

Primary malignant peripheral nerve sheath tumor of the pleural cavity: rapid progression

Piotr Misiak, Szymon Wcislo, Sławomir Jabłoński, Katarzyna Szwalbe

Military Medical Academy University Teaching Hospital – Central Veterans' Hospital in Lodz, Lodz, Poland

Kardiochirurgia i Torakochirurgia Polska 2014; 11 (2): 213-215



Abstract

Malignant peripheral nerve sheath tumor (MPNST) accounts for 5-10% of all malignant soft tissue tumors. It often affects the extremities and abdominal cavity and very rarely develops in the thoracic cavity. The incidence in the general population is 0.001%. In our paper we would like to present a case report of a 74-year-old female patient with primary MPNST of the pleural cavity which was previously misdiagnosed as a non-small cell lung cancer. Despite the combined treatment, after four months the patient developed a secondary tumor in the contralateral pleural cavity which was also treated operatively.

Key words: pleural cavity, peripheral nerve sheath tumor.

Streszczenie

Złośliwe nowotwory osłonek nerwów obwodowych (MPNST) stanowią ok. 5–10% mięsaków tkanek miękkich. Występują najczęściej w obrębie tkanek miękkich kończyn i w jamie brzusznej, niezwykle rzadko ich pierwotną lokalizacją są struktury położone wewnątrz klatki piersiowej. Częstość występowania w populacji ogólnej wynosi 0,001%. W niniejszej pracy przedstawiono przypadek 74-letniej chorej z pierwotnym guzem typu MPNST zlokalizowanym w obrębie jamy opłucnej, który na podstawie badań wstępnych został mylnie zakwalifikowany do zabiegu resekcyjnego jako niedrobnokomórkowy rak płuca. Pomimo zastosowanego leczenia skojarzonego po 4 miesiącach u chorej wykryto wtórny guz metastatyczny w przeciwległej jamie opłucnej, który został ponownie resektowany.

Słowa kluczowe: jama opłucnej, guz osłonek nerwowych.

Introduction

Primary chest wall tumors constitute approximately 0.2-2% of all tumors. More common in this localization are metastatic tumors. Primary malignant peripheral nerve sheath tumor (MPNST) situated in the pleural cavity is an extremely rare clinical entity. This type of tumor, also known as “malignant Schwannoma”, “neurofibrosarcoma” or “neurosarcoma”, is derived from Schwann cells or pluripotent cells of the neural crest. In approximately 15% of MPNSTs we can observe epithelioid or other heterogeneous components. Clinically, these tumors are aggressive, locally invasive, and highly metastatic [1, 2].

The incidence in the general population is 0.001% while the risk is approximately 4600 times higher in patients with type I neurofibromatosis. 3-13% of patients with type I neurofibromatosis will develop MPNST usually after latent periods of 10-20 years [1-3].

In our paper we would like to present a case of a 74-year-old patient without type I neurofibromatosis with

MPNST which was first misdiagnosed as a non-small cell lung cancer.

Case report

A 74-year-old woman was admitted to the Clinic of Thoracic Surgery, General and Oncological Surgery Military Academy Hospital in Łódź in 2012 due to left lung tumor which was diagnosed in the pulmonology clinic of another hospital while she was treated for lower respiratory tract infection of one month duration. Chest radiographs demonstrated a shadow involving the lower half of the left chest (Fig. 1). Further investigation with computed tomography enhanced with contrast injection showed a large mass 18 × 18 × 11 cm which was described as lung tumor of the inferior left lobe. During this examination the CT guided fine needle biopsy was performed. The result was described as a non-small cell lung cancer.

After this examination the patient was qualified for scheduled surgery. Left posterolateral thoracotomy was performed. Surprisingly, the tumor which was previously

Address for correspondence: Piotr Misiak, Military Medical Academy University Teaching Hospital – Central Veterans' Hospital in Lodz, 113 Żeromskiego St., 90-549 Łódź, tel. +48 510 115 108, e-mail: pitmis@interia.pl



Fig. 1. Chest X-ray presenting a lesion in the left pleural cavity

described as a lung tumor was a large unencapsulated tumor protruding from the chest wall which was infiltrating the diaphragm. The entire lower lobe was oppressed but not infiltrated. On the basis of the intraoperative findings the patient underwent wide local excision, including the adjacent muscles and pleura, but it was not possible to perform oncological resection with the appropriate margins especially from the diaphragm.

The resected specimen consisted of a dome-like exophytic tumor of 18 cm diameter. A cut section showed white, yellow, fleshy, lobular tumor tissue with extensive areas of hemorrhage and necrosis.

