

CASE STUDY

DIFFERENTIAL DIAGNOSIS OF SOFT TISSUE TUMORS

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Denys Kapustianskyi, Ihor Ivanytskyi, Tetiana Ivanytska, Valeriy Zhamardiy, Viktoriia Donchenko
POLTAVA STATE MEDICAL UNIVERSITY, POLTAVA, UKRAINE

ABSTRACT

The aim of the study is to identify clinical signs of neurilemmoma of the upper limb, its main ultrasound and X-ray distinctions from other benign tumors of soft tissues.
To make a preliminary diagnosis, it is advisable to use different methods of imaging, such as: ultrasound, MRI, radiography. Radiological diagnosis of soft tissue tumors should be based on the integrated use of radiological and ultrasound methods with mandatory consideration of clinical and anamnestic data such as patient age, topographic location of the tumor and its growth rate. Although ultrasound and MRI do not allow to completely differentiate the tumor, but they help to choose the right tactics of the patient. Treatment is to remove the tumor and includes cesium dissection and removal from the nerve bundle, using magnification.

KEY WORDS: neurilemmoma, schwannoma, tumorof the peripheral nerve, tumor of soft tissues, ultrasonography

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INTRODUCTION

Soft tissue tumors are a very common pathological condition. Doctors of various specialties such as surgeons, dermatologists and general practitioners have to diagnose these diseases. The most common among them are fibromas and lipomas, which usually do not cause discomfort, have different localization and slow growth, they rarely acquire characteristics of malignant tumors [1]. Because these tumors are most common, patients are often diagnosed with lipomas or fibromas when soft tissue formations are detected, even without additional examinations, which leads to misdiagnosis and, consequently, incorrect treatment.

Neurilemmomas (schwannomas) account for 5 % of benign soft tissue tumors [2]. Neurilemmoma, also called schwannoma, develops from cells of the Schwann sheath of peripheral nerves. Schwann cells (neurolemocytes) are involved in the formation of nerve fibers, forming their shells, in the peripheral nervous system [3]. In embryogenesis, Schwann cells are of neuroectodermal origin. As peripheral nerves are formed, Schwann cells migrate from the spinal ganglia parallel to the axons and close them within their cytoplasm.

Neurilemmoma is formed in the soft tissues along the nerve trunks and cranial nerves, occasionally it can be formed in the internal organs. Neurilemmoma is the most common benign tumor of the peripheral nerves sheaths. Mainly located on the small and medium-sized nerves of the head, neck, flexor surface of the extremities, sometimes affecting the skin. The presence of a soft tissue tumor near the peripheral nerve gives the doctor grounds to think about the presence of neurilemmoma.

Clinically, schwannoma is a painless encapsulated formation (single or multiple), which is observed mainly under the age of 50, affects men and women with equal frequency. In most cases, neurilemmoma is a solitary tumor, multiple schwannomas are observed in Recklinghausen's disease or schwannomatosis [2, 4]. Elbows and tibial nerves are often affected by neurilemmomas.

Neurilemmomas are the most common benign tumors of peripheral nerves, but to distinguish them from neurofibromas and neurocarcinomas in the preoperative period is quite difficult. Often the final diagnosis is established during surgery or in the postoperative period. To make a preliminary diagnosis, it is advisable to use different methods of imaging, such as: ultrasound, MRI, radiography. Radiological diagnosis of soft tissue tumors should be based on the integrated use of radiological and ultrasound methods with mandatory consideration of clinical and anamnestic data such as patient age, topographic location of the tumor and its growth rate. Although ultrasound and MRI do not allow to completely differentiate the tumor, but they help to choose the right tactics of the patient. Treatment is to remove the tumor and includes cesium dissection and removal from the nerve bundle, using magnification.

THE AIM

The aim of the study is to identify clinical signs of neurilemmoma of the upper limb, its main ultrasound and X-ray distinctions from other benign tumors of soft tissues.

