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Case Report

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Non-neural (S-100 Negative) granular cell tumor of the pericardium

Abstract

Non-neural granular cell tumors (NNGCTs) are rare neoplasms with distinct histopathological and immunohistochemical features. We present the case of an asymptomatic patient in whom a pericardial mass was incidentally discovered during coronary artery bypass graft surgery. Histopathological examination revealed a lesion composed of large polygonal cells with granular eosinophilic cytoplasm, papillary-like architectural features, smooth nuclei, and a low Ki-67 proliferation index. Immunohistochemical analysis demonstrated strong positivity for CD68, while staining was negative for S-100, neuron-specific enolase (NSE), cytokeratin, CD34, α-smooth muscle actin (α-SMA), and desmin. The diagnosis was consistent with an S-100-negative non-neural granular cell tumor of the pericardium—an extremely rare entity, not previously reported in the literature to our knowledge. This case highlights the importance of considering NNGCT in the differential diagnosis of incidental pericardial masses identified on echocardiography, computed tomography, or cardiac magnetic resonance imaging.

Keywords: Granular cell tumor, S-100 protein, pericardium, non-neural granular cell tumor, cardiac tumor

INTRODUCTION

Pericardial tumors are most commonly secondary lesions resulting from local invasion or distant metastasis. Primary pericardial tumors account for approximately 6.7% to 12.8% of all primary cardiac region tumors, with an estimated prevalence of 0.001% to 0.007% (1,2). Among benign pericardial masses, pericardial cysts are the most frequent, while the most common primary pericardial malignancy is mesothelioma (3).

Non-neural granular cell tumor (NNGCT) is a rare, low-grade mesenchymal neoplasm characterized by abundant cytoplasmic granules and an immunohistochemically S-100-negative profile, suggesting non-neural differentiation (4,5). NNGCTs most frequently arise in the skin, particularly on the back, extremities, face, and scalp. The most common extracutaneous sites are the oral cavity and tongue. To date, only three cases have been documented in the literature—two in the bronchial tree and one in the uterine corpus (4).

To the best of our knowledge, no cases of pericardial NNGCT have previously been reported. Here, we present the case of a 47-year-old man in whom a localized pericardial NNGCT was incidentally discovered during coronary artery bypass graft (CABG) surgery.

CASE REPORT

Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

A 47-year-old man was admitted for elective CABG surgery following coronary angiography. Preoperative routine laboratory and imaging studies were conducted. Transthoracic

echocardiography revealed hypokinesia of the lateral, inferior, and posterior walls, with a left ventricular ejection fraction of 30%. The end-systolic diameter measured 38 mm, and the end-diastolic diameter was 55 mm. No additional pathological findings were identified during preoperative evaluations, and the patient was cleared for surgery.

During the procedure, following median sternotomy and pericardial exposure, a yellow-gray cystic mass measuring approximately 2.0 × 1.5 × 1.5 cm was observed near the junction of the right lateral wall of the ascending aorta and the parietal pericardium (Figure 1). No other mass lesions were detected on the heart or within the pericardial cavity. A simple excisional biopsy of the lesion was performed, and the CABG procedure proceeded as planned. Off-pump CABG was completed using the radial artery for the right coronary artery and the left internal mammary artery for the left anterior descending artery.

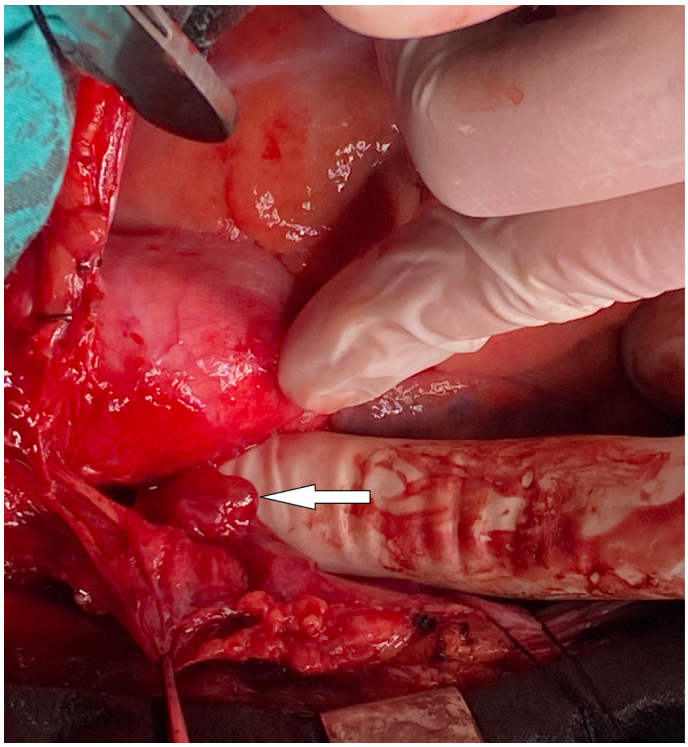

Figure 1. Intraoperative photograph showing a yellow-gray, well-circumscribed cystic mass adjacent to the right lateral wall of the ascending aorta and parietal pericardium

