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CASE REPORT

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Synovial sarcoma of the neck

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Oncology in Clinical Practice

2015, Vol. 11, No. 6, 317–321

Translation: GROY Translations

ISSN 2450–1654

www.opk.viamedica.pl

ABSTRACT

Soft-tissue sarcomas originate from the mesodermal tissue. They mainly locate within extremities (50% of cases), trunk (30%), head and neck (10%), and in the retroperitoneal space (10%). Sarcomas represent about 1% of all malignant tumours in adults. So far, causes of the soft-tissue sarcomas have not been well recognised. These neoplasms occur more frequently in persons with genetic diseases. Frequent prevalence of sarcomas is observed in subjects with Von Recklinghausen disease (neurofibromatosis), Gardner syndrome, Werner syndrome, tuberous sclerosis, and Li-Fraumeni syndrome (mutation p53). This paper presents the case of a 29-year-old patient with a large tumour of the neck and Von Recklinghausen disease, referred to the Department of Thoracic Surgery with the diagnosis of MPNST (malignant peripheral nerve sheath tumours).

Key words: soft tissue sarcoma, synovial sarcoma

Oncol Clin Pract 2015; 11, 6: 317–321

Introduction

Soft-tissue tumours are generally classified based on the type of tissue they originate from. They can arise from adipose tissue, connective tissue, muscles, as well as nervous and vascular tissue. Counterparts of some soft-tissue tumours in healthy mesenchymal tissues remain unknown. Neoplasms of soft tissues can occur at any age. The majority of benign lesions are diagnosed accidentaly or due to local symptoms of tumour. Soft-tissue sarcomas occur less frequently and are characterised by aggressive local growth and the presence of multiple distant metastases. Intermediate group consists of neoplasms with aggressive local growth affecting patients' condition, that usually without distant metastases [1, 2]. Malignant soft-tissue tumours occur rarely, and they account for less than 1% of all neoplastic proliferations in adults. However, these tumours are in the fourth place concerning prevalence among childhood malignant neoplasms, following malignancies of the haematopoietic system, tumours of nervous origin, and Wilms' tumours [3, 4].

Malignant peripheral nerve sheath tumours (MPNST) comprise 5–15% of soft-tissue sarcomas and the most frequently affect patients between 30 and 50 years of age [5]. In over 30% of cases, the MPNST develop on the base of previously existing neurofibroma in patients with neurofibromatosis type I (Von Recklinghausen disease I). Macroscopically, the MPNST is a solid tumour, moderately differentiated, and with necrotic foci and haemorrhages. The typical lesion is built of cells similar to fibroblasts, with elongated nuclei, and frequently mitoses occur. Immunohistochemically, MPNST shows the presence of markers of proliferation in the direction of nerve sheaths such as the protein S-100, Leu 7, PGP9 [6]. In disseminated disease, palliative chemotherapy (doxorubicin and ifosfamide) is used with clinical benefit observed in about 25–30% of patients.

Synovial sarcoma is a rare tumour; it comprises 5–10% of all sarcomas [7]. Most frequently, patients in the age range 25–35 years suffer from this disease. This neoplasm can arise in any location. Despite its name, connection to synovial tissues has not been proven [8]. Currently, it is assumed that it originates from the mesenchymal cell and develops in the joint area, although sometimes it grows in tissues remote from joints [9]. Histologically, several subtypes of this tumour are distinguished. The most frequent monophasic subtype is built of spindle cells creating bundles resembling a shoal of sardines [10]. Due to its monotonous bundle structure, the monophasic synovial sarcoma is often mistaken for other spindle-cell tumours, e.g. fibrosarcoma or MP-

NST. The biphasic subtype, besides the spindle cells, also contains epidermal cells creating glands, papillary structures, and solid fields. In this subtype the spindle-cell or epidermal component can predominate [10]. The third subtype of the malignant synovial sarcoma is the poorly differentiated subtype, microscopically characterised by the presence of fields comprising small round cells with numerous mitotic figures and necrosis [11]. A very rarely found subtype of synovial sarcoma is the calcifying subtype. This subtype is distinguished by the presence of multiple calcifications and better prognosis [8].

Prognosis in patients with soft tissue tumours depends on many factors. Distinction between the benign and malignant lisions is simple in many cases; however, in some cases it can turn out to be extremely difficult. A significant influence on the prognosis is exerted by: histological type of the tumour, its maturity expressed as the degree of differentiation (grading), and the size and range of invasion (stage). Some sarcomas, e.g. rhabdomyosarcoma in children, react well to combined treatment, while others, such as malignant synovial sarcomas, seem not to respond to any treatment [12]. In a substantial proportion of soft-tissue tumours, permanent chromosomal disorders are possible to diagnose using standard cytogenetic and molecular techniques. Some tumours can be identified based on translocations occurring in them that are sometimes prognostic factors.

