Case Report

Intrapericardial paraganglioma

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ABSTRACT

We report a case of intrapericardial paraganglioma accidentally discovered during surgery for mixed mitral valve disease. The 6 cm tumor was located in the aorta-caval groove, adherent to the roof of the left atrium and compressing the superior vena cava. A biopsy of the mass established the histological diagnosis of a non pressor secreting paraganglioma. The tumor was resected 2 months after the original mitral valve surgery. The patient made an uneventful recovery and remains well 2 years after surgery.

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Paragangliomas are rare neoplasms of neural crest origin. They can be chromaffin paragangliomas (pheochromocytomas) and mostly occur in the adrenal medulla. Nonchromaffin paragangliomas normally do not secrete pressor substances.[1] The latter are widely dispersed throughout the body but they are more commonly located in the anterior mediastima and costovertebral grooves. Cardiac paragangliomas are extremely rare.[2,3] In the pericardial space, they are also rarely seen. They can be an uncommon cause of superior vena caval obstruction. A case of intrapericardial non-chromaffin paraganglioma causing superior vena caval compression was described. It was accidentally discovered during mitral valve surgery. The mass was successfully resected without complications.

Case Report. A 32-year-old woman known with rheumatic heart disease presented with New York Heart Association class III exertional dyspnea. Physical examination revealed a diastolic and a systolic mitral valve murmurs on auscultation consistent with mitral valve stenosis and regurgitation. There were no signs of heart failure. There was no lymphadenopathy, distended neck veins or neck swelling. The chest x-ray demonstrated enlarged left atrium. The electrocardiogram (ECG) showed atrial fibrillation. An echocardiogram revealed moderate mitral valve stenosis and severe regurgitation. The posterior leaflet of the mitral valve was restricted in motion, however, the anterior leaflet was pliable and with no evidence of calcification. The subvalvar apparatus was thickened. There was no significant aortic or tricuspid valve disease. On the basis of these findings, mitral valve surgery was indicated. The patient underwent the usual preoperative preparation. The chest was entered through a standard median sternotomy incision. Following pericardiotomy a 4x6 cm solid and highly vascular tumor was discovered in the groove between the aorta and the superior vena cava on top of the left atrium and the right pulmonary artery. A frozen section biopsy was inconclusive. Further biopsies were taken for

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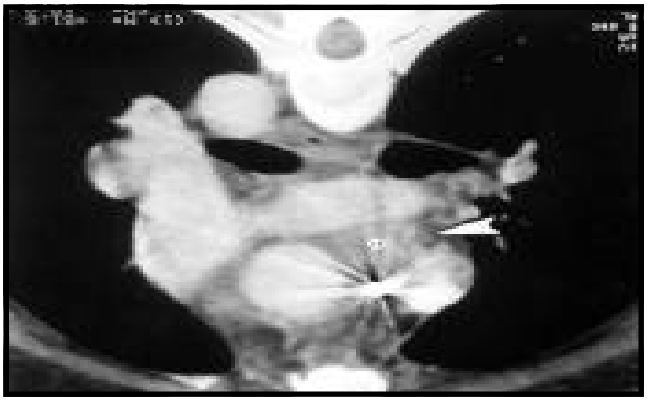

Figure 1 - A computerized tomography scan of the chest demonstrating tumor (arrow).

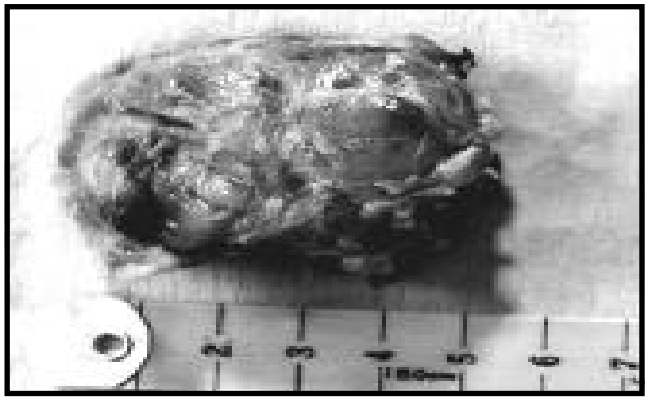

Figure 2 - Gross specimen of the resected tumor.

