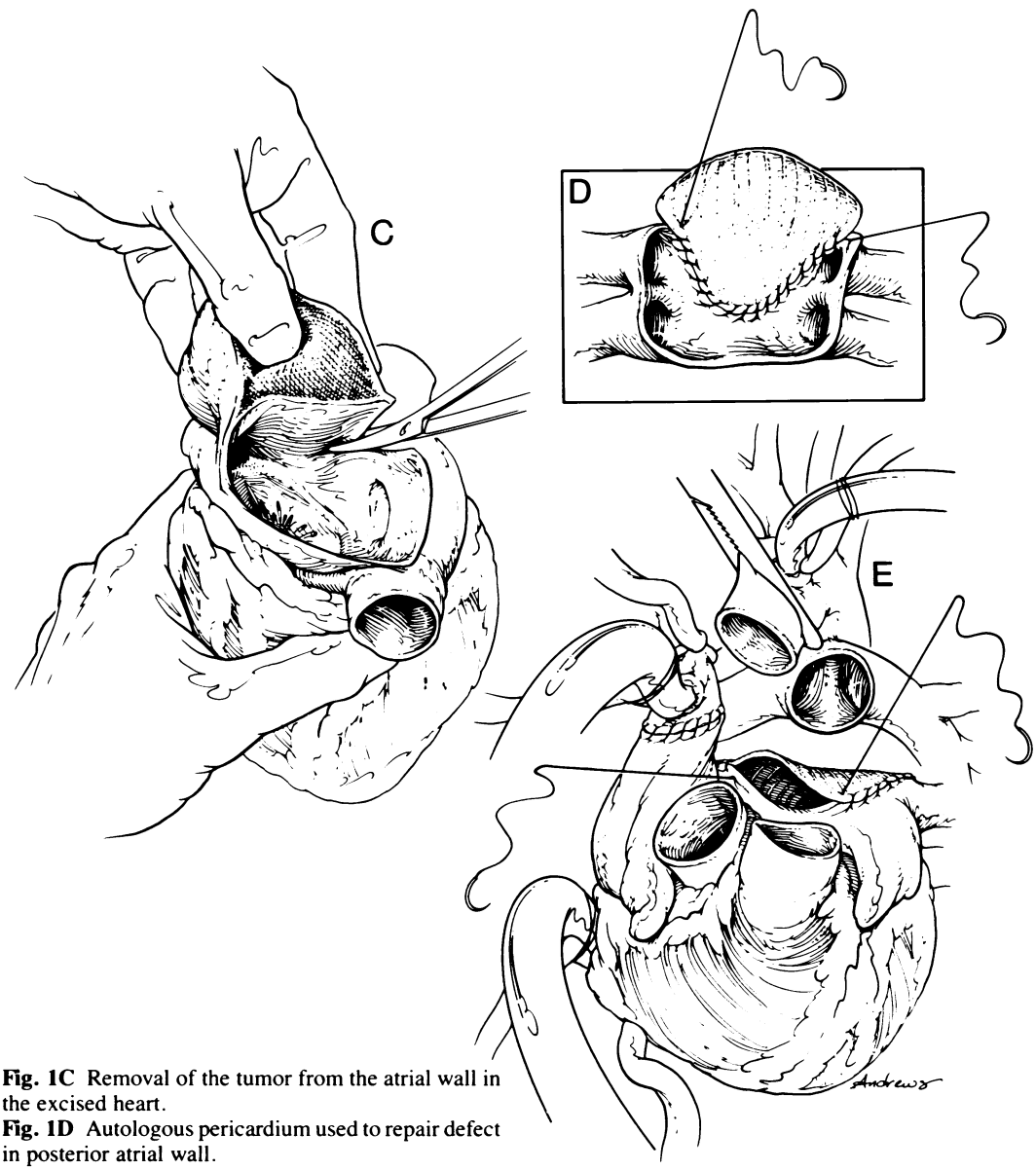

Fig. 1C Removal of the tumor from the atrial wall in the excised heart.
Fig. 1D Autologous pericardium used to repair defect in posterior atrial wall.
Fig. 1E Reimplantation technique with repair of left atrium and atriocaval attachments. Final anastomosis of main pulmonary artery and aorta completed the procedure.

