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Case Report

A Challenging Diagnosis of Unroofed Coronary Sinus, Myxoma and Left Atrial Aneurysm

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Abstract: An unroofed coronary sinus is a malformation that leads to a shunt between the coronary sinus and the left atrium. In our case, the shunt led to the formation of a gigantic left atrial aneurysm compressing the left atrium on transesophageal echocardiography. During surgery, a myxoma was incidentally discovered.

Keywords: left atrial aneurysm; unroofed coronary sinus; myxoma

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1. History of Presentation

An 83-year-old male was incidentally diagnosed with a giant left atrial aneurysm (LAA) via transthoracic echocardiography (TTE) during a routine check-up examination two years ago. At that time, he was asymptomatic and denied any shortness of breath, chest pain, or dizziness. On examination there was no cardiac murmur, and his jugular venous pressure was normal. His vital signs on admission documented a heart rate of 104 beats per minute, blood pressure of 130/70 mmHg and oxygen saturation of 98% on room air. His laboratory workup values, including pro-BNP, were within normal limits. His chest X-ray (Figure 1) evidenced an enlarged cardiac contour. He was evaluated by the cardiology team that decided to follow-up the patient with serial TTE evaluation every six months.

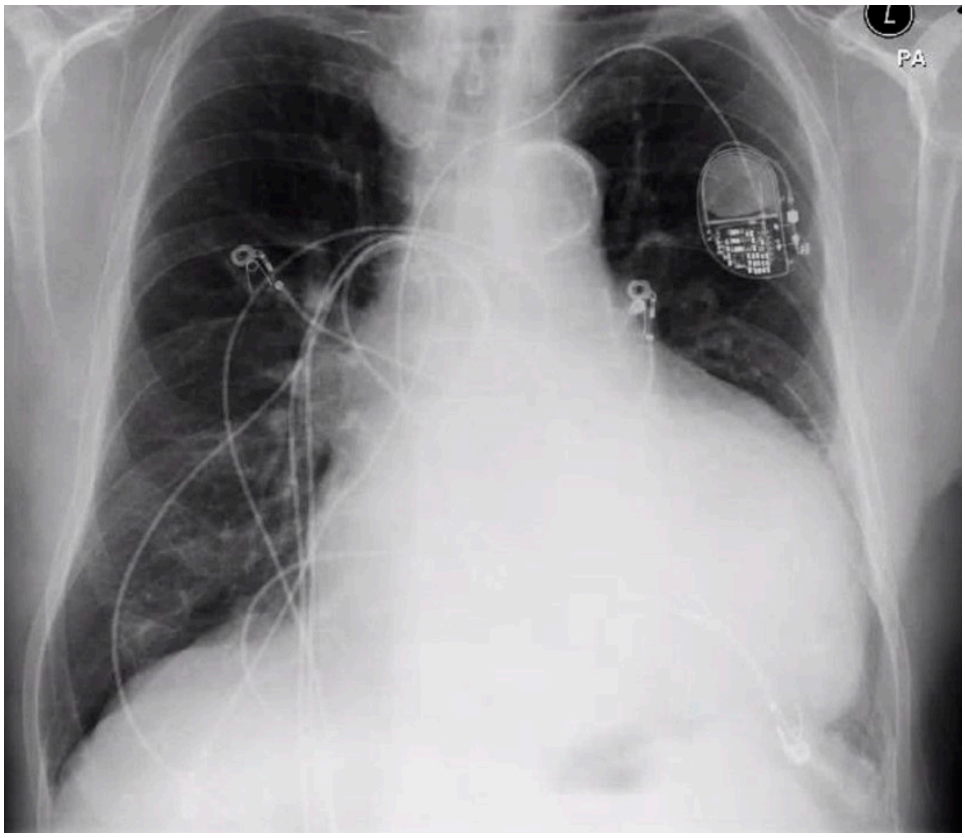

Figure 1. Chest X-ray evidencing an enlarged cardiac contour.

