Schwannoma of the phrenic nerve. A case report

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Background. Neurogenic tumors are the third most common tumors occurring within the mediastinum. Predominantly completely asymptomatic, they are most often found in the posterior mediastinum, although they may also be located, albeit rarely, in the middle mediastinum. Thus, in the cases of tumors localized in the middle mediastinum, schwannomas of the phrenic nerve must always be considered.
Case Report. In this case, a male patient presented with a tumor of the middle mediastinum. PET/CT scan determined that it was a circumscribed tumor without signs of dissemination. However, due to the tumor's location, a preoperative biopsy of the tumor was not possible. Therefore, the patient underwent videothoracoscopic extirpation of the tumor, including the necessary resection of the phrenic nerve that passed through the centre of the tumor. Subsequent histological analysis definitively confirmed a schwannoma with low proliferative activity.
Conclusions. Differential diagnosis of mediastinal tumours is very difficult; however, in the case of circumscribed tumours not invading the surrounding tissues, proceeding to surgical revision, with the complete removal of the tumour, is possible, even without determining the histological nature. Minimally invasive surgical techniques dominate the treatment of neurogenic tumors of the mediastinum; they are associated with minimal complications and allow a rapid return of the patient to their normal activities.

Key words: schwannom, nervus phrenius, mediastinum, videothoracoscopic resection, neurogenic tumors

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INTRODUCTION

Neurogenic tumours of the mediastinum represent the third most common group of mediastinal tumours, after thymomas and lymphomas, and account for about 20% of total cancer incidence. From the perspective of incidence according to localisation, they occur most frequently in the region of the posterior mediastinum – in about 75–90% (ref.[1,2]). The most common site of occurrence is along the costovertebral sulcus[3-5]. In recent literature, there are only a few reported cases of a schwannoma of the phrenic nerve[6]. Here we present a patient case with an incidental finding of a schwannoma of the phrenic nerve in the area of the middle mediastinum. The tumor was completely removed using a minimally invasive videothoracoscopic technique.

METHODS AND RESULTS

A 51-year-old male patient was referred to our department with an incidental finding of a right mediastinal mass, discovered as a result of a lung X-ray which was part of a preoperative examination, prior to arthroscopic surgery of the right knee. The patient was otherwise clinically uncomplicated. A contrast enhanced CT scan of the chest was then performed and showed a 62x43 mm tumor in the right pleural cavity arising from the middle mediastinum (Fig. 1). A preoperative biopsy was not possible due to the location of the tumor. For this reason, a whole-body PET/CT scan was performed, as a part of the differential diagnosis and to exclude generalisation of the disease. There was an increased accumulation of glucose

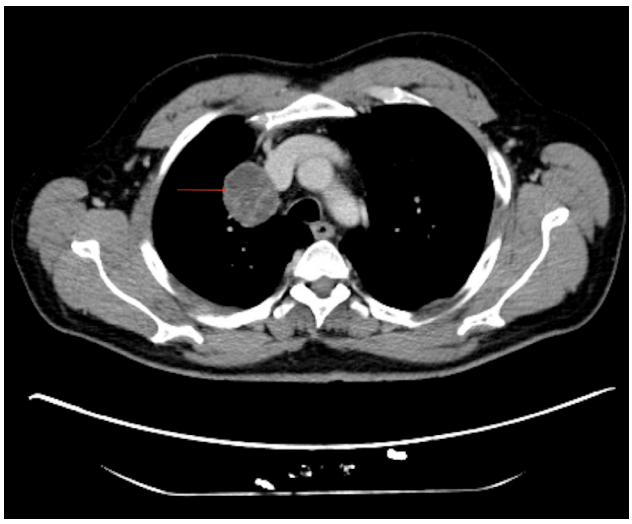

Fig. 1. Chest CT with middle mediastinal tumor.

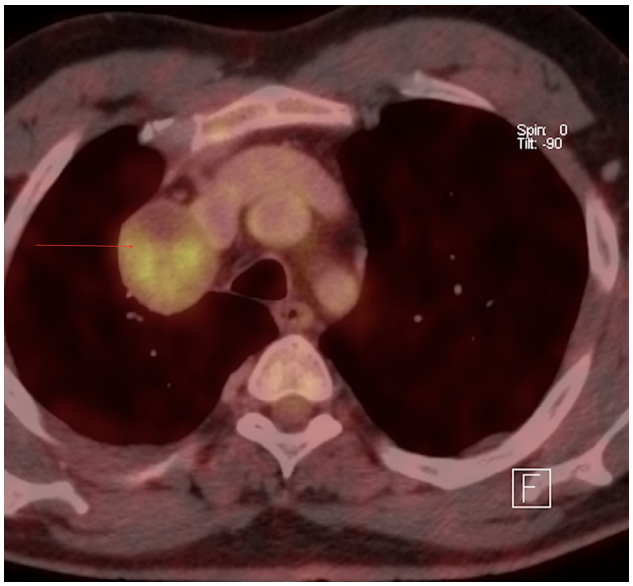

Fig. 2. PET/CT scan.