The patient's postoperative course was uneventful, and he was discharged on postoperative day six without complications. Approximately one week later, histopathological examination of the excised mass revealed features consistent with a non-neural granular cell tumor (NNGCT) (Figure 2).

Histomorphological examination revealed a well-demarcated lesion composed of large polygonal cells with abundant granular eosinophilic cytoplasm, forming papillary-like structures. The nuclei were round to oval, smooth, and showed mild pleomorphism, with occasional nuclear prominence. The Ki-67 proliferation index was low, indicating minimal mitotic activity.

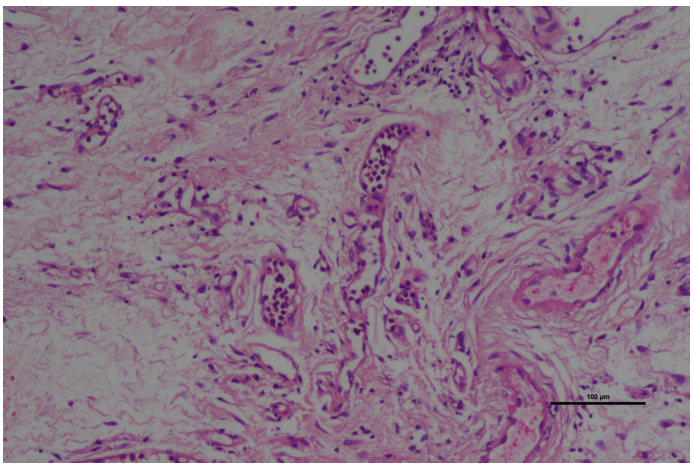

Figure 2. Histological section stained with hematoxylin and eosin (H&E), demonstrating large polygonal cells with abundant granular eosinophilic cytoplasm. Original magnification ×100

Immunohistochemical analysis showed strong positivity for vimentin, CD68, and factor XIIIa. Staining for smooth muscle actin (SMA), S-100 protein, CD34, desmin, anaplastic lymphoma kinase (ALK), neuron-specific enolase (NSE), and HMB-45 was negative, supporting a diagnosis of NNGCT.

At the six-month postoperative follow-up, no residual or recurrent mass was detected on transthoracic echocardiography. The patient remains under clinical surveillance.

DISCUSSION

Histopathologically, NNGCTs are composed of spindle to oval cells with granular eosinophilic cytoplasm and vesicular nuclei. Immunohistochemically, they may exhibit positivity for markers such as NSE, CD68, factor XIIIa, NKI/C3, HMB-45, SMA, vimentin, and ALK, while S-100 protein is typically negative (5).

In our case, histopathological analysis revealed large polygonal cells with granular eosinophilic cytoplasm and occasional nuclear prominence. Immunohistochemistry showed strong positivity for CD68 and factor XIIIa, with negative staining for S-100 protein, supporting the diagnosis of NNGCT.

Cutaneous NNGCTs are generally asymptomatic and exhibit benign biological behavior (4–6). Conservative surgical excision is the treatment of choice. In rare cases, regional lymph node metastases have been reported; these were managed with surgical excision. One case described the development of axillary lymph node metastasis four years postoperatively, which was treated with re-excision (6). In a pediatric case of bronchial NNGCT, acute respiratory failure occurred due to tumor location; however, no recurrence was observed following lobar resection (7).

CONCLUSION

Due to its benign nature, surgical management of pericardial NNGCT may be approached similarly to other benign pericardial tumors, with treatment tailored according to tumor size and anatomical location. In this case, the lesion was incidentally identified in an asymptomatic patient, and complete excision was achieved owing to its small size, accessible location, and non-aggressive behavior.

NNGCT is a rare pathological entity, and to the best of our knowledge, no prior case of pericardial origin has been reported. We suggest that this rare diagnosis be considered in the differential evaluation of incidentally detected pericardial masses during imaging studies.

Conflict of Interests

The authors declare no conflicts of interest related to any financial organization or material discussed in this manuscript.

Financial Disclosure

No financial or material support was received from any pharmaceutical company or manufacturer of medical devices and materials related to the subject of this study.

Patient Informed Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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