Case presentation

A 29-year-old man with the diagnosis of MPNST was admitted to the Department of Thoracic Surgery in order to secure airway patency and to consider the possibility of palliative tumour resection. The histopathological diagnosis was made based on open biopsy of the tumour performed in the referring institution. Before being admitted to the Department of Thoracic Surgery the patient underwent palliative radiotherapy of the neck tumour area. In 2012, the patient was diagnosed with multifocal Von Recklinghausen disease. At admission, the patient complaint of cough and shortness of breath during light effort (Fig. 1, 2). The physical examination showed an oval hard lump, 15 cm in diameter, movable against the base, in the neck area, slightly on the right side. Moreover, on the skin, on the right side in the area of loins, numerous lumps (neurofibromas) and a hairy birthmark were found. Bronchoscopy proved translocation of the larynx towards the left side, and 2 cm below the vocal folds right-side compression of trachea causing stenosis of its diameter by 50%. No other changes were found in the bronchi. In the area of the stenosis, a coated, self-expandable stent of 16 × 60 mm was placed. Correct position of the stent was confirmed radiologically. The patient was discharged in good general condition.

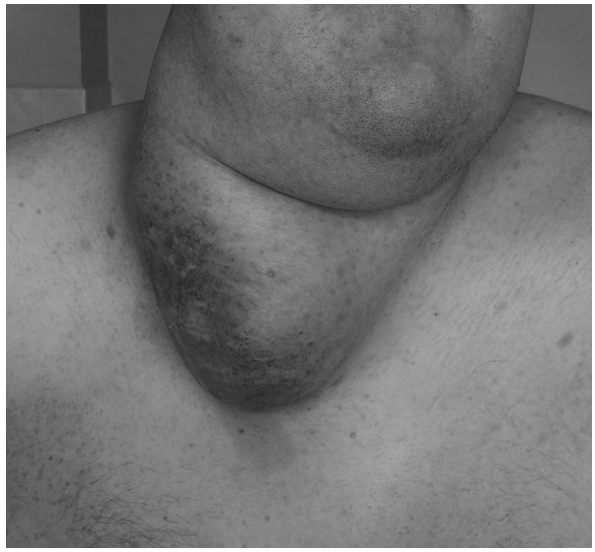

Figure 1. Neck tumour in 29-year-old patient

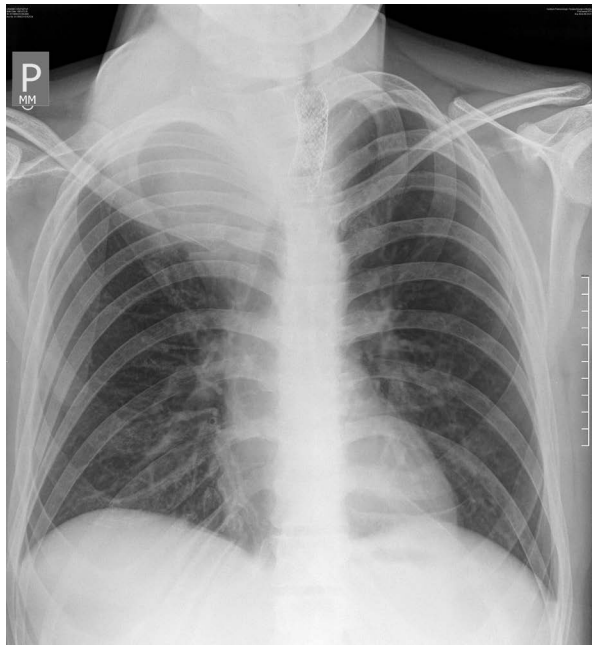

Figure 2. Control X-ray of the ribcage in the PA view — after placing a self-expandable stent in the trachea

In the scope of the follow-up treatment and oncological supervision, the patient received palliative chemotherapy (ifosphamide), and five months later he was again admitted to the Department of Thoracic Surgery in order to remove the tracheal stent, which had moved downwards, thus ceasing to fulfill its previous role. In a neck and mediastinum CT scan (using spiral technique, collimation 40 × 0.6 mm, with the Somatom Definition Edge apparatus, before and after the contrast), which was a comparative examination in