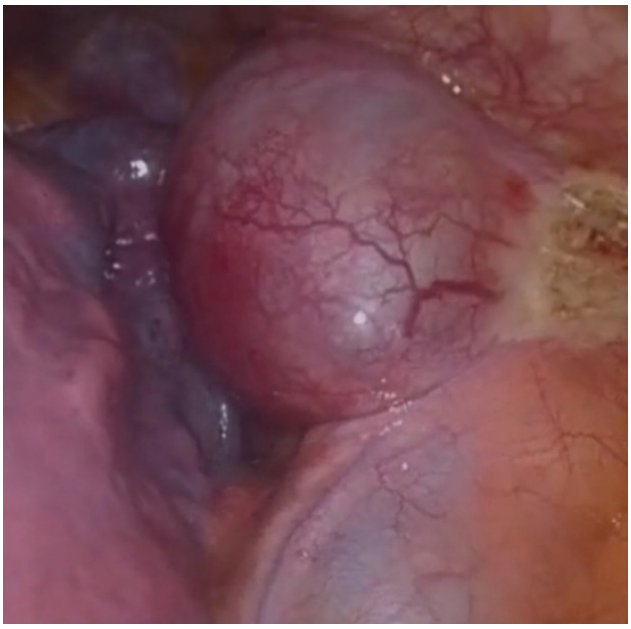

Fig. 3. Intraoperative finding of a schwannoma of the middle mediastinum (alternative: Intraoperative finding of the middle mediastinum schwannoma).

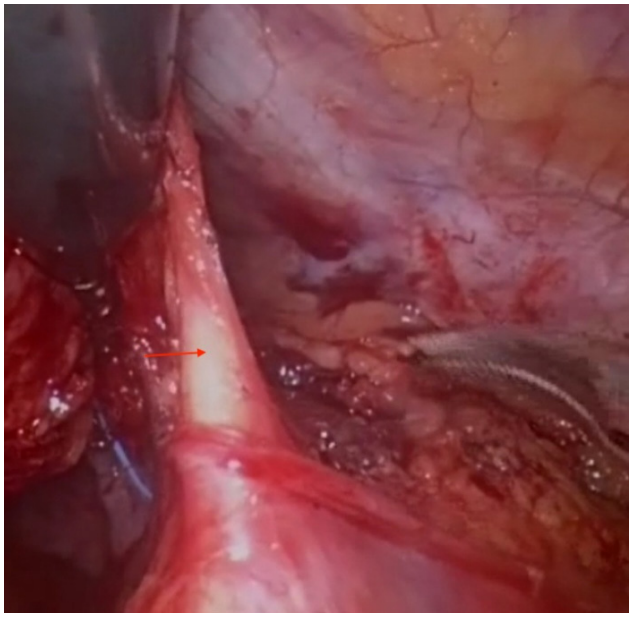

Fig. 4. Phrenic nerve going to the centre of the tumor.

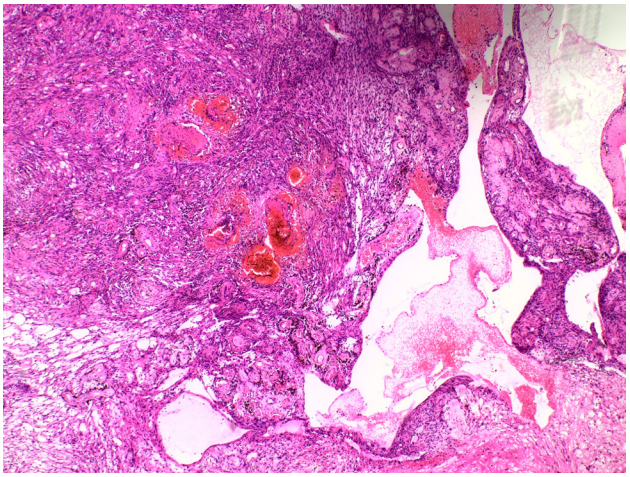

Fig. 5. Regressive changes.

in the mass, which indicated the presence of viable tumor tissue (Fig. 2). Circulating oncomarkers from the patient's blood were investigated, and a preoperative pulmonary examination was completed to assess respiratory functions, which were intact.

