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Primary malignant peripheral nerve sheath tumour of the heart

Case report

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Abstract: A malignant peripheral nerve sheath tumour (MPNST) is a rare variety of soft-tissue sarcoma of ectomesenchymal origin. The World Health Organisation created the term MPNST to replace previous terminology such as *malignant schwannoma, malignant neurilemmoma, neurogenic sarcoma*, and *myxofibrosarcoma* for tumours of neurogenic origin with similar biological behavior [1-3]. The vast majority of these tumours develop in extremities. They also tend to be located in unusual sites of the body, such as the pelvic retroperitoneum, infratemporal fossa, intrapericardium, and mediastinum [1,3,4]. This case study presents a patient with an extremely rare primary cardiac MPNST.

Keywords: *Malignant peripheral nerve sheath tumour*

1. Case presentation

A 44-year-old woman was referred to the Cardiology Department with heart rhythm disturbances. An electrocardiogram showed atrial fibrillation and a ventricular rate of 124–174 beats per minute. An echocardiographic study found a very large tumor mass in the left atrium. The lower edge of the tumor reached out to the mitral valve opening and mitral valve anterior leaflet was touching the tumor in systole. On the visible surface of the anterior leaflet another formation was seen. The reading suggested that tumor transcended the anterior mitral valve leaflet. A second degree of mitral valve regurgitation was recorded (Figure 1). Cardiovascular magnetic resonance (CMR) imaging revealed a large, irregularly shaped left atrial mass measuring 7.0x6.0 cm. The mass was composed of two formations, one of which had invaded the posterior atrial wall. The second formation invaded and occluded the left superior pulmonary vein (Figure 2). In the tumor's structure foci, calcification were visible. Mitral valve mobility and structure were normal – the tumor did not affect the mitral valve. In both pleural cavities the presence of liquid was seen. There were no bronchial pressure symptoms. The patient underwent an extensive body examination, and the results showed no significant change. Therefore, it was concluded that the tumor in the left atrium was of primary origin without metastases, and surgery was scheduled.

A left anterior lateral thoracotomy was performed. A significant volume of fluid was aspirated upon opening the pleural cavity. The pericardium was normal and no pericardial adhesion or fluid was observed. The left atrial appendage was free, but the tumor had spread to its base, conjoined the posterior wall of the left atrium,

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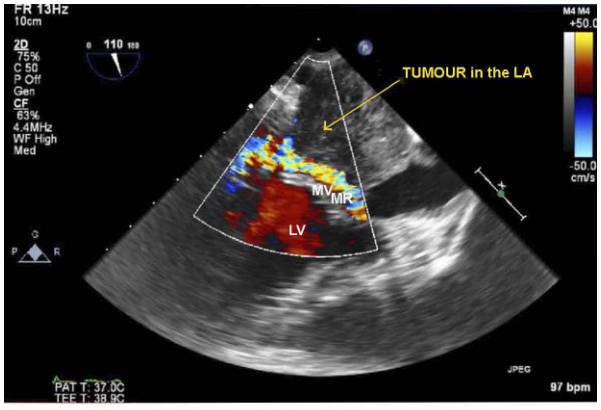

Figure 1. Two-dimensional trans-oesophageal echocardiography of the tumor in the left atrium (LA). LV, left ventricle; MV, mitral valve; MR, mitral regurgitation.

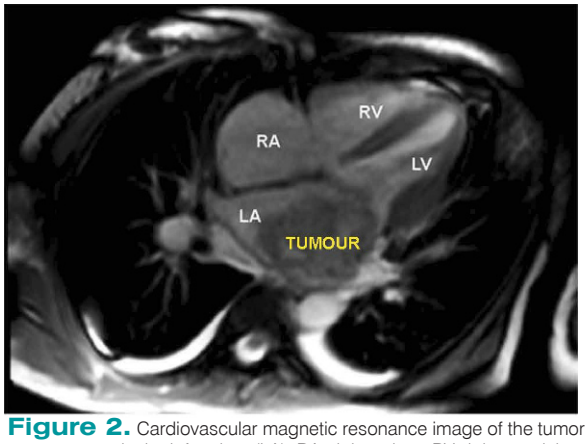

Figure 2. Cardiovascular magnetic resonance image of the tumor in the left atrium (LA). RA, right atrium; RV, right ventricle.