CLINICAL CASE

Given the relatively high percentage of diagnosed neurilemmoma in the postoperative period and the need

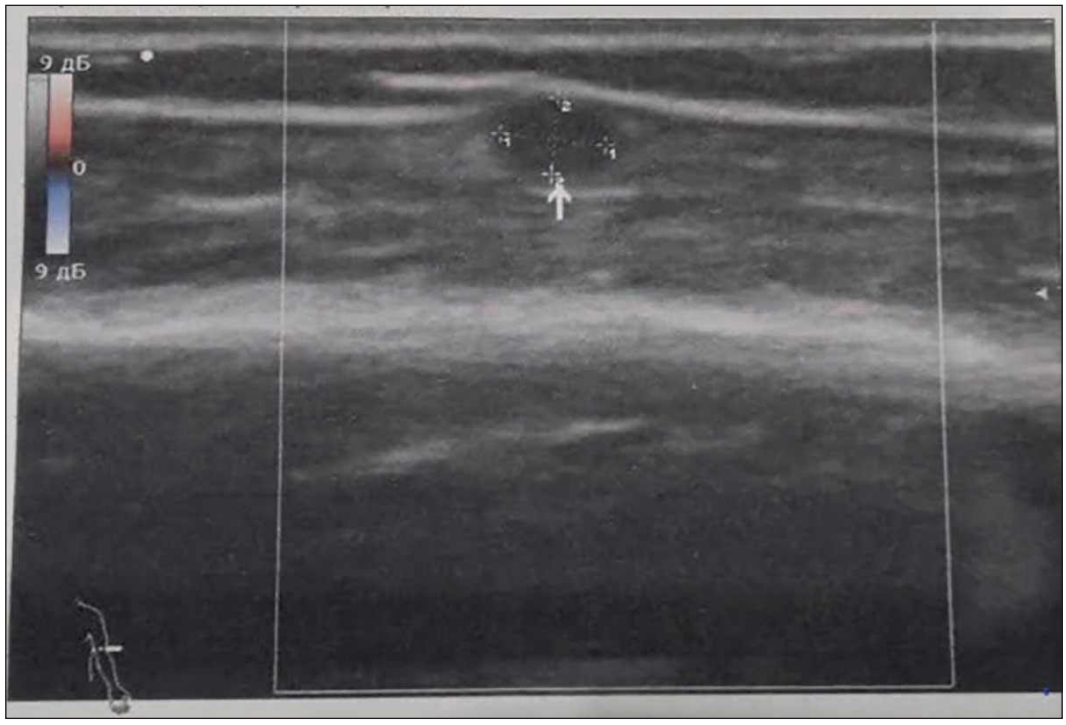

Fig. 1. Ultrasound of shoulder neurilemmoma, showing a hypoechoic encapsulated formation

for great caution in its surgical treatment to preserve the integrity of the nerve, we consider it necessary to describe the clinical case from our own experience.

Patient A, 22 years old, complained to the family doctor about the appearance of a small painful formation in the elbow area on the right. Application of pressure on the formation lead to significant increase in pain which irradiated in the shoulder and forearm, limiting the atient's movements. Patient first noticed a slight pain six months ago after strenuous exercise in the gym (squeezing off the floor). Palpation revealed a small, up to 5 mm, painful formation, the occurrence of which is associated with exercise. On examination: on the extensor surface of the shoulder 2 cm above the elbow joint a formation with a diameter of about 7 mm was palpated, which was sharply painful after slight pressure and percussion, the pain irradiated to the shoulder and fingertips. The patient was referred for soft tissue ultrasonography. Ultrasound examination revealed in subcutaneous adipose tissue a hypoechoic formation of a round shape with clear, smooth contours, homogeneous structure, hyperechoic capsule, not associated with the surrounding soft tissues. The formation was visualized near the triceps tendon. It shifted together with the tendon during active and passive movements. Between the tendon and the formation revealed a thin (diameter less than 1 mm) hyperechoic structure associated with the formation. It was seen by us as *nervus cutaneus brachii posterior* (Fig. 1). When performing compression elastometry, streinratio was 4.8 when compared to adipose tissue and 3.9 when compared to muscle tissue, indicating increased stiffness of tumor tissue. Performing ergot Doppler mapping revealed the absence of blood vessels in the tumor. We concluded that there is a high probability of tumor origin from the *nervus cutaneus brachii posterior* and the need for its surgical removal.

