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(CASE REPORT)

Cardiac ventricular fibroma with surgical excision, late recurrence and refractory heart failure: Case report and a brief review of the literature

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Abstract

Primary cardiac tumors are exceedingly rare in the pediatric population, with cardiac fibromas (CFs) representing the second most common benign neoplasm. Although histologically benign, these intramural tumors frequently precipitate life-threatening, intracavitary obstruction, and sudden cardiac death. We present the case of a 9-year-old male who presented with exercise-induced syncope and was found to have a large, arrhythmogenic interventricular septal fibroma. The patient underwent successful complete surgical excision with patch reconstruction, resulting in immediate resolution of his arrhythmia burden. However, 5 years postoperatively, he developed a late, infiltrative recurrence at the patch margin, precluding complete re-resection. Despite partial debulking and implantable cardioverter-defibrillator placement, the patient experienced progressive left ventricular dysfunction and refractory heart failure, ultimately succumbing to the disease at age 19 while awaiting heart transplantation. This case underscores the unpredictable long-term biological behavior of cardiac fibroma. It highlights the critical diagnostic role of multimodal imaging, the surgical challenges of septal reconstruction, and the rare but devastating potential for late recurrence. Furthermore, it emphasizes the necessity of lifelong surveillance and the complex interplay between repeated surgical interventions, chronic arrhythmia, and progressive myocardial failure in pediatric patients with recurrent cardiac fibromas.

Keywords: Cardiac fibroma; Benign; Recurrent; Ventricular arrhythmias; Progressive myocardial failure

1. Introduction

Primary cardiac tumors are exceptionally rare, with an estimated incidence of 0.03% to 0.32% in the general population. [1] Among these, cardiac fibromas (CFs) are the second most common benign primary cardiac tumor in children, following rhabdomyomas. [1,2] Despite their histologically benign nature, CFs pose significant clinical challenges due to their propensity to cause intracavitary obstruction, conduction system disruption, and life-threatening ventricular arrhythmia. [1,3] While complete surgical resection is the definitive treatment and generally confers an excellent long-term prognosis, tumors involving the interventricular septum present formidable surgical complexities, often requiring extensive myocardial resection and patch reconstruction. [4]

A critical gap in the current literature remains the long-term biological behavior of these tumors following complete gross resection. Late recurrence is exceedingly uncommon and poorly characterized, with limited data guiding the

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management of recurrent, infiltrative lesions that precipitate progressive ventricular dysfunction. [5] This report detailed a rare case of a pediatric ventricular septal fibroma that, despite initial complete surgical excision, exhibited late, aggressive recurrence leading to refractory heart failure.

This case is reported to highlight the diagnostic nuances, the surgical dilemmas of septal involvement, and the imperative for lifelong surveillance due to the unpredictable potential for late recurrence and subsequent myocardial failure.

2. Case Presentation

2.1. Clinical Presentation

A 9-year-old male was referred to a pediatric cardiology center following an episode of exercise-induced syncope during a school physical education class. His parents reported a two-month history of intermittent palpitations and reduced exercise tolerance, previously attributed to deconditioning. Review of systems revealed two prior fainting episodes over the preceding year, each dismissed as vasovagal in nature. There was no prior cardiac diagnosis, though his mother recalled being told at his 4-year-old well-child visit that a heart murmur was present and then "resolved." He had no prior cardiac imaging. Family history was noncontributory, with no known cardiac disease, sudden death, or neurocutaneous syndrome. He presented now following an emergency department visit where an EKG was performed, revealing a significant finding that prompted urgent cardiology referral.

2.2. Physical Examination, History, Investigations & Differential Diagnosis

On examination, the child appeared well-nourished and in no acute distress. Heart rate was 88 beats per minute with occasional irregularity; blood pressure was 104/62 mmHg; oxygen saturation was 98% on room air. An II/VI systolic murmur was audible along the left lower sternal border. No skin lesions, ash-leaf macules, or café-au-lait spots were identified, effectively reducing the likelihood of tuberous sclerosis complex at clinical examination. Neurological assessment was unremarkable.

The ECG demonstrated sinus rhythm with frequent unifocal premature ventricular contractions exhibiting a left bundle branch block morphology, suggesting a right ventricular or septal origin, along with diffuse T-wave abnormalities across the precordial leads. A 48-hour Holter monitor recorded a PVC (Premature Ventricular Contractions) burden of 22% with two runs of no sustained ventricular tachycardia. Laboratory investigations, including complete blood count, comprehensive metabolic panel, thyroid function, and inflammatory markers, were all within normal limits. Brain natriuretic peptide (BNP) was mildly elevated to 98 pg/mL.

