

Alveolar rhabdomyosarcoma of the thenar eminence in a 7-year-old child. A case report.

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ABSTRACT

Rhabdomyosarcoma is a highly malignant soft tissue tumor that develops from muscle cells. It is the most common soft tissue sarcoma in children and adolescents and rarely occurs in the hand. Alveolar rhabdomyosarcoma is the commonest histological subtype seen and has the worst prognosis. We report a case of a 7-year-old child, with an alveolar rhabdomyosarcoma of the right thenar eminence, which was treated by wide surgical excision, followed by adjuvant chemotherapy and radiotherapy. Seven years after the operation, no recurrence or metastasis has been observed and the patient remains tumor-free.

KEY WORDS: tumor; soft tissue; rhabdomyosarcoma; hand; thenar eminence

Introduction

Rhabdomyosarcoma (RMS) is a highly malignant tumor that develops from muscle cells. It is the most common sarcoma in children and adolescents, where accounts for more than 50% of all soft tissue sarcomas [1]. RMS can occur in different body areas, most commonly in the head, neck, genitourinary system and retroperitoneum. About 15% of the reported cases involve the extremities and only 7% of them the upper extremity. Primer hand involvement is rare [2].

There are four histological subtypes: embryonal (in infants and young children), alveolar (in older children and adolescents), botryoid (in infants and young children, typically in the vagina) and pleomorphic (older patients 40-70 years old) [2]. It is presented as a rapidly growing, painless mass and a series of tests and procedures are necessary for its diagnosis, including x-rays, ultrasonography (US), computerized tomography (CT), magnetic resonance imaging (MRI) and tumor biopsy (TB) [3].

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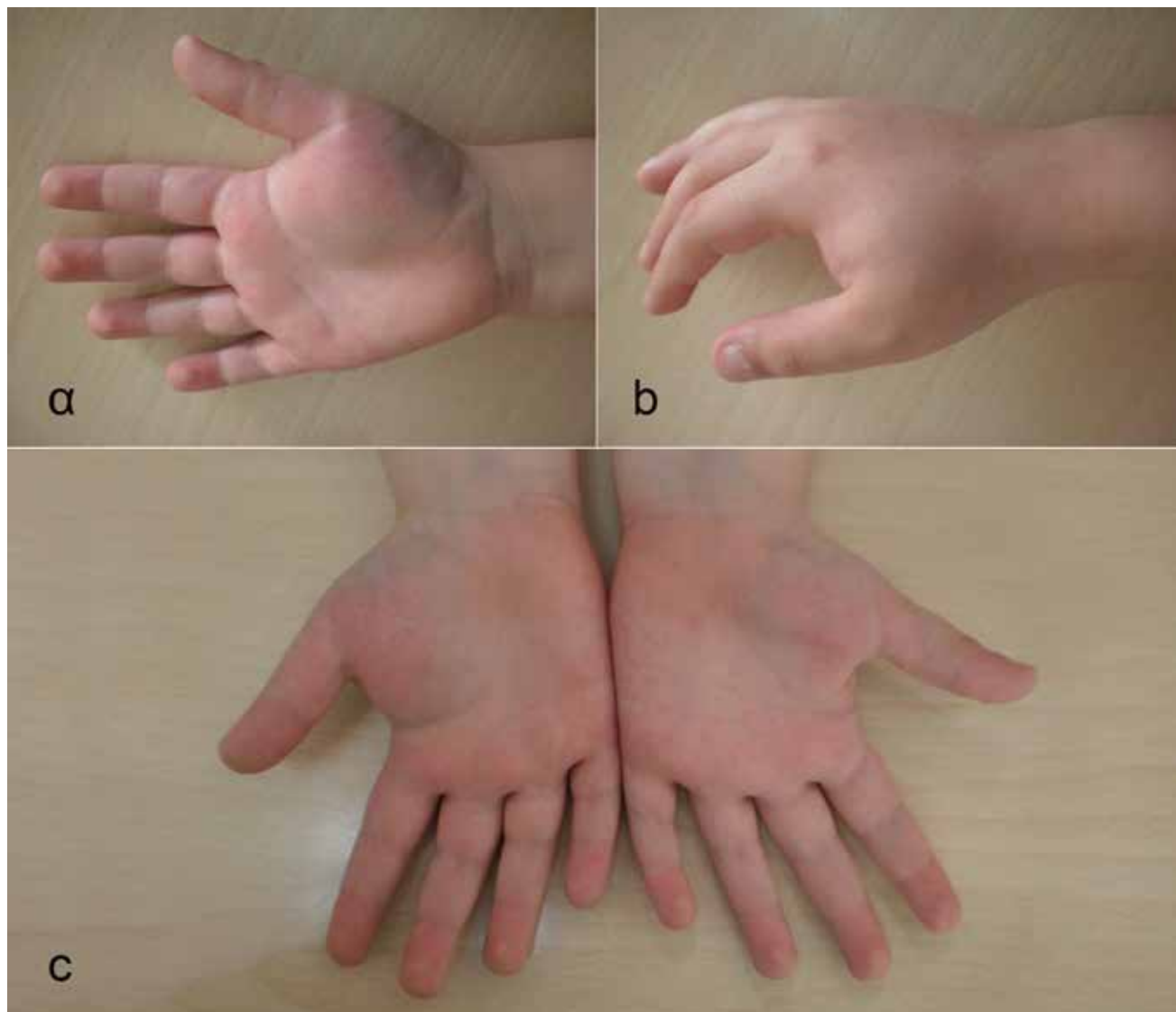


