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RIGHT ATRIAL BENIGN SCHWANNOMA

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Primary schwannoma of the heart is a rare disease. It arises from vagus nerve branches and plexus. Most schwannomas are benign tumors, but sometimes they can be primary malignant neoplasms. In the MedLine database, we found only 21 publications on benign and 13 publications on primary malignant cardiac schwannoma. Moreover, according to localization in the right atrium, only eight benign schwannoma observations are described. We report a 73-year-old woman who, with echocardiography and magnetic resonance imaging of the heart, revealed a right atrial tumor with sprouting of the right atrial free wall. The tumor was radically excised through cardiopulmonary bypass and pharmaco-cold cardioplegia. Pathohistological and immunohistochemical examination of the excised tumor showed that it is benign schwannoma.

Keywords: schwannoma, cardiac tumor, malignant neoplasms.

INTRODUCTION

The heart schwannoma (also known as neurilemmoma, neurinoma) is an extremely rare tumor. It develops from Schwann cells producing myelin and located in the peripheral nerve sheaths [1]. In the heart, the schwannomas are formed from the cells of the membranes of the cardiac plexus nerves and the nervus vagus branches [2]. One of the first reports of benign schwannoma, which grew from the wall of the right atrium, was published in 1972 [3]. We have found only 21 publications on benign schwannomas of the heart and 13 publications on primary malignant ones in the MedLine database. Besides, benign schwannomas in the right atrium are described only in eight observations. The purpose of the present article is to describe such a rare pathology as a benign right atrial schwannoma which has been removed through a radical surgery in our center.

CASE STUDY

Patient K., female, aged 73, was admitted to our Center on January 30, 2019 complaining of fatigue, increased blood pressure and palpitation episodes. The medical history showed that the examination at the place of residence for hip joint pain revealed a neoplasm in the right atrium. The cardiovascular surgeon recommended an operative therapy. Concomitant and previous diseases: hypertension of the 3rd degree, stage 3, risk 4; crystalline humors replacement in both eyes (2012), appendectomy.

At the admission, the patient's condition is of moderate severity. Clear consciousness. The skin and visible mucous membranes are of normal color. Vesicular breathing is heard throughout the full lung field, no rales. BP – 140/80 mm. Hg. Heart tones are muffled, the heart rhythm is regular, the heart rate is 60 beats per minute. A slight systolic murmur is heard over the heart region. The liver is not enlarged, the spleen is non-palpable. The CVA tenderness of the lumbar region is not manifested on either side. The bowel and bladder functions show no abnormalities. No edema.

ECG: sinus rhythm, heart rate – 60 beats per minute.

The plain chest X-ray showed no focal and infiltrative lesions; the phrenicocostal sinuses are free, the heart shadow is not enlarged. Coronary angiography: the right type of the myocardium blood supply, no hemodynamically significant lesions of the coronary arteries. Echocardiography revealed a hyperechogenic neoplasm in the right atrium, fixed to the lower third of the atrial septum. MRI (30.01.2019) of the right atrium cavity revealed an additional fixed neoplasm between the inferior vena cava entry and the coronary sinus, while the results of multispiral computed tomography of the heart showed signs of a moderate accumulation of the Ultravist 370 contrast (Fig. 1). A neoplasm of 2.5 × 1.5 × 2.7 cm is attached to the lower third of the atrial septum with a base of 9 mm, deforming and narrowing the entry of the inferior vena cava and the coronary sinus (Fig. 2, 3).

Preoperative clinical diagnosis: neoplasm of the right atrium; circulatory deficiency II A; hypertension of the 3rd stage, degree 3, risk 4.

Intraoperatively, a moderate amount of clear serous fluid was found in the pericardial cavity. The ascending aorta diameter is 3.4 cm, the pulmonary trunk diameter of up to 3.2 cm. The pulmonary veins typically enter the left atrium. CPB was connected by the "aorta–vena

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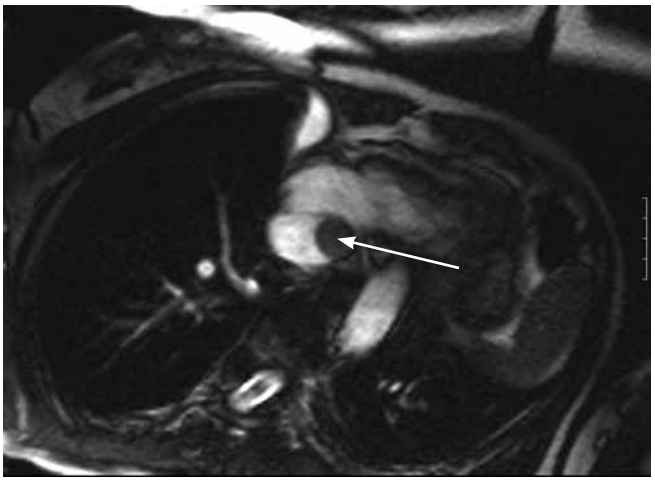