Transthoracic echocardiography (TTE) demonstrated a large, well-circumscribed, echogenic, non-mobile mass measuring 4.1 × 3.6 cm within the interventricular septum, extending into the apical left ventricular myocardium without intracavitary protrusion. Biventricular systolic function was preserved with an ejection fraction of 62%. No outflow tract obstruction was identified. Cardiac MRI with gadolinium confirmed a solitary intramyocardial mass that was markedly hypointense on both T1- and T2-weighted sequences, consistent with a densely fibrous composition. Late gadolinium enhancement was uniformly distributed across the lesion. Computed tomography of the chest demonstrated faint central dystrophic calcification within the mass, a finding highly characteristic of cardiac fibroma.

The radiographic differential diagnosis was narrow given the characteristic constellation of findings. CF was the leading and favored diagnosis based on the pediatric age, solitary intramyocardial location, T2 hypointensity, homogeneous fibrotic LGE pattern, and CT calcification. Cardiac rhabdomyoma was considered but excluded based on solitary presentation, absence of tuberous sclerosis stigmata, and the T2 hypointense signal; rhabdomyomas are typically T2 hyperintense. Hamartoma of mature cardiac myocytes was listed as a remote possibility. Primary cardiac sarcoma was considered but regarded as exceedingly unlikely given the patient's age, and benign imaging characteristics.

2.3. Multidisciplinary Tumor Board Discussion

The case was reviewed at the pediatric cardiac tumor board attended by pediatric cardiologists, congenital cardiac surgeons, pediatric radiologists, and pathologists. Discussion centered on the indications for surgical intervention versus continued surveillance. Given the tumor's large size relative to the heart, the significant PVC burden with documented NSVT (Non-Sustained Ventricular Tachycardia) conferring a risk of sudden cardiac death, and the progressive symptomatology, the board reached consensus in favor of surgical resection with the goal of complete excision. The anticipated need for patch reconstruction of the interventricular septum was discussed. Cardiac transplantation was noted as a contingency if intraoperative findings precluded safe, complete resection. Preoperative

genetic tests for TSC1/TSC2 and PTCH1 were negative, supporting a diagnosis of sporadic fibroma. Biopsy prior to resection was not recommended, given the characteristic imaging and the surgical plan already established.

2.4. Management, Pathology, IHC & Final Diagnosis

The patient underwent median sternotomy with cardiopulmonary bypass. Intraoperatively, the mass was identified as a firm, white, tan, well-demarcated lesion without a true capsule, adherent at its base to the dense fibromuscular tissue of the interventricular septum. The surgical team achieved grossly complete resection with negative margins, confirmed on intraoperative frozen section. Septal reconstruction was performed using a glutaraldehyde-fixed autologous pericardial patch. The aortic cross-clamp time was 94 minutes. Intraoperative TTE confirmed no residual mass and preserved biventricular function.

Gross pathological examination revealed a firm, rubbery, white-gray mass measuring 4.3 × 3.8 × 3.1 cm with a whorled cut surface, interspersed with areas of chalky calcification. No necrosis or hemorrhage was identified. Histological examination on permanent sections demonstrated a paucicellular, densely collagenous stroma composed of bland fibroblastic spindle cells arranged in interlacing bundles. Entrapped islands of residual cardiomyocytes were identified at the peripheral margin. Mitotic figures were absent. No nuclear atypia, necrosis, or increased cellularity was observed. (Figure 1 A, B)

Immunohistochemistry (IHC) studies were performed to confirm the diagnosis and exclude malignancy. The spindle cells showed positivity for vimentin and CD34, consistent with fibroblastic differentiation. Desmin, SMA, and myogenin were negative, excluding a myogenic neoplasm. Pan-cytokeratin (AE1/AE3) was negative. TLE1 was negative, effectively excluding synovial sarcoma. S-100 was negative. The Ki-67 proliferative index was less than 1%, indicating a biologically indolent lesion. No molecular studies were indicated, given the unambiguous morphological and IHC findings. The final pathological diagnosis was ventricular cardiac fibroma, completely excised, with dystrophic calcification, without cytological atypia or malignant features.

2.5. Outcome, Recurrence, Complications & Unfavorable Prognosis

The patient's immediate postoperative course was uncomplicated. Epicardial temporary pacing was required for 48 hours due to a transient junctional rhythm, which was resolved spontaneously. He was discharged on postoperative day nine in stable condition. Holter monitoring at six weeks post-discharge documented a dramatic reduction in PVC burden to less than 2%, with no further episodes of NSVT. Cardiac MRI at six months confirmed no residual or recurrent lesion and normal biventricular function. The patient resumed full physical activity at eight months postoperatively and was followed annually with cardiac MRI and echocardiography per institutional protocol.