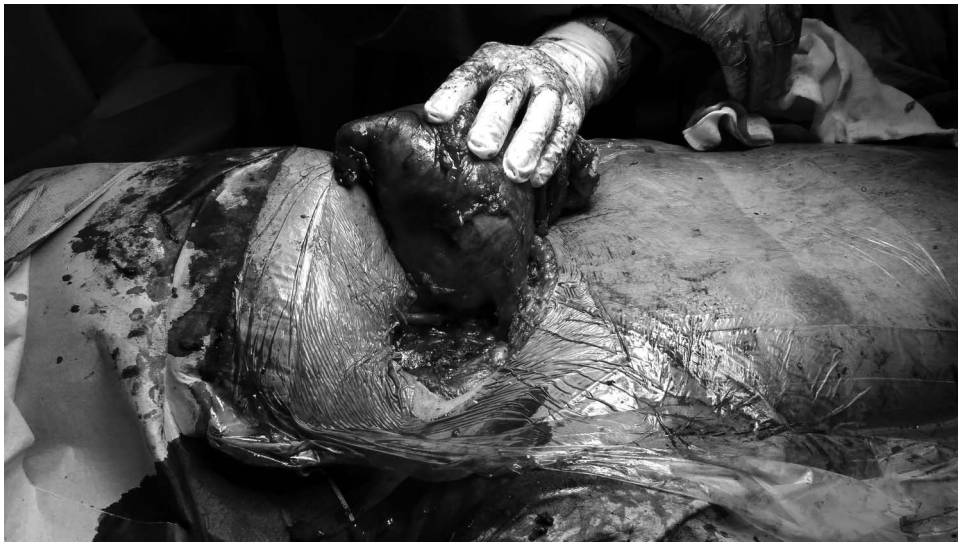

Figure 3. Tumour before en bloc resection

relation to the CT scan performed eight months earlier, progression of the tumour size was found. The current largest transverse dimension of the tumour was 13 cm, while in previous examination it was about 7 cm. The tumour caused translocation and pressure to the trachea on the level of the upper opening of the rib cage. The width of the tracheal opening in the coronal plane was 10 mm (previously 16 mm). Just below, in the rib cage area, the internal tracheal stent was visible, reaching the place of the trachea division. It can be supposed that lowering of the stent had a direct connection with the growth of the tumour. In the left above-clavicle area, a mildly enhancing 76 × 36-mm tumour was found. In the immediate area of the right common carotid artery, several smaller satellite lisions measuring up to 2 cm in diameter were present. The tumour within the upper mediastinum caused translocation to the left side of the lower and central pharynx. Conclusion of this examination was progression of the size of lisions by about 35% in comparison to previous examinations. The tracheal stent was removed. After the procedure, the patient did not complain of shortness of breath. The patient was discharged in stable general condition.

After three weeks, the patient was again admitted to the Department for qualification for surgical palliative treatment. An magnetic resonanse imaging (MRI) was performed to assess the topography of vessels (MRI of the neck in coronal and sagittal planes, in sequences T2/FR/FSE, T1/SE, T2/FS, after intravenous administration of the contrast medium, sequences T1/FSE and T1/FS were conducted, assessment of vessels after intravenous administration of the contrast medium — TRIX method). On MRI the presence of a large pathological mass was found — the lision had polycyclic outlines, regular edges, and lack of infiltration to neighbouring structures. On the front medial surface of the tumour, external and internal common carotid arteries of regular diameter and walls without indications of infiltration were visible. The internal vein, passing through the back surface of the tumour was severely compressed due to the pressure. There was no evidence of infiltration. The tumour was localised over the sternoclavicular joint, and it does not cause infiltration to the bone structures. From the top, the tumour penetrated the submandibular space, without infiltration to the submandibular salivary gland. The neck lymph nodes were not visible. After intravenous administration of the contrast medium, the lision underwent heterogeneous contrast enhancement. The next focus was localised in the left above-clavicle area without infiltration to the neighbouring bone structures, subclavian vessels, and muscles. In segment I/II of the right lung, soft-tissue well-distinguished infiltration was found, also undergoing contrast strengthening. After taking into account the patient's tests results, the patient was qualified to palliative surgery (Fig. 3). Transverse skin incision was performed, cutting through the hypodermis and platysma muscle to the layer of surface neck fascia, on the length allowing the tumour to be exposed. The skin incision was then deepened; in this way the tumour was exposed. Sharply and bluntly alternatively dissecting along the right common carotid artery, the tumour was dissected from the surrounding tissues. After dissection, vascular loops were placed around the internal and external carotid artery, which would enable stopping and directly controlling potential haemorrhage. The main vessels were divided, tightened, and secured. The tumour was dissected in monoblock. Haemostasis control was performed. Drainage of the operative field was provided. The surgical wound was closed in layers. On postoperative day 1, massive