Fig. 1. Signs of moderate accumulation of a contrast agent in tumor (arrow) located in the cavity of the right atrium between the inflow of the inferior vena cava and the coronary sinus. Multislice computed heart tomography

cava" scheme. Start of artificial blood circulation. The tourniquet loops are placed on the vena cava. Spontaneous cooling to 34.6 C. Aortic clamp. Cardioplegia to the aortic root, Custodiol, 2,000 ml. The right atrium was opened with a longitudinal incision. The revision of the right atrium between the inferior vena cava entry and the coronary sinus showed a dense neoplasm, whitish, 3 × 4 cm, with a wide base of 2.5 × 3 cm, encapsulated, covered by the endocardium and tightly connected with the lateral surface of the coronary sinus, interatrial septum and the free lower wall of the right atrium. The neoplasm narrows the coronary sinus and the entry to the inferior vena cava.

Fig. 4 shows a neoplasm of the right atrium deforming the coronary sinus. It was removed through resection of the endocardium at the place of its attachment. Also the partial resection of free wall of the right atrium

Fig. 2. Compression of the inferior vena cava by a neoplasm of the right atrium. Multislice computed heart tomography

was performed at the neoplasm extension site (Figs. 5, 6). A slit-like defect of the free lower wall of the right atrium of 3 × 1.5 mm formed. The defect is sutured with Teflon gaskets. The plastic repair of the endocardial resection area was performed with a xenopericardial patch. The incision of the right atrium wall was sutured tightly with a double-row continuous twisted suture (Prolen 4/0). The patient was warmed. Tourniquets on the vena cava were loosened. The aortic clamp was removed. Restoration of the cardiac activity with a single defibrillator discharge; sinus rhythm with a heart rate of 75 beats per minute. With the satisfactory hemodynamics parameters, artificial circulation was stopped. Decanulation of the lines in the machine for artificial blood circulation. Hemostasis.

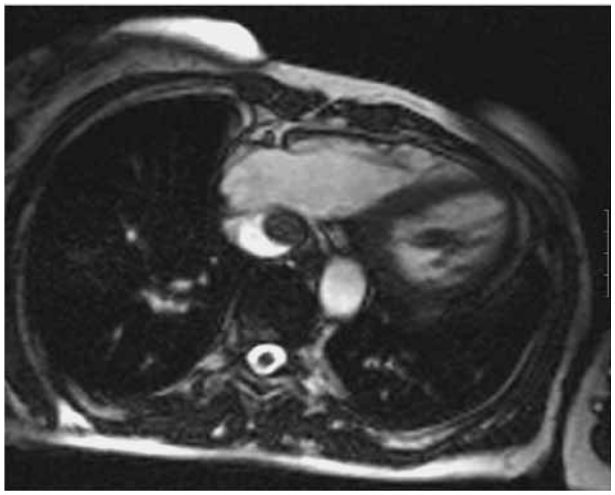

Fig. 3. Compression of the coronary sinus by a neoplasm of the right atrium. Multislice computed heart tomography

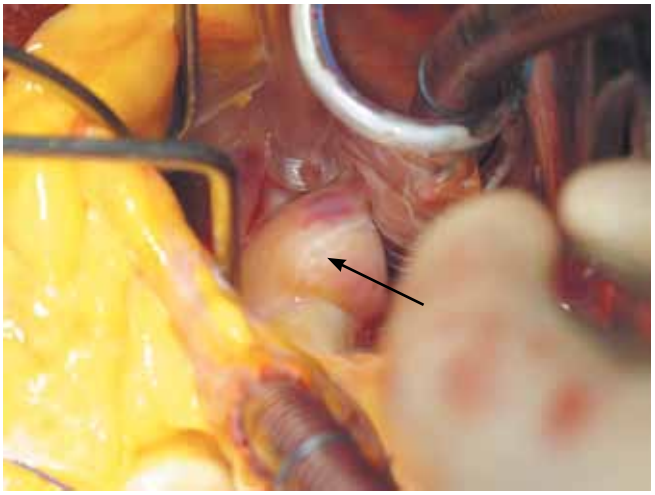

Fig. 4. Neoplasm of the right atrium fixed to the atrial septum and the free wall of the right atrium