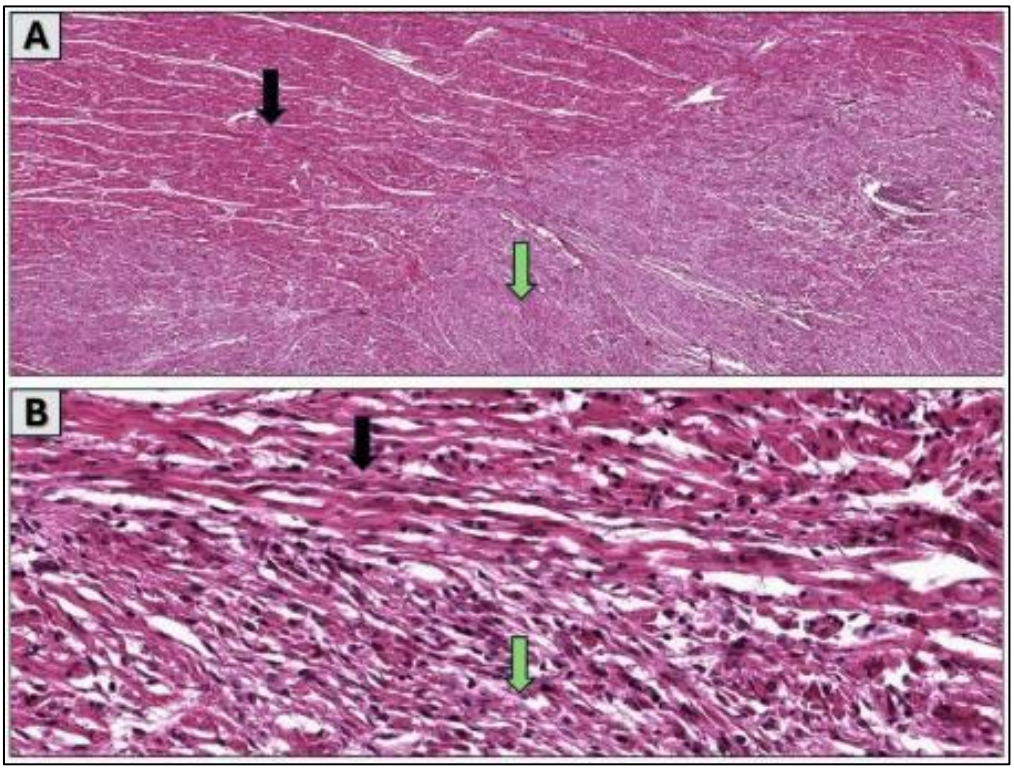
At five-year follow-up, surveillance cardiac MRI identified a 1.4 cm nodular area of T2-hypointense signal with gadolinium enhancement at the apical margin of the pericardial patch. This was initially regarded as scar tissue or a benign fibrous nodule, and the patient remained asymptomatic. Serial MRI at six- and twelve-month intervals over the subsequent two years documented progressive growth of this nodule to 2.9 cm. Cardiac CT confirmed the absence of calcification in the new lesion, a departure from the original fibroma, and FDG-PET demonstrated mild metabolic activity. A second surgical resection was undertaken at seven years post-initial surgery.

Intraoperatively, the recurrent lesion was found to be adherent to the pericardial patch and deeply infiltrative into the adjacent ventricular myocardium, precluding complete excision without critically compromising ventricular integrity. A partial debulking was performed. Histopathology confirmed recurrent cardiac fibroma with identical morphological features to the original lesion; no malignant transformation was identified. The IHC profile was unchanged, and TLE1 remained negative.

At age 14, the patient experienced an episode of sustained ventricular tachycardia, leading to the implantation of a dual-chamber implantable cardioverter-defibrillator (ICD). Two years later, at age 16, he developed progressive left ventricular dysfunction. This decline was attributed to a combination of factors: myocardial loss from repeated surgeries, the ongoing arrhythmia burden from a residual fibroma, and chronic reliance on right ventricular pacing from his ICD. Over the two years following the partial resection, his LVEF decreased to 32%, despite being on optimized guideline-directed medical therapy including sacubitril-valsartan, carvedilol, and spironolactone.