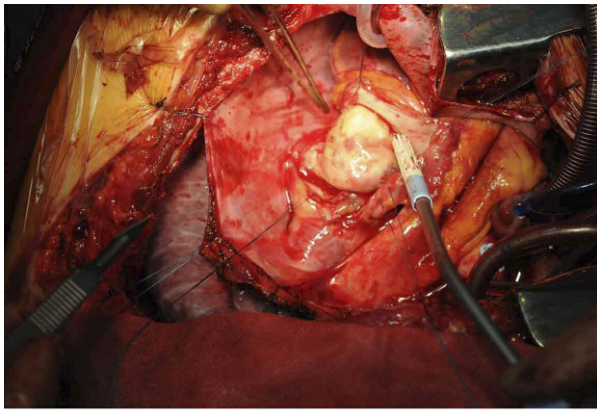

Figure 3. Intraoperative image of the tumor in the left atrium.

and extended to the left superior pulmonary vein. The left atrium was opened, and a white brilliant tumor was found (Figure 3). Closer inspection revealed that the tumor consisted of two conjoined oval formations occupying the left atrial cavity. Although ultrasound had shown damage to the mitral valve anterior leaflet, the valve appeared normal. It was concluded that the tumor had spread from the left superior pulmonary vein. The

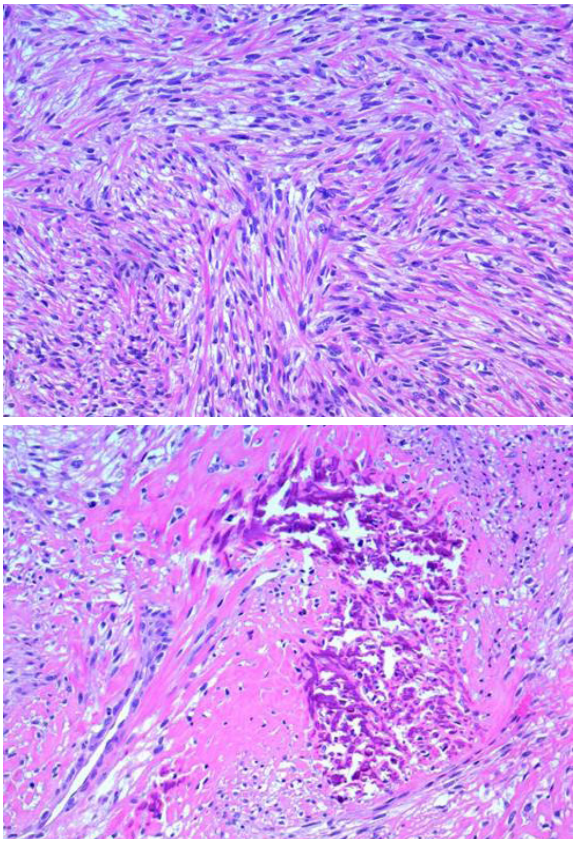

Figure 4,5. A histological examination of the tumor confirmed the diagnosis of MPNST

vein was completely closed by the tumor. The tumor was resected macroscopically radical. Resection margin was left atrium, reconstruction of lower lung vein and resection of upper left lung vein together with upper left lobectomy, microscopically (after examination of pathologist) tumor was found in the field of resection of upper left lung vein. Size of resected tumor was 9.0x4.5x4.5 cm and two more tumors in the upper left lung vein 4 cm diameter. The free wall of the left atrium was reconstructed using a pericardial patch that measured 3.0x4.0 cm. Cardiopulmonary bypass time was 177 minutes, aortic clamping time was 87 minutes, and the lowest temperature was 28°C. A histological examination of the tumor confirmed the diagnosis of MPNST (Figure 4, 5).