The patient was indicated for a minimally invasive videothoracoscopic tumor extirpation. A relatively large, circumscribed tumor was found in the pleural cavity (Fig. 3), with an unexpected discovery of the right phrenic nerve entering the tumor mass. Due to a high risk of malignancy, extirpation of the tumor along with the corresponding phrenic nerve was performed (Fig. 4). The tumor was extracted in an endobag, having previously been divided into two parts for easier extraction via a minithoracotomy. The postoperative period was uneventful and without complications, and the chest drain was removed on the second postoperative day. Elevation of the right diaphragm, as a result of the resection of the phrenic nerve, was already visible on the control radiograph. The patient was discharged to home care on the sixth postoperative day, without breathing difficulties, which may have been a possible complication following the resection of the right phrenic nerve. Histological findings confirmed the diagnosis of a schwannoma of the phrenic nerve, with S100 protein positivity, significant regressive changes, central disintegration and proliferative activity of Ki67 in only 5% of tumor cells (Fig. 5). The patient is participating in follow-up care at the pneumo-oncology clinic at regular intervals, with control CT examinations, and there are no signs of recurrence. The patient's lifestyle and quality of life has not been negatively affected by the paresis of the right diaphragm.

DISCUSSION

Neurogenic tumors of the mediastinum most often occur in the posterior mediastinum, but in rare cases they can also occur in other locations. Malignancy of these mostly benign tumors is very rare in adults; it is significantly more frequent in children[3,4]. The posterior mediastinum is rich in nerve fibres, which is why these tumors most frequently occur here, where they arise from peripheral and especially intercostal nerves, from ganglia and paraganglia of the sympathetic or vagus nerve, and rarely, more ventrally, from the phrenic nerve. In the available literature, schwannoma of nervus phrenicus is only described in a few cases, and it is, like other neurogenic mediastinal tumors, mostly asymptomatic[6]. Certain nonspecific symptoms such as chest or back pain are rarely mentioned in the literature. Very rarely, specific symptoms such as dyspnea due to diaphragmatic paresis and medically uncontrollable singultus may appear[7,8].

The histopathological analysis of a given tumor is the basis of establishing a definitive diagnosis. Preoperative transparietal biopsy helps to confirm the diagnosis and should therefore be performed, wherever possible, as a part of the differential diagnosis. A CT scan of the chest is required as standard practice, and preferably would be followed by a full body PET/CT scan to determine the possible presence of distant metastases in the case of an aggressive form of tumor. A concurrent examination of oncomarkers in the serum is necessary, and in male patients, alpha-fetoprotein and human chorionic gonadotropin must not be omitted in order to rule out malignant germ cell tumors. In the case of circumscribed, surgically removable tumors, it may not always be necessary to perform a preoperative biopsy. In most cases, minimally invasive surgical techniques are the predominant treatment method[9]. The only limitations of the procedure may be the size of the tumor which, according to the literature, would need to exceed 6cm in size, and the additional factor of an unclear anatomical relationship between the tumour and the intrathoracic organs. Both of these factors would need to be considered[10]. One of the first robotic operations in thoracic surgery was for neurogenic tumors. The primary advantages of robotic surgery include perfect orientation in confined spaces, such as the mediastinum, and a low risk of possible complications[11].

According to the WHO, schwannomas are classified as Grade 1 tumors; benign and slow growing associated with long term patient survival. However, no staging system has been created. Recurrences of these tumors are uncommon and they almost exclusively occur in patients with Recklinghausen's disease. Malignant schwannomas may also occur in these patients, therefore regular monitoring is required[12-14]. In the case of schwannomas, extirpation without breaking the tumor capsule using a minimally invasive surgical technique is entirely sufficient. Aggressive neurogenic tumors require a radical operation with resection of the nerve and surrounding tissue with a sufficient safety margin[13]. There is one reported case of a malignant schwannoma of the phrenic nerve in literature[15]. In our case, according to the definitive histological analysis, it was a tumor with a low degree of malignancy; however, as the nerve ran through the centre of the tumor, a radical resection was performed. This was feasible due to the patient's above average respiratory parameters. In the subsequently monitored postoperative period the patient was able to return to his sports activities, without respiratory limitation during physical exertion.

CONCLUSION

Neurogenic mediastinal tumours occur most often in the area of the posterior mediastinum, only rarely are they localized in the middle mediastinum. In the majority of cases they present as a schwannoma of the phrenic nerve. Minimally invasive surgical techniques dominate the therapy of these tumors, due to both safety reasons and to minimize possible complications.

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