Fig.1: (a-c) Preoperative pictures of the right thenar eminence.

Alveolar RMS arising in the hand has been associated with poorer prognosis (50% five year survival) than other subtypes. In some cases, its small size and asymptomatic behavior can delay tumor detection and lead to the presence of metastases at the time of its initial diagnosis. Thus, early detection and application of the appropriate treatment of RMS are crucial for patient's survival [4].

We report a case of a 7-year-old child, with an alveolar RMS of the right thenar eminence, which

was treated by wide surgical excision, followed by adjuvant chemotherapy and radiotherapy. Seven years after the operation, no recurrence or metastasis has been observed and the patient remains tumor-free.

Case Report

On April 2011, a 7-year-old boy was presented to our department with an enlarging, painless, immobile, hard mass of his right thenar eminence, which was first noticed by his mother about one

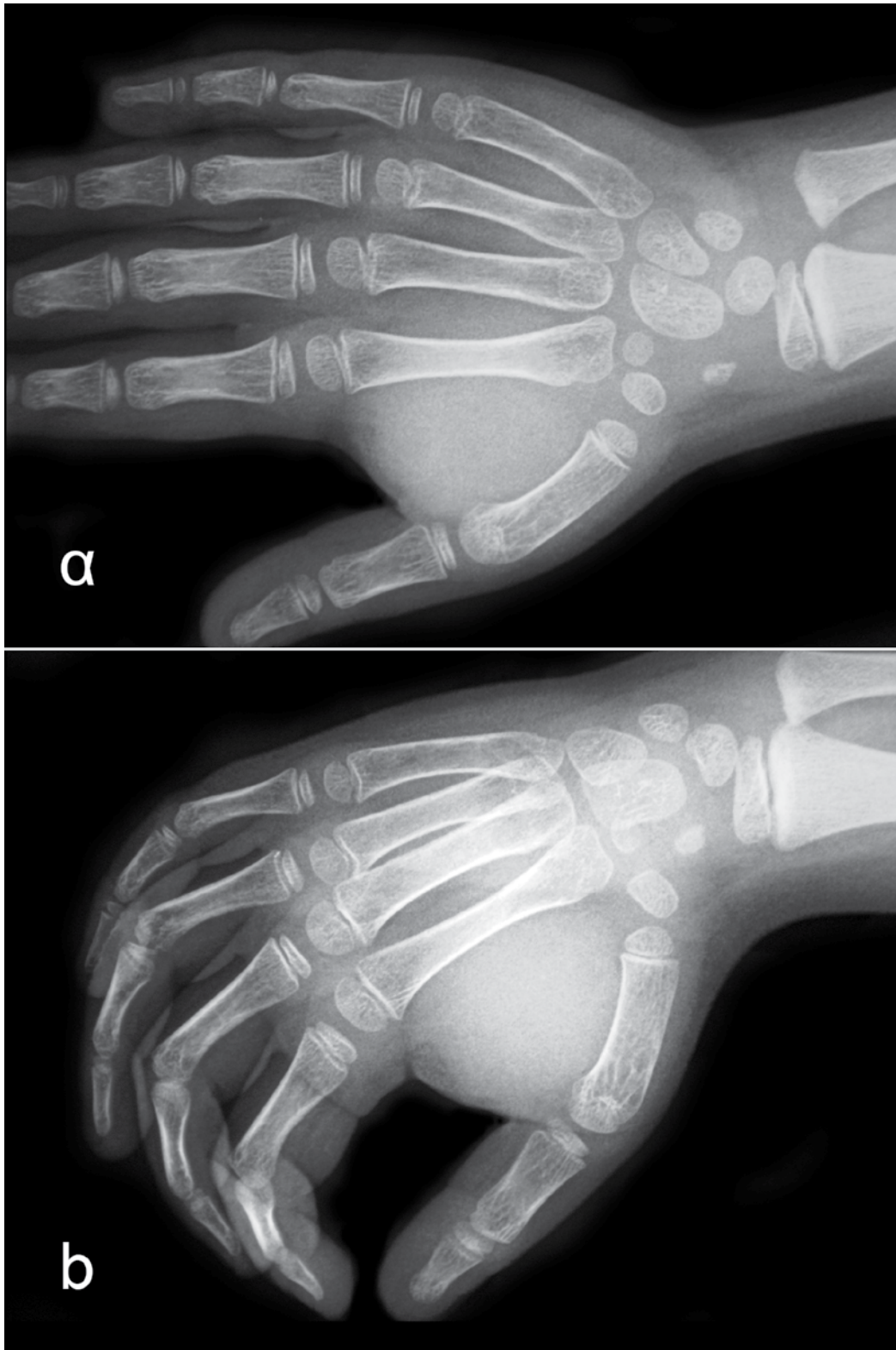


Fig. 2: (a) Anteroposterior and (b) lateral preoperative radiographs of the right hand show a soft tissue shadow in the region of the right thenar eminence.



