Extra-adrenal pheochromocytoma involving the left atrium: An infrequent pathologic occurrence

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Cardiac paraganglioma is extremely infrequent. About 20 cases have been reported, some of them more than once. Our patient presented with the clinical signs, symptoms, and biochemical evidence suggestive of pheochromocytoma. Computed tomography showed a tumor situated above the left atrium that was successfully removed by means of cardiac surgery.

Clinical Summary

A 37-year-old woman experienced hypertensive crisis while being anaesthetized for cholecystectomy. An anterior mediastinal mass, compatible with pheochromocytoma, was revealed by means of computed tomographic scanning and vanillylmandelic acid determinations.

Mediastinal sternotomy was performed to reach a theoretic anterior mediastinal mass. However, during the operation the mass was found to be embedded in the left atrium, extending into the wall of the right upper lobar segmental arteries. The only way to remove the tumor was by means of rear total left atrial exeresis followed by substitution with a large polytetrafluoroethylene* patch (10 × 12 cm) and resection of the pulmonary affected arteries (Figure 1). The technique was performed with cardiopulmonary bypass (CPB).

The postoperative course was uneventful, and the patient was discharged 11 days later after echocardiographic confirmation of normalized left atrial function. At 6 months, echocardiography showed complete patch endothelialization, and oral anticoagulation was suspended.

Histologic findings included tumor node formation with complete delimitation by a connective capsule. Microscopic findings showed organoid clustering of neoplastic cells forming a “zellballen.” A sustentacular cell population can be demonstrated by staining for S-100 protein at the periphery of the nests. Tumor cells were oval or polygonal with granular cytoplasm. This cytoplasm presented strongly positive immunoreactivity for chromogranin A. We observed tumor cells connecting with the external muscular wall of the left atrium (Figure 2).

Discussion

Since the beginning of this century, there have been descriptions of paraganglia in various sites near the base of the heart and great vessels.¹ This subset of paraganglioma might require special surgical management techniques, such as CPB with cardioplegia² and even human cardiac explantation and autotransplantation when the paraganglioma is large.³ In our case the aortopulmonary paraganglioma arises from the paraganglia, which might be intracardial, usually at the level of the atria, or extracardial. These tumors account for about 1% of all primary mediastinal neoplasias.⁴

Occasionally cardiac paragangliomas are associated with paragangliomas in other sites. The incidence of malignancy with metastasis has been estimated at 13%.⁵

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Figure 1. Resection of total rear atrium and substitution with a polytetrafluoroethylene patch.

Surgical resection might necessitate CPB with removal of portions of the heart affected by the tumor, as in our case, and intraoperative deaths have occurred because of excessive bleeding.

This case was exceptional for the complexity of the surgical technique, the semispherical patch modeling, and the normalization of left atrial function without loss of sinusal rhythm.

Figure 2. Histologic findings typical of paraganglioma.

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