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Intrapericardial Paraganglioma, an Exceedingly Rare Finding: A Case Report

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Abstract

Paragangliomas are rare tumors that arise from the sympathetic or the parasympathetic ganglia. Parasympathetic paragangliomas are usually nonfunctional. They are often found in the pre aortic and paravertebral sympathetic plexus or at the base of the skull. 80% of these are Glomus jugulare and carotid body paragangliomas. Intrapericardial paragangliomas are exceedingly rare. Less than 2% of paragangliomas are found in the chest and most of them are situated in the posterior mediastinum. As such, there are no clear guidelines on how to treat intrapericardial paragangliomas. We present here the case of a patient with an intrapericardial paraganglioma who was evaluated by cardiology, cardiac surgery, and endocrinology. In this case cardiac surgery opted for a conservative approach and did not offer surgery to the patient. There is some literature supporting a surgical approach but no specific guidelines about this have been written. The patient is currently being monitored since she did not have metastatic disease and her paraganglioma was nonfunctioning. With this case we want to add to the body of evidence that this type of cases does not need an urgent surgical approach.

Keywords

Paraganglioma, Intrapericardial, Nonfunctioning

1. Introduction

This is a case report about an intrapericardial paraganglioma. These are very rare tumors, especially in this anatomic location. Several cases have been reported but most paragangliomas happen outside the heart and the amount of available literature for intrapericardial paragangliomas is not enough to delineate clear guidelines on how to approach these tumors. One of the main issues

that have been reported is whether patients should undergo surgery or not. With our case we want to add to the body of evidence about these rare tumors and how our patient has been managed conservatively given the low suspicion for malignancy and the high surgical risk these tumors pose due to their high vascularization.

2. Case Report

We present here the case of a 25-year-old female with an intracardiac mass suspicious for intrapericardial paraganglioma.

Patient was initially diagnosed with a large jugular paraganglioma on October 2022 when she presented to the emergency department with nausea, vomiting, headaches and vertigo. The paraganglioma was non secretory per work up. After her diagnosis, she was referred to a geneticist who diagnosed her with hhereditary PGL/PCC (paraganglioma/pheochromocytoma) syndrome. This was secondary to a germline pathogenic variant in SDHB gene (deletion exon 8). As part of the full work up, patient underwent a cardiac MRI (Figure 1) on February 2023 which revealed an intensely enhancing intrapericardial lesion along the anterior/superior aspect of the left atrium. The imaging features were most suggestive of an intrapericardial paraganglioma, given positive genetic analysis and concurrent imaging findings of neck glomus tumor. No obvious intraluminal infiltration into the left atrium/aorta. Her ejection fraction was reported as normal and there were no areas of late gadolinium enhancement to suggest myocardial inflammation/fibrosis/scar. Of note, because her tumor was non functional she had minimal symptoms.

Patient was subsequently referred to cardiothoracic surgery on March 2023

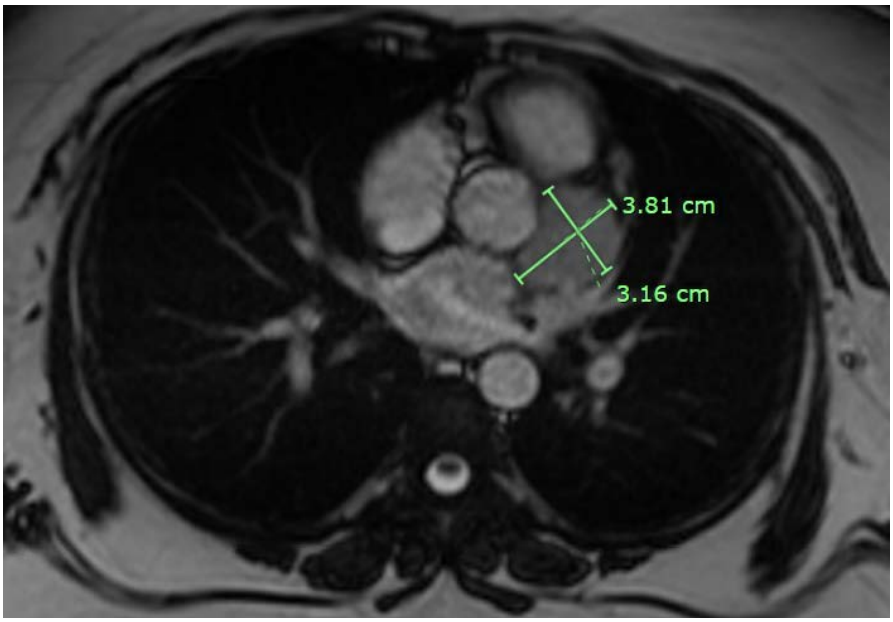

Figure 1. Intensely enhancing intrapericardial lesion along the anterior/superior aspect of the left atrium. Imaging features most suggestive of an intrapericardial para ganglioma, given positive genetic analysis and concurrent imaging findings of neck glomus tumor.

and at that time it was decided that they were not going to pursue any surgical intervention after discussing risks and benefits to the patient including the risk of hemorrhage. The plan was to obtain a CT thorax in 5 months and have the patient follow up in 6 months.