After clarifying the topographic and anatomical features of the location of the tumor it was removed. Under local infiltrative anesthesia with 2 % lidocaine solution, the skin was dissected over the tumor, then with the help of a surgical magnifying glass under visual control the tumor was isolated and removed. We needed to remove the tumor as accurately as possible with minimal nerve damage, to prevent further neurological dysfunction. The removed tumor was placed in a preservative and sent for histological examination. According to the results of histological examination, the diagnosis was «Neurilemmoma». In the early postoperative period, the patient noted the absence of clinical symptoms, the condition improved significantly, the pain disappeared, there was no violation of neurological functions. The postoperative wound healed with primary tension. Sensitivity in the area of innervation of the nervus cutaneus brachii posterior was not disturbed. At next examination of the patient after 1 month any features or pathology in the course of the postoperative period were not revealed.

DISCUSSION

Schwannomas or neurilemmomas are rare tumors. They are often single and benign formations, but can be multiple and associated with neurofibromatosis type 1 and schwannomatosis [5]. Neurilemmomas are most common in the head, neck, including the brachial plexus and spinal nerves. Limbs are affected less often. In this case, tumors are mainly

found on the flexor surfaces of the limbs. Schwannomas of the upper extremities are quite common, manifested by mild symptoms and lack of neurological manifestations [6]. It is important to make a differential diagnosis of neurilemmoma from the most common soft tissue tumors.

The most common benign soft tissue tumors are lipomas formed by matured adipose tissue. They can be single or multiple, including systemic lipomatosis. They are distributed in the population regardless of gender and age. Clinically, these are painless soft-elastic formations. The skin over them is not changed. A common feature of lipoma on radiographs and echograms is the density of the shadow or echogenicity, which is the same as subcutaneous fat. Liposarcomas have increased echogenicity [7, 8].

Benign formations of fibrous origin – desmoids (invasive fibromas). These are peculiar tumors, which are characterized by local infiltrative growth and constant recurrences. A characteristic clinical feature of fibromas is a very dense consistency with limited mobility. Radiologically they are characterized by the density similar to the muscle tissue, elongated shape and infiltration of surrounding tissues, including subcutaneous fat. In ultrasonography, they are homogeneous and hypoechoic.

The main clinical sign of neurilemmoma is pain on pressure with irradiation along the corresponding nerve. The radiograph reveals a small formation with a clear contour of the ovoid shape in the subcutaneous fat or in the muscles in the projection of the main nerve. At ultrasonic research of a schwannoma is revealed as homogeneous hypoechoic formation.

Diagnosis of neurilemmoma at the preoperative stage is quite a difficult task, because the tumor grows slowly and in most cases has few clinical manifestations. Proper diagnosis is essential for proper planning of surgery, which aims not only to remove the tumor, but also to preserve the integrity of the affected nerve. Neurilemmomas have many features in common with soft tissue tumors, which are the cause of misdiagnosis and, accordingly, incorrect treatment tactics. Differential diagnosis of neurilemmoma should also be performed with neurofibromas, ganglion cysts, lipomas, xanthomas and malignant tumors [6]. For example, neurofibroma is indistinguishable from schwannoma during objective examination. The final diagnosis can be established only histologically. There are no specific symptoms to distinguish these tumors. The slow growth of benign tumors of the nervous tissue allows it to adapt to being under pressure without significant dysfunction. The onset of the first symptoms is more related to the location than the size of the tumor, because the symptoms of neuronal compression occur with increasing tumor mass, and this process can take several years. Unlike schwannoma, neurofibroma often becomes malignant, turning into neurosarcoma [8]. Because this process is long and the body adapts to it, it may not cause sufficient vigilance of the primary care physician. Therefore, it is advisable to conduct a comprehensive diagnosis of any detected tumor to prevent adverse effects.