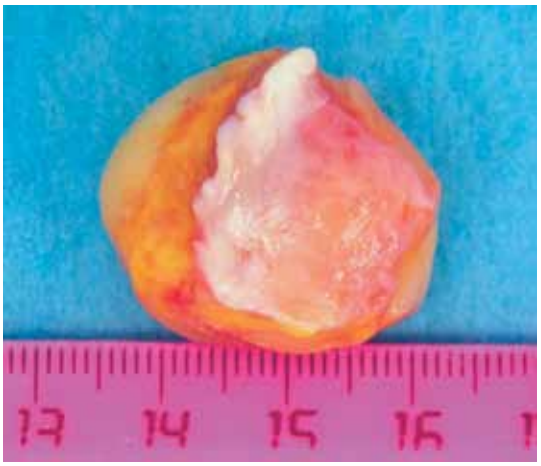

Fig. 5. Resected mass of the right atrium

Fig. 6. Neoplasm of the right atrium in the section

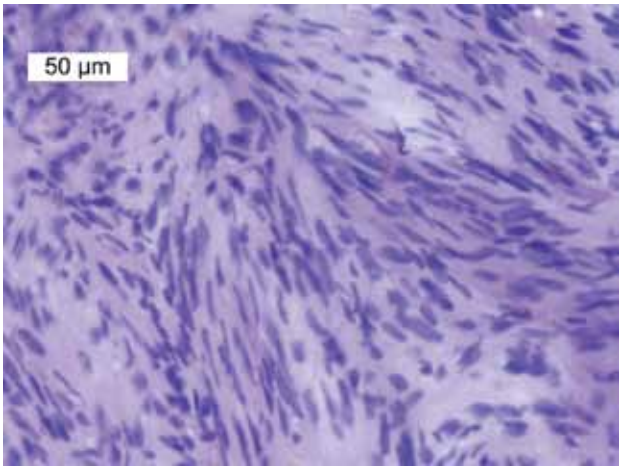

Fig. 7. Fragment of the shwannoma Antoni A (H&E stain, ×400)

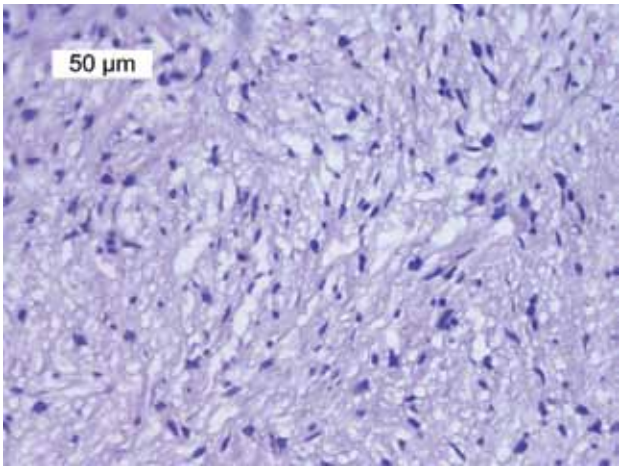

Fig. 8. Fragment of the shwannoma Antoni B (H&E stain, ×400)

The pericardium was sutured with a continuous twisted suture in the upper and middle thirds. The sternum was closed with four 8-shaped wire ligatures. Hemostasis. Layered suturing of the wound. Aseptic sticker.

The remoted neoplasm of whitish-pink color was densely elastic and sized 3 × 2 × 2 cm, having a smooth surface, in the section it was of a nodal structure and whitish-yellow color (Fig. 5, 6).

Morphological examination with histological staining (hematoxylin and eosin, trichrome Masson). The expression of S100 protein, vimentin, CD34, desmin, smooth muscle actin, MyoD1, Ki 67, GFAP was studied with the immunoperoxidase method (preparations were provided by I.B. Kaplanskaya, a leading scientist of the Pathology and Anatomy Department at P.A. Herzen Moscow Cancer Research Institute, Ph.D.).

The histological examination showed that the tumor was a benign mixed-structure schwannoma – Anthony type A and type B. In A type sections, the tumor cells were arranged in dense blocks, their nuclei were elongated and hyperchromatic (Fig. 7). In B type sections, the tumor cells were located less densely, their nuclei were polymorphic, slightly elongated, rounded, oval, and irregular in shape (Fig. 8). Multiple thin-walled