2. Medical History

His past medical history was remarkable for chronic atrial fibrillation and tachycardia-bradycardia syndrome. Due to two previous syncopal episodes the patient underwent an implantable cardioverter-*defibrillator* implantation. Other medical conditions included chronic kidney disease, two previous episodes of deep venous thrombosis, and rheumatoid arthritis.

3. Methods

The patient signed the consent for the disclosure of the medical information for publication.

3.1. Investigations

A preoperative electrocardiogram evidenced a ventricular-paced rhythm with occasional premature ventricular complexes (Figure 2). A transesophageal echocardiography (TEE) evidenced a LAA, 10.2 × 7.7 cm connected to the left atrium. There was severe biatrial enlargement with an enlarged right ventricle. A bubble study investigation did not evidence any communication between the left and right atrium at the level of the atrial septum. A follow-up TEE reported the LAA to be greater in size (14 × 7 cm), echo dense, and heterogeneous with cystic and solid components. A shunting flow communication between the right atrium and the LAA was discovered by color Doppler (Video S1). No persistent left superior vena cava (PLSVC) was noticed. Following CT-scan imaging (dual-source 64-slice scanner, Definition, Siemens, Germany) the LAA appeared to be irregular and lobulated (11 × 14 × 11 cm), compressing the left atrium, with the presence of a prominent coronary sinus (Video S2). However, it did not evidence a fistulous connection between the LAA and the cardiac chambers.

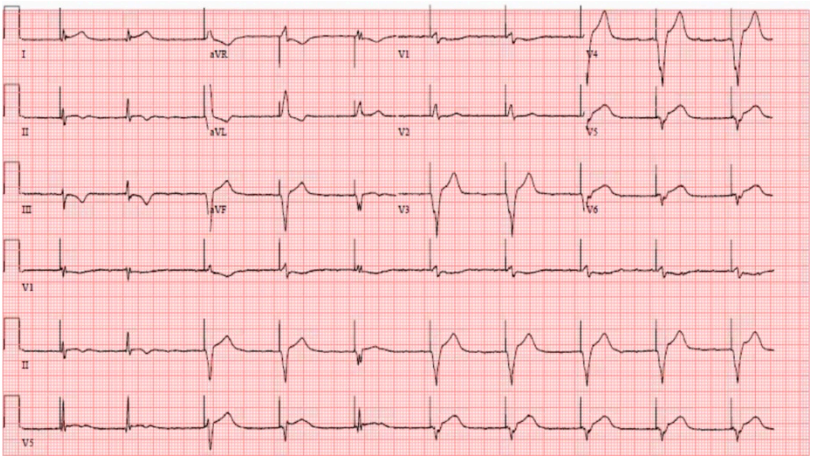

Figure 2. Electrocardiogram evidencing a ventricular-paced rhythm with occasional premature ventricular complexes.

3.2. Differential Diagnosis

Due to the conflicting imaging reports the differential diagnosis included LAA, diverticulum of the left atrium, isolated pericardial cyst, angiosarcoma, teratoma, echinococcosis, and unroofed coronary sinus (UCS).

3.3. Management

The patient was admitted to the inpatient service at the cardiac surgery department. We performed a left hemi-clamshell thoracotomy to have better surgical exposure in case an angiosarcoma or a teratoma was to be found. Due to the poor surgical exposure, we extended the incision, performing a full clamshell incision. The intraoperative, direct observation macroscopically indicated a LAA. A primary cavity incision resulted in continuous bleeding with hemodynamic instability. Therefore, an emergent cardiopulmonary bypass was established through arterial and venous femoral cannulation, and the heart was arrested after aortic cross-clamping. LAA cavity exploration led to large clots evacuation and a 2 cm myxoma removal. Indeed, a large UCS was present and other two smaller foramen were diagnosed (Figure 3). No PLSVC was present. The UCS communication with the left atrium and the mass was closed with a pericardial bovine patch (Braile Biomédica®), while the other two foramens were primarily closed (Video S3).