At age 18, the patient was listed for orthotopic heart transplantation. Bridging with a left ventricular assist device (LVAD) was not feasible due to the anatomical distortion of the ventricular septum and apical myocardium from repeated surgical interventions. He experienced two hospitalizations for decompensated heart failure over the following year. While awaiting transplantation at age 19, he suffered an out-of-hospital cardiac arrest with pulseless

ventricular tachycardia; ICD shock successfully restored rhythm, but he sustained hypoxic-ischemic encephalopathy with moderate neurological sequelae. He died at age 19 while awaiting a suitable donor organ, following a decision by the family and the palliative care team to transition to comfort-focused care after a second resuscitated arrest left him with severe neurological impairment. The primary cause of death was recorded as refractory heart failure secondary to recurrent cardiac fibroma with extensive myocardial replacement.



1A: Low power view showing uninvolved myocardial muscle (Black arrow) and the cardiac fibroma, no presence of necrosis (Green arrow) (H&E X20).; **1B:** High power view showing uninvolved myocardial muscle (Black arrow) and the cardiac fibroma with densely collagenous stroma composed of bland fibroblastic spindle cells arranged in interlacing bundles (Green arrow) (H&E X40).

Figure 1 Microscopic examination of the excised ventricular cardiac fibroma

3. Discussion

3.1. Background (History, epidemiology, risk factors, and WHO classification)

The entity of cardiac fibroma (CF) was first recognized in the mid-20th century, with early surgical excisions reported in the 1960s. [6] Epidemiologically, primary cardiac tumors are rare, with a prevalence ranging from 0.0017% to 0.28% in autopsy series. [1] CFs predominantly affect the pediatric demographic, with approximately 90% of cases diagnosed in infants and children. [1]

While most cases are sporadic, approximately 3% to 5% of patients with nevoid basal cell carcinoma syndrome (Gorlin-Goltz syndrome), an autosomal dominant disorder linked to mutations in PTCH1 or SUFU, develop cardiac fibromas. [1] Anatomically, these tumors are almost exclusively intramural and solitary, with the left ventricular free wall and the interventricular septum being the most typical locations. [1,3] According to the 2021 World Health Organization (WHO) classification of cardiac tumors, fibromas are categorized as benign, non-neoplastic hamartomas composed of monomorphic fibroblasts and dense collagenous stroma. [1,7]

Unlike rhabdomyomas, fibromas do not spontaneously regress. Their biological behavior is characterized by slow growth that typically ceases as the heart reaches maturity; however, their rigid, noncontractile nature within the myocardium frequently serves as an arrhythmogenic focus, predisposing patients to ventricular tachycardia and sudden cardiac death. [1,3]

3.2. Pathogenesis, Pathophysiology

The pathogenesis of cardiac fibromas is rooted in the localized, uninhibited proliferation of monomorphic fibroblasts and the subsequent excessive deposition of dense collagenous extracellular matrix. [11] While the majority of cases are sporadic, the molecular etiology is closely linked to the Sonic Hedgehog (SHH) signaling pathway. Specifically, mutations or loss of heterozygosity in the PTCH1 tumor suppressor gene, a hallmark of Gorlin-Goltz syndrome, lead to constitutive activation of the SHH pathway, driving aberrant fibroblastic growth. [12]

Pathophysiologically, the clinical manifestations of cardiac fibromas are dictated by their rigid, noncontractile nature and their anatomical location. When situated in the interventricular septum or ventricular free walls, these large, unyielding masses can cause significant mass effect, leading to intracavitary obliteration, inflow or outflow tract obstruction, and progressive diastolic and systolic dysfunction. [11] Furthermore, cardiac fibromas are highly arrhythmogenic. Histopathologically, the tumor margins frequently exhibit "interdigitating myocardial tongues", areas where dense fibrotic tissue intertwines with viable, entrapped cardiomyocytes. [13] This architectural distortion disrupts normal electrical conduction, creating areas of slow conduction and heterogeneous repolarization. These interdigitating zones serve as an ideal electrophysiological substrate for re-entry circuits, which are the primary mechanism underlying the life-threatening ventricular tachycardias and sudden cardiac death frequently observed in these patients. [13,14]

3.3. Comparative Analysis of Our Case with Existing Literature. (Clinical, radiology, pathology, Lab, diagnosis, management, and outcome)

The presented case exhibited classic initial features of a pediatric cardiac fibroma but diverged significantly in its long-term clinical trajectory. Consistent with established literature, the patient presented with ventricular arrhythmias and syncope, which are the most common manifestations of septal fibromas. [1,3] The diagnostic utility of cardiac MRI, demonstrating characteristic T2 hypointensity and homogeneous late gadolinium enhancement, and CT imaging revealing dystrophic calcification aligned perfectly with standard radiological profiles. [1] The initial clinical decision-making favored complete surgical resection, the gold standard that typically yields excellent arrhythmia-free survival. [4,8] However, the late recurrence observed in this case was highly atypical.