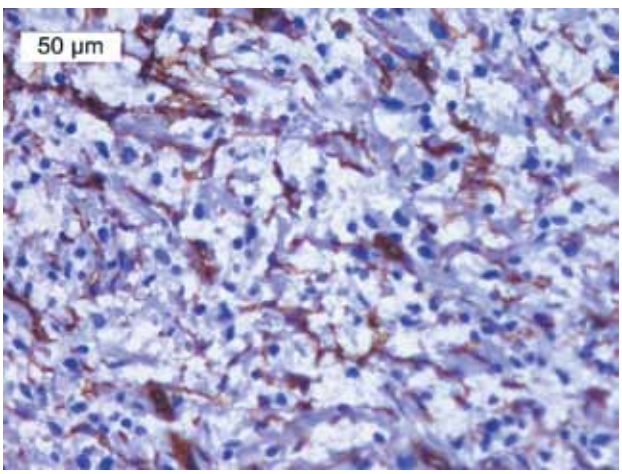

Fig. 9. The blood supply to the tumor was provided by numerous blood vessels whose visualization is well achieved by staining with the marker CD34. Immunoperoxidase method

vessels and muscle-type arteries provided for the blood supply to the tumor (Fig. 9). A nerve trunk was located deep in the tumor (Fig. 10), Schwann cells resulted in the formation of a schwannoma.

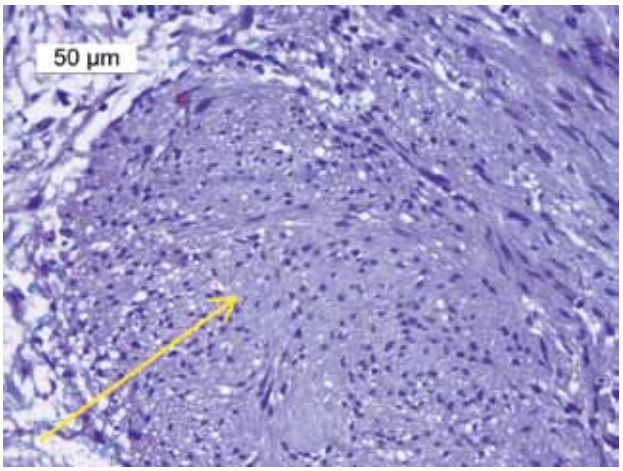

Fig. 10. Nervous trunk in schwannoma (arrow) (Masson's trichrome stain)

The immunohistochemical examination confirmed the diagnosis. The tumor manifested diffuse staining of S100 protein (Fig. 11) and vimentin (Fig. 12). There was no desmin or smooth muscle actin in tumor cells, but it produced positive staining in the arterial wall of muscle type (Fig. 13, 14). The reaction with antibodies to MyoD1, Ki 67 and GFAP (glial fibrillary acidic protein) were negative.

No complications in the postoperative period. The surgical wound was healed by the first intention. Echocardiography showed no pathological masses in the heart cavities. Normokinesis. No pathological flows on the interatrial septum. The patient was discharged in a stable condition for the follow-up by a cardiologist on the 8th day after surgery.

Final clinical diagnosis: benign right atrium schwannoma.

DISCUSSION

Schwannomas of the heart are extremely rare [4]. Therefore, the present study of a benign schwannoma

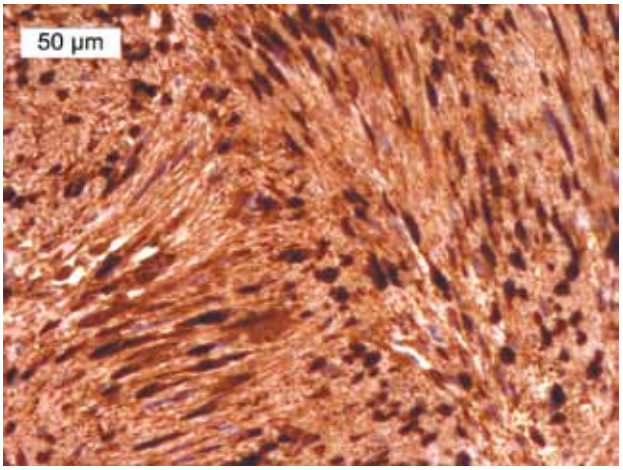

Fig. 11. Intensive positive reaction to S100 protein (×400)

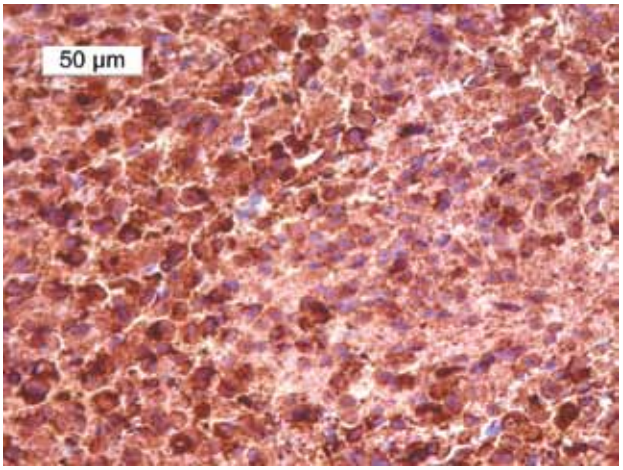

Fig. 12. Intensive positive reaction to vimentin (×400)