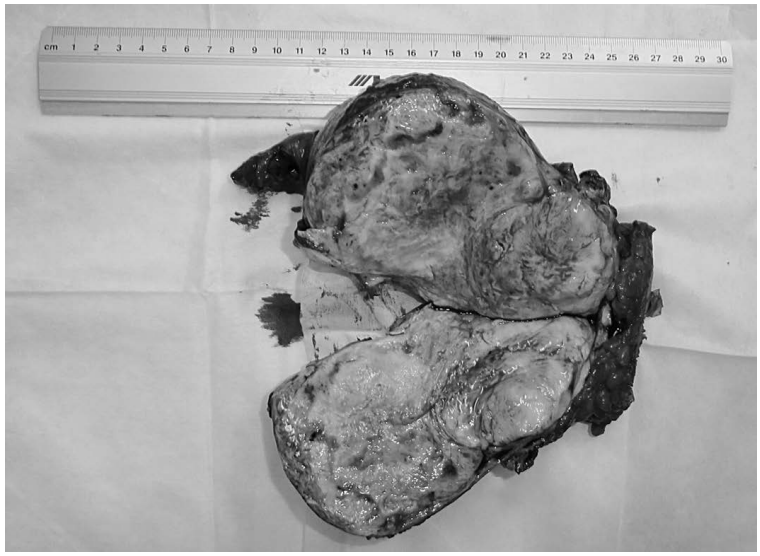

Figure 4. Microscopic view of the tumour

haemorrhage from the right common carotid artery that delaminated occurred. The patient underwent emergency surgery — during the revision the bleeding vessel was secured. The approximate blood loss was 1500 ml — two units of PRBCs of compatible blood group were transfused with no complications. After the surgery, the patient was conscious without symptoms of CNS damage. On postoperative day 2, a third unit of PRBCs was transfused. The patient was discharged on postoperative day 9 in good general condition. The dissected tumour was passed to histopathological examinations (Fig. 4). Examination description: a tumour of dimensions of 14 × 12 × 7 cm, quite well separated with the pseudocapsule, does not infiltrate the skin. In cross-section yellowish, with areas of necrosis. On the tumour's periphery, nodular structures resembling lymph nodes are visible. The immunohistochemical examination showed spindle-cell epidermal tumour tissue of the phenotype CD99(+), bcl-2(–), CD68(–), CK AE1/AE3(–), S-100(±), CD34(+), Actin (–), Desmin (–). High proliferation activity of the tumour — the Ki-67 reaction was positive in about 40% of nuclei. The entire histopathological picture matches the diagnosis of monophasic synovial sarcoma — malignant synovial sarcoma.

Summary

There is a small group of neoplasms of unknown histogenesis. This group includes the alveolar soft part sarcoma, epidermal sarcoma, and malignant synovial sarcoma. The malignant synovial sarcoma discussed in the above case report is the most frequent. It comprises 10% of all soft-tissue sarcomas. In the majority of synovial sarcomas the characteristic (and present only in this type of tumour) chromosomal translocation t(X;18) (p11.2;q11.2) is present. This translocation is associated with the recently identified irregularities within the so-called malignant synovial sarcoma-accompanying genes. It seems that these genes influence the morphology and biology of these malignancies [13]. The malignant synovial sarcomas are neoplasms of different sizes — from small ones, seemingly separated lisions, to large infiltrating masses. The rarely occurring monophasic synovial sarcoma is the most malignant type. Determination of keratin or the epidermal membrane antigen (EMA) is helpful in making the diagnosis. The best factor confirming the diagnosis is proving the chromosomal translocation t(X;18) [13].

The backbone of treatment in soft-tissue sarcomas localised in different anatomical areas is surgical dissection. Initial evaluation of the possibility of performing full resection, taking into account the patient's safety — evaluation of local advancement, infiltration to adjacent structures, is the most appropriate approach. The en bloc resection of the tumour, with the widest possible margins, is preferred. However, the extent of the surgery needs to be adjusted to avoid any damage to important structures, while trying to include appropriate margins [14]. After the surgery, it is the decision of the qualifying oncologists whether to continue the treatment with chemotherapy or radiotherapy. The standard procedure is to perform adjuvant radiotherapy, despite the relatively low radiosensitivity. Moreover, currently, combined treatment regimes are analysed with preoperative radiotherapy, which concerns non-small cell soft tissue sarcomas, including synovial sarcomas.

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