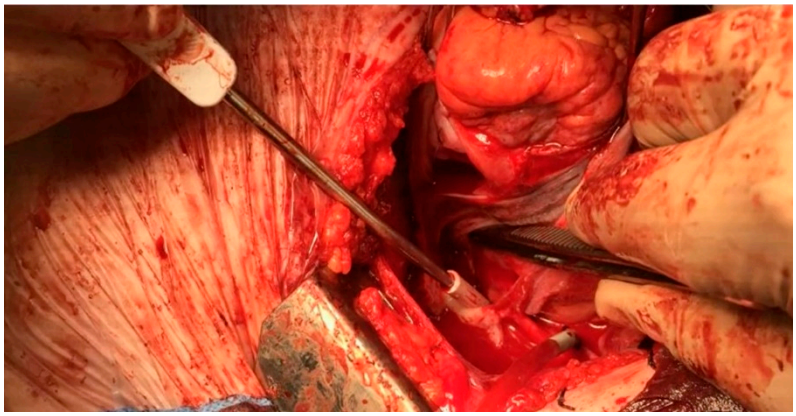

Figure 3. Intraoperative view of the unroofed coronary sinus.

3.4. Discussion

We described a rare case of an adult man with clinically silent UCS, LAA, and myxoma that was found during a routine check-up requiring surgery to perform a differential diagnosis.

UCS syndrome is a cardiac abnormality in which a communication occurs between the coronary sinus and the left atrium, because of the partial (either focal or fenestrated) or complete absence of the roof of the coronary sinus. The morphological types of UCS have been classified into four groups: type I, completely unroofed with PLSVC; type II, completely unroofed without PLSVC; type III, partially unroofed mid portion; and type IV, partially unroofed terminal portion. The abnormality is often associated with PLSVC and other forms of complex congenital heart disease, usually heterotaxia syndromes [1]. Kim et al. found that in patients with UCS, the mean diameter of the coronary sinus standardized to the patient's body surface area was 15 ± 4 mm/m^2. This is similar to that of patients with a PLVCS without UCS (15 ± 6 mm/m^2) and significantly greater than patients with normal anatomy (7 ± 2 mm/m^2) [2]. However, the diagnosis is often difficult because of nonspecific clinical features. The diagnosis of this lesion is important to the prognosis of the patient because of the consequences of brain abscess or cerebral emboli that may result from a right-to-left shunt [3]. Delayed diagnosis may lead to left-to-right shunt and right atrial and ventricular enlargement. In long standing right to left-to-right shunt, the left atrial intracavitary pressure increases by switching the shunt right-to-left. Transthoracic echocardiography imaging demonstrates right-sided heart chambers enlargement. After injection of agitated saline via the left arm, prominent contrast can be seen in the left heart with minimal opacification of right-sided chambers. This confirms the presence of an unroofed coronary sinus (CS) defect. If agitated saline is given via the right arm, greater contrast is seen in the right atrium and ventricle, in comparison to the left-sided chambers. This confirms the presence of a right-to-left intracardiac shunt [4–6]. Cardiac CT-scan imaging and Magnetic Resonance Imaging are also valuable tools to diagnose the UCS and can complement echocardiographic imaging to demonstrate the PLSVC and any possible anomalous pulmonary venous drainage [7,8]. Acquired pathologic LAA is a rare condition, and patients may present with atrial tachyarrhythmias. In our case tachyarrhythmias led to the implantation of an ICD. Surgical obliteration and LAA closure through a minimally invasive approach is the mainstay of treatment for preventing potentially fatal cardiovascular adverse events, such as stroke. In our case, due to the large mass and patient comorbidities, we opted for the hemiclamshell approach. The actual medical literature does not describe any reported case of a myxoma inside a LAA associated with an UCS. Therefore, the echocardiographic diagnosis is challenging especially for novel cardiologists, anesthesiologists, and surgeons. In this case report, we provide an insight of echocardiographic and surgical evidence on what are the parameters and characteristics of this unusual congenital combination. In our case, all the preoperative imaging workup did not evidence an UCS.

