remaining fourth aneurysm in the left side of the chest had increased from 3.7 cm to 5.1 cm over 6 months (Fig 2C, 2D). Attempts at embolization were again unsuccessful. The patient was taken to the operating room where the aneurysm was excised (Fig 3A, 3B). She again made an uneventful recovery and was discharged.

Comment

This is the first case report to document intercostal artery aneurysmosis. The patient had no connective tissue or systemic vascular disease, except that she had undergone endovascular repair of an infrarenal abdominal aortic aneurysm 3 years earlier. Patients with more than 1 intercostal artery aneurysm have associated diseases such as neurofibromatosis, coarctation of the aorta, or Kawasaki’s disease [2–5]. Although we expected to find neurofibromas along the intercostal nerves, this was not the case. In addition, close review of the computed tomographic scan of her chest did not reveal an undiagnosed coarctation. Intercostal artery aneurysm formation and hemorrhage have been associated with systemic lupus erythematosus and Ehlers-Danlos syndrome, but our patient had neither of these based on further testing [10, 11]. Because specific genetic testing for COL3A1 for vascular Ehlers-Danlos syndrome was not performed, there is a possibility that the patient carries these germline mutations. Testing for TGFBR1 and TGFBR2 for Loeys-Dietz syndrome was performed and was negative.

Interestingly, the aneurysms were along multiple different intercostal arteries, including the lower ones. With coarctation of the aorta, the aneurysms involve the upper intercostal arteries. There is a single case report in the literature of a mycotic intercostal artery pseudoaneurysm, but in our patient the fungal culture results were negative and she did not have any evidence of a systemic infection [12].

Endovascular management of these aneurysms has shown success in the small number of patients reported in the literature [2, 6, 8, 13–15]. We also had success using endovascular embolization to occlude flow into several of the aneurysms in our patient. Unfortunately, the feeding intercostal arteries tend to be elevated and stretched over the aneurysmal sac, which can make embolization difficult. Surgical repair was, and will remain, a safe alternative approach if embolization is not successful.

References

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Cystic Atrioventricular Node Tumor Excision by Minimally Invasive Surgery

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Cystic tumor in the atrioventricular (AV) node region is a rare cardiac primary tumor that can lead to sudden death. Our patient was a 43-year-old woman who presented with dyspnea and a first-degree AV block seen on electrocardiography. Echocardiography revealed a cystic mass attached to the inferior portion of the interatrial septum. After surgical excision of the mass, placement of a permanent pacemaker was required for complete heart blockage. Histopathologic examination revealed the mass to be a cystic tumor of the AV node region. To our knowledge, this is the first report of this condition.
diagnosed ante mortem and treated successfully with minimally invasive surgery.


Cystic tumors of the atrioventricular (AV) node are rare primary cardiac tumors derived from endoderm and located at the base of the atrial septum in the region of the AV node. These tumors can cause various degrees of heart blockage and are the smallest tumors of the heart capable of causing sudden death [1,2]. The precise incidence of these tumors remains unknown given that the cardiac conduction system receives scarce attention at autopsy. However, incidentalomas found at autopsy have been reported, suggesting that cystic tumor of the AV node can remain clinically silent. Ante mortem diagnosis and successful excision of this type of tumor are extremely rare [1–3]. We report a case of cystic tumor of the AV node region in which the tumor was detected preoperatively and was successfully excised with cardiopulmonary bypass through a right minithoracotomy.