Microscopically the neoplasm was composed of spindle cells arranged in interlacing fascicles, whorls and a palisading pattern. Spindle cells showed hyperchromatic nuclei with increased mitotic activity. Immunohistochemistry



Fig. 2. Tumor of the right pleural cavity

stains: S-100 positive in spindle cell areas, Desmin-negative, CK7-negative, EMA-negative. The final diagnosis was established as G3 high grade malignant peripheral nerve sheath tumor. Multiple biopsies from the infiltrated part of the diaphragm also showed malignant cells. The postoperative course was uneventful and the patient was referred to the oncologist for adjuvant chemotherapy (according the ADR schedule). Despite the chemical treatment after four months she was once again referred to our clinic for consultation. The control CT scan showed multiple metastases to both lungs. Moreover, in the right pleural cavity the radiologist described a pathological mass (20 × 12 × 16 cm) (Fig. 2). The mass compressed the mediastinal structure to the left and the liver downwards. The patient was qualified for a palliative operation to alleviate respiratory and circulatory disorders caused by compression of the lung, mediastinal structures and vena cava. During the surgical procedure we removed the tumor. We did not interfere with the lung tissue because the tumor was only oppressing it, not infiltrating. We used argon coagulation to diminish the visible place of infiltration at the chest wall and diaphragm. The second postoperative course was also free of major complications. After the operation indicators of respiratory function improved. It was also accompanied by disappearance of shortness of breath. The patient was referred for palliative radiotherapy. Despite this combined treatment she died 8 months after the second surgery due to multiple lung metastases.

Discussion

Malignant peripheral nerve sheath tumors are rare, aggressive and highly metastatic soft tissue sarcomas of ectomesenchymal origin. These tumors are derived from Schwann or pluripotent cells of neural crest origin and arise from peripheral nerve branches or sheaths of peripheral nerve fibers. They can arise de novo or from preexisting benign neurofibromas. MPNSTs account for approximately 5-10% of all soft tissue sarcomas [1-4].

The most common anatomical sites include proximal portions of upper and lower extremities, the trunk (arising from the brachial plexuses, sacral plexuses or the sciatic nerve). In our case the tumor originated from the sheet of the intercostal nerve, which makes this case more uncommon.

The diagnosis of MPNST is very difficult. Needle biopsy of the suspected tumor is the most popular diagnostic tool but according to various authors the result of this examination can be easily misdiagnosed, as in our case. The radiologic findings are various. CT examination is the method of choice; it provides fine features of the relationship between the tumor and surrounding structures, but there are no specific CT findings which may clearly establish the final diagnosis. Also MRI has limited utility in distinguishing MPNSTs from benign peripheral nerve lesions [4, 5].

The histogenesis of this kind of tumor is still unclear, but there is higher incidence in patients with neurofibromatosis type I. The majority of MPNSTs are unencapsulated infil-

trating tumors composed of spindle cells arranged in a fascicular growth pattern. S-100 protein is the most commonly used antibody to identify various nerve sheath tumors, but only 50-70% of MPNSTs exhibit S-100 staining. The reactivity is grade related. In lower-grade tumors the staining is more extensive while in high-grade tumors it can be found in single cells [1, 2, 4].

The treatment of MPNSTs is difficult. Aggressive surgical resection and complete removal of the tumor with histologically clear margins is the first line of therapy, and most important, because the effect of postoperative therapy is not clear. The role of chemotherapy has never been adequately studied, but the effect of radiation therapy in local control of MPNSTs has been proved by various studies [2-5].

Biologically, MPNSTs are mostly high-grade sarcomas with very high potential to recur and metastasize. The 5-year local recurrence and distant metastasis rates have been reported as approximately 40% and 65% respectively. The long-term prognosis remains poor with 5-year survival ranging from 16 to 53%. Hematogenous metastatic spread occurs most commonly to the lungs; metastasis via the lymphatic route is very rare [4, 5].

A number of different prognostic factors have been cited in the literature: the primary location of the tumor (better prognosis when the tumor originates at extremities), the tumor size (< 5 cm), the tumor grade and the histological type, prior irradiation, neurofibromatosis type I, and oncological margin status [2].

Most reports agree that a negative surgical margin is the most significant prognostic factor for survival and local control of the disease. Therefore it is so important to perform complete surgical resection of the tumor [1-5].

Conclusions

Malignant peripheral nerve sheath tumor originating from the intercostal nerve is a very rare clinically entity, especially when the patient has no coexisting type I neurofibromatosis.

The incidence in the general population is 0.001%. However, we should consider this type of tumor in the differentiation of other cases, localized in the chest cavity, because fast radical surgery can lengthen a patient's life.

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