permanent sections and it was decided to leave the mass at that stage and proceed with the mitral valve surgery. On cardiopulmonary bypass and with cardioplegic cardiac arrest the mitral valve was approached through a left atriotomy in the interatrial groove. The valve was successfully repaired by commissurotomy, mobilization of subvalvar apparatus and a Carpentier ring. The post operative recovery was uneventful. The studies on the tumor biopsies including immunohistopathology revealed a benign non pressor secreting (nonchromaffin) paraganglioma. A computerized tomography (CT) scan of the chest defined a solid, vascular tumor in aorta-caval groove **(Figure 1)**. Due to the surrounding structures being compressed by the tumor, it was elected to go back 2 months later and resect the tumor. At the operation, the tumor was found very adherent and dissection resulted in laceration to the left atrium and right pulmonary artery. Cardiopulmonary bypass was instituted through femoral vessels cannulation. The tumor was then completely resected with a small segment of the left atrial wall **(Figure 2)**. The left atrium and right pulmonary artery lacerations were repaired. The patient was successfully weaned from cardiopulmonary bypass and thereafter made an unremarkable recovery. She remains well, asymptomatic and with no evidence of recurrence 2 years after her surgery.

Discussion. Mediastinal paragangliomas arise from paranganglia of the pulmonary artery and the aortic arch (anterior mediastinal tumor) or from paraganglia associated with the sympatheric chain (posterior mediastinal tumor). These are uncommon tumors and less than 100 cases have been reported in the English literature.[4] Fifty percent of patients are asymptomatic.[4] Malignant behavior is found in 10% of all paragangliomas and there are no definite microscopic criteria to distinguish the benign from the malignant form. The lesions are often discovered on routine chest x-ray that was indicated for other reasons, or during cardiothoracic surgery as in this case report. In the majority, the patients are more than 40-years-old. They are more common in women. A familial tendency has been noted. Our patient was a 32-year-old female and required a mitral valve repair which permitted an unexpected and early discovery of the tumor. Symptoms when present are secondary to local mechanical or compressive effects of the tumor. These patients present with hoarseness, dysphagia or non-specific chest discomfort.[5] Only 4 cases of a vena caval obstruction have been described among the 90 cases of mediastinal gangliomas reported in the literature.[6,7] Hence, despite the proximity of the tumor to the superior vena cava, signs of compression are rare. The heart is an unusual site for this tumor.[2,3] Extension to the right atrium has also been noted.[8] Gopalakrishnan et al[2] reported a case of nonfunctioning paraganglioma that was thought to be benign but the patient died 4 years postoperatively of multiple metastases. Radiologically, the presence of calcification is uncommon. In contrast, these tumors are shown to be highly vascular and fed from the subclavian, internal mammary or intecostal arteries. The detection of mediastinal paragangliomas is possible by non-invasive imaging techniques, such as echocardiography, CT scan and magnetic resonance imaging. The pathological diagnosis is usually confirmed following a resection or by a postmortem examination. Complete excision is the recommended treatment. The operative difficulties are related to bleeding and injury of surrounding structures. Therefore, we recommend that a standby cardiopulmonary bypass circuit be available during the resection of intrapericardial paraganglioma. There is no consensus regarding screening for recurrence. The controversial point in our case is the argument that one could have resected the tumor at the time it was discovered with the mitral valve

surgery. Though this is quite feasible we did not feel comfortable tackling the tumor without knowing the diagnosis as the frozen section histology result was inconclusive. The patient was symptomatic enough from her mitral valve disease and we felt obliged to go ahead with the mitral valve repair.

References

1. Abell MR, Hart WR, Olson JR. Tumors of the peripheral nervous system. *Hum Pathol* 1970; 1: 503-551.
2. Gopalakrishnan R, Ticzon AR, Cruz PA, Kennedy FB, Duffy FC, Barmada B et al. Cardiac paraganglioma (Chemodectoma): a case report and review of the literature. *J Thorac Cardiovasc Surg* 1998; 76: 183-189.
3. Apaydin AZ, Wu D, Greipp RB. Cardiac Paraganglioma: Case Study. *Asian Cardiovascular and Thoracic Annals* 2001; 9: 212-214.
4. Sharma SK, Sanjiv S, Mukhopadhyay S. Mediastinal paraganglioma as an intracardiac mass with superior vena caval obstruction. *Thorax* 1993; 48: 1181-1182.
5. Enzinger FM, Weiss SW. Paraganglioma. In: Enzinger FM, Weiss SW, editors. Soft tissue tumors. 2nd ed. St. Louis, Washington (DC): The CV Mosby Company; 1988. p. 836-860.
6. Benjamin SP, Mc Cormack LJ, Effler D, Groves LK. Primary tumours of the mediastinum. *Chest* 1972; 62: 297-303.
7. Olson JL, Salyer WR. Mediastinal paragangliomas (aortic body tumour). A report of four cases and a review of the literature. *Cancer* 1978; 41: 2405-2412.
8. Hodgson SF, Sheps SG, Subramanian R, Lie JT, Carney JA. Cathecholamine secreting paraganglioma of interatrial septum. *Am J Med* 1984; 77: 157-161.