posterior left atrial wall. Because of the extensive size of the tumor and its extension superiorly into the transverse sinus space, this approach proved inadequate. Subsequently, the aorta distal to the coronary arteries, the main pulmonary artery, and the high right atrium inside the superior vena cava cannula were divided and the heart removed (Fig. 1B). The portion of the left atrial wall that contained tumor was excised, and no involvement of the mitral valve was present (Fig. 1C). No further evidence of tumor was found in the excised heart, and it was placed in a bath of iced saline. The tumor, which measured approximately $5 \times 6 \times 6$ cm, involved the posterior left atrial wall superior to the pulmonary veins, extending into the transverse sinus, and was easily approached (Fig. 1B). Complete excision was complicated by an extremely vascular bed. Reimplantation of the heart was done using approximately the same technique as for cardiac transplantation. A flap of pericardium was used to expand the posterior and superior left atrial wall near the right superior pulmonary vein and transverse sinus (Figs. 1D and E). The low and high right atrial division points were anastomosed with continuous polypropylene sutures. The great vessels were then anastomosed. The aortic cross-clamp was removed, and air was aspirated from the ventricles as the patient was rewarmed to 37.5° C. Spontaneous cardiac activity resumed, and the patient was weaned from cardiopulmonary bypass without difficulty. Caval canulae were removed. Despite reversal of heparin with Protamine Sulfate, excessive bleeding from the rich vascular network, mainly originating from the posterior mediastinum, was encountered, and full control was not possible. Despite multiple blood transfusions, blood loss led to ultimate circulatory failure.

Pathologic examination of the tumor revealed a highly vascular paraganglioma that proved to be catecholamine-producing by electron microscopy. Both thick-walled and sinusoidal vascular spaces were observed (Fig. 2).