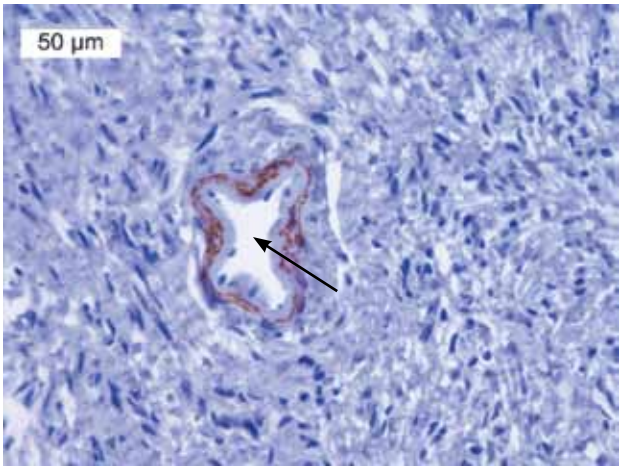

Fig. 13. Positive reaction to desmin in the muscle-type arterial wall of the schwannoma (×400)

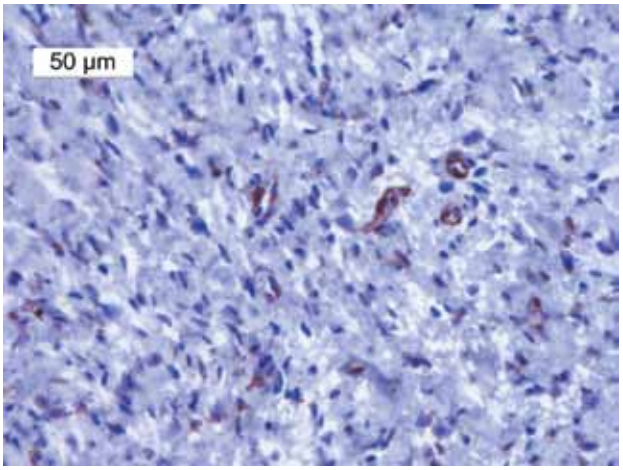

Fig. 14. Positive reaction to smooth muscle actin in the vessel wall of schwannoma (×400)

in the right atrium between the inferior vena cava entry and the coronary sinus is of great interest. The neoplasm narrowed the coronary sinus and the inferior vena cava entry, however, there were no expressed symptoms, and the tumor was discovered accidentally during the examination.

Our study of schwannomas in the right atrium is the ninth of the available sources published (Table). The right atrium is a typical location of the heart schwannomas [5]. The age of patients ranged from 33 to 72 (Table), and in our study, the patient was 73.

Table

Benign schwannomas of the right atrium

№	Author, publication date	Localization	Gender	Age
1	T.H. Gleason et al., 1972	Right atrium	–	–
2	B. Monroe et al., 1984	Right atrium	M	70
3	F. Bizzarri et al., 2001	Right atrium	M	72
4	D.S. Jassal et al., 2003	Right atrium	F	49
5	K. Nakamura et al., 2003	Right atrium	F	33
6	N.A. Stolf et al., 2006	Right atrium	F	56
7	S.A. Early et al., 2007	Right atrium	M	57
8	S. Koujanian et al., 2017	From the right atrium to the right ventricle	F	47
9	Observation	Right atrium	F	73

The size of the schwannoma in this study was small (3 × 2 × 2 cm), although such tumors can reach a giant size (12 × 8 cm) [6]. Despite its modest size, the schwannoma is visualized in ECG, CT or MRI, which may be associated with the density of its structure [7]. However, a definitive diagnosis can only be established based on histological and immunohistochemical tumor tests [8]. The intense staining of the S-100 protein confirms the diagnosis of benign schwannomas [5], and a positive response to CD56 shows the malignant nature of the tumor [9].

Radical tumor resection is necessary to prevent life-threatening complications, such as compression of vital structures, embolism, valve obstruction and sudden death [6]. In our study, the patient underwent successful complete removal of neoplasm in the right atrium within healthy tissues and plastic reconstruction of the atrial septum and the free wall of the right atrium with a patch of xenopericardium. The patient has an excellent prognosis, as schwannoma proved to be benign.

The authors declare no conflict of interest.

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