She was seen by cardiology on april 2023. During the visit patient reported having palpitations and dizziness mostly when standing up and only on certain occasions and not every day. She did not report palpitations but did say her resting heart rate was usually in the 100's.

3. Discussion

Paragangliomas are chromaffin tumors that arise from either the sympathetic or the parasympathetic ganglia. Paragangliomas affecting the heart are extremely rare. Paragangliomas themselves are rare but even more so those affecting the heart. Upon reviewing the available literature, we found that about 2% or less of paragangliomas affect the chest. Most of those tumors are found in the posterior mediastinum. The very small proportions of them found in the heart are commonly seen arising from the roof of the left atrium but then can affect any of the cardiac structures [1].

During the past decades, genetic studies have been able to identify mutations associated with paraganglioma syndromes (PGL). Two of the genes identified are succinate dehydrogenase subunit D (*SDHD*) gene which causes PGL type 1 (PGL-1), and succinate dehydrogenase subunit B (*SDHB*) gene as the susceptibility gene for PGL type 4 (PGL-4). Our patient had a mutation in SDHB gene which seems to have higher risk of malignancy compared to those with SDHD mutation [2].

The clinical presentation can vary depending on whether the tumor is functional or not. In patients with functional tumors, we can expect catecholamine related symptoms such as headaches, sweating, palpitations, and associated high blood pressure. The symptoms can also be related to the location. Intracardiac tumors can potentially disrupt blood flow or valvular function [3].

In a retrospective review of 12 patients with intrapericardial paragangliomas, 10 of them were located very close to the left atrium and 50% percent of these were found to have myocardial invasion which was confirmed by surgery or biopsy [4].

Regarding the diagnosis, the imaging studies that are most helpful are MIBG which is a nuclear scan that uses iodine-123 meta-iodobenzylguanidin, CT or MRI. Regarding tissue diagnosis, it is suggested that because they are highly vascularized, a biopsy is not always best since the risk of hemorrhage is likely high [5].

The problem with this approach is that approximately 10% of these tumors can be malignant hence a surgical resection is most of the times discussed with the patient to make the final diagnosis. More often though malignant tumors can be identified without tissue diagnosis by finding metastatic lesions on imag-

ing studies such as CT or MRI [6].

The treatment also varies depending on the type of tumor and location. In nonfunctional paragangliomas, surgery is one of the most effective treatments and sometimes the only one. Surgery for this tumor can most of the time represent a very challenging case not only because they have a very rich vascularization but also because of their location in the heart and its surrounding structures [7].

Although in a literature review of 92 cases, the invasion of contiguous structures in the heart, the diameter of the paraganglioma, and the functional status did not appear to affect the survival and this remain the same whether the patient had surgery or not [8].

In another study of 158 patients with cardiac paragangliomas, 76 of them had a complete resection of the tumor and 15 of them had incomplete resection. 2 of the patients ended up having heart transplant surgery. 100 of these patients were follow up and at 5 years, 80% of them were alive. In this study also it is suggested that because most of the cardiac paragangliomas are benign, surgical resection is the best treatment approach with an expected good recovery and prognosis. For malignant cardiac paragangliomas, the therapeutic approach is more uncertain [9].

In the case of our patient, she does not seem to have evidence of metastatic disease and her paraganglioma seems to be nonfunctional. Cardiothoracic surgery decided after discussing with the patient that it is better not to perform surgery as of now and continue with 6-month surveillance.

The patient is also expected to follow up with endocrinology and oncology.

4. Conclusion

The management of intrapericardial paragangliomas remains a challenge across the world. The reported cases in the literature do not demonstrate a homogeneous approach. The main question is still whether a patient with this type of tumor would benefit from surgery or not. Our case shows the complexity of this tumors and how this requires a multidisciplinary approach. The patient continues to do well and currently follows with surgical and non-surgical teams.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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