3.5. Follow-Up

At the 3-month follow-up, the patient remained well compensated.

4. Conclusions

In adults, the combination of an UCS, LAA, and myxoma represent a challenging subset of patients due to the rarity and heterogenicity of this combination. This makes the complex understanding of this abnormality essential to customize an appropriate surgical approach. In this case, we demonstrated that a rigorous preoperative workup and a heart-team collaboration is essential to perform an adequate patient-customized treatment.

Supplementary Materials: The following supporting information can be downloaded at: https://www.mdpi.com/article/10.3390/hearts3030010/s1. Video S1: Transesophageal echocardiography evidencing the LAA and a shunting flow communication between the right atrium and the LAA. Video S2: CT-scan evidencing the LAA and the presence of a prominent coronary sinus. Video S3: Intraoperative diagnosis of the UCS, the LAA, and the myxoma.

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Informed Consent Statement: Informed consent was obtained from the patient involved in the study.

Data Availability Statement: The data presented in this study are available in the article or supplementary material.

Conflicts of Interest: The authors declare no conflict of interest.

Abbreviations

LAA	Left atrial aneurysm
TTE	Transthoracic echocardiography
TEE	Transesophageal echocardiography
PLSVC	Persistent left superior vena cava
UCS	Unroofed coronary sinus

References

1. Thangaroopan, M.; Truong, Q.A.; Kalra, M.K.; Yared, K.; Abbara, S. Images in cardiovascular medicine. Rare case of an unroofed coronary sinus:Diagnosis by multidetector computed tomography. *Circulation* **2009**, *119*, e518–e520. [CrossRef] [PubMed]
2. Kim, H.; Choe, Y.H.; Park, S.W.; Jun, T.G.; Kang, I.S.; Yang, J.H.; Eo, H.; Lee, H.J. Partially Unroofed coronary sinus: MDCT and MRI findings. *Am. J. Roentgenol.* **2010**, *195*, W331–W336. [CrossRef] [PubMed]
3. Murthy, A.; Jain, A.; El-Hajjar, M. Unroofed Coronary Sinus Presenting as Cerebral Abscess: A Case Report. *Cardiol. Res.* **2013**, *4*, 116. [CrossRef] [PubMed]
4. Ootaki, Y.; Yamaguchi, M.; Yoshimura, N.; Oka, S.; Yoshida, M.; Hasegawa, T. Unroofed coronary sinus syndrome: Diagnosis, classification, and surgical treatment. *J. Thorac. Cardiovasc. Surg.* **2003**, *126*, 1655–1656. [CrossRef]
5. Xu, B.; Betancor, J.; Cremer, P.C.; Renapurkar, R.; Pettersson, G.B.; Asher, C.R.; Rodriguez, L.L. Multimodality Cardiovascular Imaging of Unroofed Coronary Sinus Defects. *JACC Cardiovasc. Imaging* **2018**, *11*, 1027–1030. [CrossRef]
6. Watanabe, N.; Yanagita, Y.; Matsuura, H.; Nishino, S.; Nishimura, M.; Yano, M.; Ashikaga, K.; Kuriyama, N.; Shibata, Y. Unroofed coronary sinus detected by 2D/3D echocardiography in a patient referred to catheter ablation for atrial fibrillation. *J. Cardiol. Cases* **2016**, *14*, 111–114. [CrossRef]
7. Sun, L.; Juan, Y.H.; Chen, J.; Zhuang, J.; Xie, J.; Li, H.; Liu, H. Evaluation of Unroofed Coronary Sinus Syndrome Using Cardiovascular CT Angiography: An Observational Study. *Am. J. Roentgenol.* **2018**, *211*, 314–320. [CrossRef] [PubMed]
8. Raj, V.; Joshi, S.; Ho, Y.C.; Kilner, P.J. Case report: Completely unroofed coronary sinus with a left superior vena cava draining into the left atrium studied by cardiovascular magnetic resonance. *Indian J. Radiol. Imaging* **2010**, *20*, 215–217. [CrossRef] [PubMed]