The literature suggests that complete resection of cardiac fibromas is generally curative, with recurrence being exceedingly rare and almost exclusively associated with incomplete initial debulking. [5,8] In contrast, our patient experienced an aggressive, infiltrative recurrence five years after a grossly complete resection with negative margins. Furthermore, the recurrent lesion lacked the characteristic calcification of the primary tumor, presenting a diagnostic challenge. The subsequent clinical decline highlighted the compounded risks of repeated surgical interventions, chronic right ventricular pacing, and residual tumor burden.

While recent large cohort studies indicate that most children maintain left ventricular function post-resection. [4,9], our patient developed progressive, refractory heart failure. This trajectory underscored the limitations of surgical debulking in recurrent, infiltrative septal lesions. It illustrated the complex transition from an arrhythmic threat to irreversible myocardial failure, ultimately necessitating consideration for orthotopic heart transplantation, a recognized but rare endpoint for unresectable fibromas. [10]

4. What Have We Learned from This Case?

This case provided several critical clinical insights into the management of pediatric cardiac fibromas. Diagnostically, it reinforced the necessity of multimodality imaging; the classic triad of T2 hypointensity, late gadolinium enhancement, and central calcification remains highly reliable for initial diagnosis. [1] However, clinicians must recognize that recurrent lesions may present atypically, such as lacking calcification. They must maintain a high index of suspicion for any new myocardial mass adjacent to a previous resection site.

A vital lesson was the unpredictable long-term behavior of these tumors. Even after grossly complete resection with negative margins, late recurrence remains a significant risk. This challenged the prevailing notion that complete excision is uniformly curative, mandating rigorous, lifelong surveillance with serial cardiac MRI. From a management perspective, the case highlighted the profound implications of septal involvement. While initial resection successfully mitigated the immediate arrhythmic risk, the structural distortion from patch reconstruction, combined with subsequent recurrences and chronic pacing, precipitated irreversible left ventricular dysfunction. [9] This emphasizes that in cases of extensive septal fibromas, the long-term prognosis may be dictated not only by the tumor itself but by the cumulative myocardial loss and mechanical desynchrony resulting from surgical interventions. Consequently, early

multidisciplinary evaluation for advanced heart failure therapies, including transplantation, should be integrated into the long-term care plan for patients with recurrent or unresectable disease. [10]

Abbreviations

- Cardiac fibromas (CFs);
- Transthoracic echocardiography (TTE);
- Immunohistochemistry (IHC);
- NSVT Non-Sustained Ventricular Tachycardia (NSVT).

5. Conclusion

In conclusion, this case of a pediatric ventricular septal fibroma highlighted the rare but devastating potential for late, infiltrative recurrence following complete surgical excision. While initial resection successfully eradicated the immediate arrhythmic threat, the subsequent development of a recurrent mass and progressive, refractory heart failure underscored the complex long-term morbidity associated with extensive septal tumors.

This report added significant value to the existing literature by documenting a highly unusual clinical trajectory that challenged the prevailing paradigm of uniform surgical curability following complete fibroma excision. It explicitly reinforced the necessity for lifelong, high-resolution cardiac MRI surveillance, even in patients who achieve initial complete resection with negative margins. Furthermore, this case demonstrated that the cumulative effects of repeated surgical reconstruction, chronic right ventricular pacing, and residual tumor burden can precipitate irreversible myocardial failure in a manner that is distinct from the tumor's primary arrhythmogenic threat.

Clinicians managing pediatric patients with septal fibroma must therefore adopt a longitudinal, multidisciplinary approach that integrates early recognition of recurrence, timely escalation of heart failure therapies, and proactive consideration for orthotopic heart transplantation when disease progression renders the myocardium irreversibly compromised.

Compliance with ethical standards

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All authors make the following declarations:

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Statement of ethical approval

Ethical review and approval were not required for this study involving human participants. The paper has been sufficiently anonymized to maintain the patient's confidentiality.

Data access statement

All relevant data are included in the paper.

Author contributions

All authors contributed equally to producing this manuscript.

Statement of informed consent

This study was conducted as a retrospective review of archival pathology material collected during routine clinical care. All data were fully de-identified prior to analysis. No patient contact or intervention was involved, and informed consent was not required in accordance with institutional policies.

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