Following seven days in intensive care unit (due to hypotension, heart rhythm disorder) the patient was transferred to hospital ward. In a few weeks the patient was well enough and discharged for further rehabilitation treatment. Adjuvant treatment started from postoperative radiotherapy, because within two months after surgery, chest computed tomography and an echocardiographic examination showed local recurrence of the tumor, with no distant metastasis. Postoperative radiotherapy 66 Gy was given to the patient, because that reoperation of recurrence was impossible. Lung metastases were

found within 6 months after surgery. The patient received 6 cycles of Ifosfamide. Tumor recurrence finally decreased as a result of this course of treatment. The patient had no major complaints, only slight pain in the field of operation incisions and arrhythmia.

2. Discussion

A malignant peripheral nerve sheath tumor is a very rare tumor with an incidence of 1 per 100 000 population and represents between 3 to 12% of all soft tissue sarcomas [1,3]. Typically this tumor has spread from a major or minor peripheral nerve branches or sheath of peripheral nerve fibers. Therefore from 5% to 42% of MPNSTs have an association with multiple neurofibromatosis Type-1 [1,3]. Another interesting clinical feature of this tumor is the multifocality and development of second primary tumors of same histology – as a s*chwannoma, ganglioneuroma*, or *pheochromocytoma* [1]. However, most cardiac MPNSTs seem to spread sporadically [1,3,4,7]. As well as in our case the patient had no features of neurofibromatosis Type-1 also there was not any primary tumor.

Cardiac tumors manifest themselves with a variety of symptoms. These are related to obstruction, embolism, myocardial invasion and damage. In this case study, the main symptom was paroxysmal atrial fibrillation as the tumor spread into the left atrium. Several studies have shown that together with the pulmonary veins, many extrapulmonary vein areas such as left atrial appendage and left atrial posterior wall may be the source of initiation and maintenance of atrial fibrillation [8].

Often MPNST in structure has heterogeneous elements, such as, bone, and typically form endoluminous masses [4,9]. According to localization cardiac MPNST most frequently found in the left atrium [4,7,9].

Echocardiography is the initial and of course the main study confirming the diagnosis of cardiac tumor. CMR reveals the tumor tissue-specific and sufficiently accurately define their boundaries as well as to diagnose the presence and extent of local and distant metastases. However, only histological examination can differentiate between tumor morphology and confirm the final diagnosis of MPNST [2,4,9].

There is no doubt, a radical surgical resection is the treatment of choice in the treatment of MPNST. Currently, all research on cardiac sarcomas report an improved rate of survival after the tumor is radically removed. In addition, it is usually required to for the reconstruction of cardiac chambers [10-15]. Therefore, this is best achieved when tumors are encapsulated and well demarcated [16-19]. However, most research note local recurrence of tumor or metastasis from 2 to 10 months following the operation. Early recurrence of the tumor has been recorded as a poor prognostic factor for overall survival [3,4,9]

The cases of cardiac transplantation, when primary cardiac tumors are present, are limited as it is difficult to determine the operability of the tumor before surgery.

Unfortunately, MPNST is the most pathological of all cardiac sarcomas. Median survival ranges from 3 months to 1 year and prognosis correlates with the histological grading, on tumor location and rate of growth [2,4,6] Adverse prognostic factors include large size (>5 cm), high-grade tumor, advanced histological characteristics, surgical margin with tumor invasion, and neurofibromatosis Type-1 [3,9].

In this case study, the tumor had recurred just after more than two months and metastasis in the lung developed after six months following the surgery.

Therefore, primary MPNST of the heart is a highly aggressive tumor which has the highest recurrence rate of any other sarcoma. The prognosis for patients with such cardiac tumor is poor and the median survival ranges from 2 to 12 months [1,3,4,9].

Currently, postoperative radiotherapy is recommended by the Oncology Consensus Group as part of a uniform treatment policy for MPNST [1]. Due to the rare incidence of MPNST, large trials of the effectiveness of chemotherapy in MPNST are impossible and most current data are based on case reports, small case series, or regimens proven to be successful for other soft tissue sarcomas.

3. Conclusion

This is a very rare case of primary cardiac MPNST, rising from upper left lung vein penetrating into the left atrium. Treatment of these patients is challenging and of course treatment strategy should be controversial in multidisciplinary team. In case of local spread, with no distant metastasis, surgery should be a part of treatment along with chemoradiation.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare that they have no competing interests.

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