Clinically, neurilemmoma has the appearance of a rounded almost painless formation. For neurilemmoma Tinel's positive symptom is characteristic, there can also be paresthesias and local increase in sensitivity. Sometimes patients complain of spontaneous pain, weakness in the extremities. Often this formation is detected by chance in the absence of clinical symptoms. Neurilemmoma rarely leads to impaired motor activity. Impaired motor function is often caused by neurosarcomas [9]. The presence of the formation that causes pain when pressed, shifts in the transverse direction relative to the axis of the limb and is almost motionless with its longitudinal displacement, percussion of the formation causes paresthesias along the affected nerve, which is analogous to the symptom of Tinel – these are the most characteristic clinical signs of neurilemmoma at the initial examination [10]. Quite often the correct diagnosis is made already in the operative or postoperative period that can have undesirable consequences for the patient. Untimely diagnosis of neurilemmoma in clinical practice is associated with nonspecificity and sometimes absence of symptoms. Schwannomas have a soft consistency, are mobile, grow slowly, and are sometimes completely painless. They are often diagnosed as lipomas, fibroids or xanthomas, which is the reason for their improper management. Visualization of the formation with the help of magnetic resonance imaging, radiography or ultrasonography will allow you to make a correct diagnosis in time. Ultrasonography is the primary method of visualization of soft tissue tumors, as it is widespread, accessible and fast to perform. In ultrasonography neurilemmoma has the form of a clearly defined hypoechoic and homogeneous mass with the presence of acoustic amplification.

Magnetic resonance imaging is the best way to diagnose neurinoma. These tumors may vary in their manifestations but, as a rule, most of them are isointensive in T1 mode and hyperintensive in T2 mode. Variations may be due to the content of fat and melanin in some neurinomas. About 40 % of schwannomas contain a cystic component. All neuromas accumulate contrast [11]. Unfortunately, these patterns of signal intensity do not help to distinguish tumors of neurogenic origin, but make it possible to clarify the neurogenic origin of the tumor. Neurilemmomas have a low risk of recurrence or malignancy. Surgical treatment is the careful removal of the tumor using optical lenses for magnification [10, 11]. After accurate, correctly performed surgery, neurological defects are virtually absent.

CONCLUSIONS

Thus, we conclude that it is necessary to conduct a differential diagnosis of soft tissue tumors using existing imaging methods (radiography, ultrasound, magnetic resonance imaging), which will help prevent misdiagnosis and prevent possible resection of the affected nerve. Peripheral nerve tumors should be included in the list of possible causes of soft tissue tumors. Only the presence of close cooperation between a family doctor, ultrasound diagnostician or radiologist and surgeon will qualitatively increase the possibilities of early diagnosis and treatment of atypical soft tissue diseases. Ultrasound diagnosis allows to identify

the differential signs of soft tissue tumors, the association of a tumor with a peripheral nerve, and to choose the correct tactics of patient management. Treatment of schwannomas is precise dissection and its removal from the nerve bundle, using a magnifying glass.

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ORCID and contributionship:
Denys Kapustianskyi: 0000-0002-7633-7713 [B-D,F]
Ihor Ivanytskyi: 0000-0002-0583-2303 [B-D]
Tetiana Ivanytska: 0000-0002-2556-7658 [B-D]
Valeriy Zhamardiy: 0000-0002-3579-6112 [A,E,F]
Viktoriia Donchenko: 0000-0002-9665-7204 [A,E,F]

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CORRESPONDING AUTHOR
Viktoriia Donchenko
Poltava State Medical University
23 Shevchenko st., 36011 Poltava, Ukraine
tel: +380662674172
e-mail: vik.donchenko@gmail.com

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