A 43-year-old woman was admitted to our department with dyspnea and fatigue. A resting electrocardiogram and 24-hour Holter monitoring revealed sinus rhythm and first-degree atrioventricular (AV) block. Transthoracic and transesophageal echocardiography exposed a 22 × 21 mm right atrial hyperechogenic lesion with a broad connection to the interatrial septum. After inducing general anesthesia with a double-lumen endotracheal tube, a 14F venous cannula was placed percutaneously through the right jugular vein into the upper vena cava. The patient was positioned supine with slight elevation of the right hemithorax. A 6-cm port was made along the inframammary fold lateral to the midclavicular line. After heparin infusion, arterial and venous cannulation was performed in the right side of the groin. A 17F arterial cannula and a 21F venous cannula were introduced over a guidewire. Bypass was begun with bicaval venous drainage, and the patient was maintained in a normothermic state (36.3°C). The atrium was examined by transesophageal echocardiography to assess the site of tumor attachment. Without cross-clamping the aorta, the atrium was opened to reveal a 20-mm round cyst attached to the interatrial septum in the area of Koch’s triangle near the septal leaflet of the tricuspid valve. The cyst was completely resected and the atrium was closed. Throughout the procedure, a spontaneous rhythm with first-degree AV block was observed. The patient was easily weaned from cardiopulmonary bypass. Postoperative echocardiography showed a good left ejection fraction (64%) and trivial tricuspid and mitral valve regurgitation. A complete AV block appeared only on postoperative day 2. In agreement with cardiologists, the nodal rhythm ruled out the immediate implantation of a pacemaker in an effort to spontaneously restore sinus rhythm. Afterward, the persistent AV block and history of dyspnea and fatigue likely caused by the tumor led to our decision to implant a permanent pacemaker.

On gross inspection, the cut surface of the 2.5 × 1.7 cm nodule showed a cystic structure. It was processed for histologic examination with sections entirely along the minor transverse axis. At histologic examination, a cystic tumor of the AV node was found, consisting of a principal cyst lined by squamous, cuboidal, and focal transitional-like cells; other irregularly shaped microcysts of various sizes were present (Fig 1). The cysts were filled with

Fig 1. Gross examination and histologic findings of the cystic tumor of the atrioventricular node. (A) Tumor mass after resection. The epithelium is (B) squamous (hematoxylin and eosin; ×200), (C) cuboidal (hematoxylin and eosin; ×200) (arrow) or transitional-like (hematoxylin and eosin; ×200) (asterisk). (D) Small solid nests of squamous and sebaceous cells are shown (hematoxylin and eosin; ×400).
diastase-resistant proteinaceous material that stained positive with periodic acid-Schiff (Fig 1). They were surrounded by dense fibrous stroma showing a focal lymphocytic reaction and small solid nests of squamous and sebaceous cells (Fig 1). At immunohistochemical analysis, all the cells expressed wide-spectrum cytokeratins and epithelial membrane antigen; sporadic cells were positive for calcitonin and serotonin; staining was negative for calretinin (Fig 2) [3–6].

**Comment**

Cystic tumor of the AV node can be defined as a congenital multicystic lesion located at the base of the interatrial septum. The differential diagnoses can include bronchogenic cysts, ectopic thyroid (struma cordis), teratomas, and metastatic adenocarcinomas. The mean age at presentation is 38 years (range, birth to 78 years) and women are more frequently affected than men (approximately 3:1). Two thirds of patients present with complete heart block, 15% with lesser degrees of AV block, and 10% with sudden death even without a history of heart block. The literature reports tumor sizes varying from 0.5 mm to 30 mm. To make an ante mortem diagnosis, special mention should be paid to female patients with electrocardiographic evidence of heart block with narrow QRS complexes (limited to the AV node). The cause of lethal arrhythmia in patients with cystic tumor of the AV node region is still controversial, without a clear relationship between tumor size and the occurrence of arrhythmia. Hypotheses that explain why lethal arrhythmia occurs are based on excessive distention of the ventricle with subsequent ventricular fibrillation. Furthermore, postmortem studies have shown that pacemaker implantation does not prevent sudden death in patients with heart block caused by this type of tumor. Thus, surgical intervention should always be indicated.

To our knowledge, only 5 ante mortem diagnoses and resections of cystic tumors of the AV node region have been reported [7,8]. In our case, the anterior minithoracotomy approach was an excellent route for the right atrium and provided us with a satisfying field of view.

In conclusion, because the worst complication with this tumor is sudden death, we believe that complete resection is essential, even if subsequent pacemaker implantation becomes necessary.

**References**