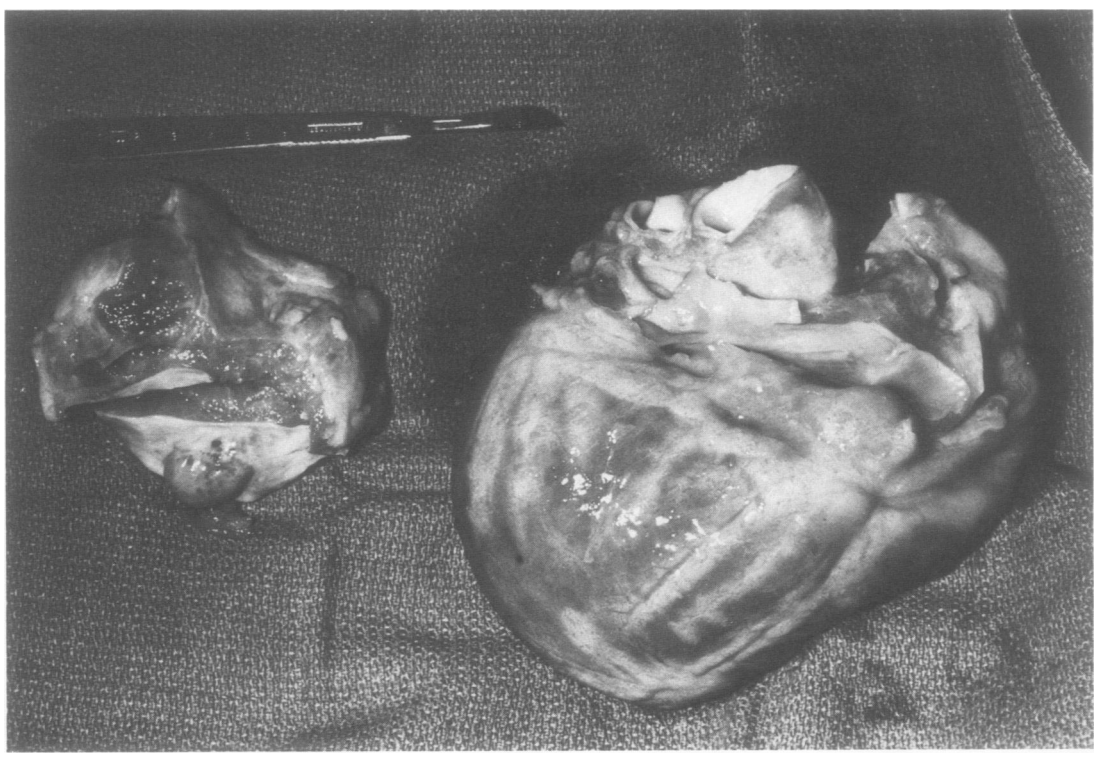

Fig. 2 Photograph showing the excised pheochromocytoma (paraganglioma) and the explanted heart.

DISCUSSION

Paragangliomas may be divided into chromaffin (pheochromocytoma) and nonchromaffin (chemodectoma) tumors, based on the presence of catecholamine secretion (pheochromocytoma) or its absence (chemodectoma). Both tumors are members of the APUD (amine precursor uptake and decarboxylation) system. These tumors arise in the mediastinum from the chemoreceptor tissue around the aortic arch, the aorticosympathetic chains, and the vagus nerve.[5]

The tumor in this case was a norepinephrine-producing tumor, and was therefore in the pheochromocytoma class. Mediastinal pheochromocytomas occur most commonly in men between 20 and 40 years of age.[5] Although the tumors are hormonally active, they tend to be clinically silent. Unfortunately, the histologic picture is an unreliable predictor of malignancy, which must be determined on the presence of metastasis and the clinical course of the tumor.[6] The incidence of malignancy is approximately 30% in extraadrenal locations.[6] Since intrapericardial pheochromocytomas are exceedingly rare[3] and the histological appearance is unreliable as a determinant of malignancy, this tumor should be considered to be biologically benign. A complicating feature of the intrapericardial position is the mechanical distortion of the heart, as evidenced by left atrial obstruction and mitral regurgitation in this patient. Since no other means of cure or control was possible, surgical resection offered the only hope of cure.

Autotransplantation of the heart in the laboratory has revealed satisfactory function despite disconnection from lymphatics and autonomic nerve supply.[7-10] Technical experience gained in laboratory and clinical cardiac transplantation indicates that the removal and reimplantation of a heart can be accomplished clinically. Indeed, successful human autotransplantation in the treatment of Prinzmetal's angina has been reported.[11,12]

In our case, an initial attempt was made at operation to avoid totally explanting the heart by dividing the inferior vena cava right atrial junction and the left atrial wall. The heart was then displaced superiorly and anteriorly on a pedicle of superior vena cava and great vessels. This resulted in good exposure of the posterior left atrial wall; however, it was inadequate for this tumor, which was located superior to the pulmonary veins and was larger than others previously described.[3] The heart was then removed and the tumor easily approached. A plastic repair on the left atrium with an autologous pericardial patch was used to replace the excised atrial wall. Weaning from cardiopulmonary bypass was accomplished without problems. Persistent bleeding from the highly vascular bed led to the patient's death. Although the final outcome of this, our first case of cardiac autotransplantation, was unfavorable, we believe the technique of autotransplantation deserves consideration in resecting large tumors which encroach upon the heart. To our knowledge, this technique has not been used previously or reported for surgical excision of cardiac tumors.

ADDENDUM

After this article was submitted for publication, an interesting report from Orringer and associates was published in the *Journal of Thoracic and Cardiovascular Surgery* (89:753-757, 1985) entitled, "Surgical Treatment of Cardiac Pheochromocytomas." Four cases were presented, and the authors stated that their increasing experience with the diagnosis and management of cardiac pheochromocytomas suggest that thoracic surgeons will be called upon to treat them more often in the future. We concur, and hope that our technique will be of some use.

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