Fig. 3: (a) T1 weighted coronal and (b) axial MRI; a solid hypointense mass lesion is observed. (c) Contrast enhanced coronal and (d) axial MRI show dense contrast enhancement.

month earlier. There was no history of trauma, fever, chills or weight loss. Clinical examination revealed a non-tender, fixed, hard, solid mass, with no irritation or changes of the color and trophism of the overlying skin (**Fig. 1a-c**). Local neurovascular elements and the range of motion of the thumb were not disturbed.

During imaging tests, on the anteroposterior and lateral radiographs, a soft tissue shadow was observed in the region of the right thenar eminence (**Fig. 2a-b**). The US examination showed a solid, lobulated mass, with intense vascular flow and no bone infiltration. The MRI scan revealed a heterogeneous, solid, lobular contoured mass,

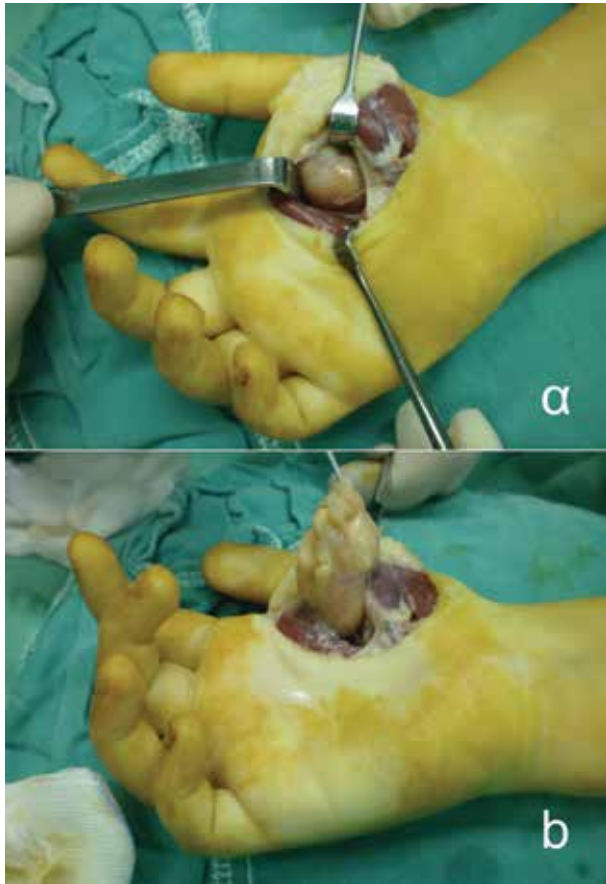


Fig. 4: (a-b) Intraoperative pictures of wide surgical tumor excision.

of about 4.5 x 2.0 x 1.5 cm in size, with low signal intensity on T1 images and high signal intensity on T2 images (**Fig. 3a-d**).

A wide surgical excision of the tumor was performed, followed by a histopathological examination (**Fig. 4a-b**). Histologically, the mass was composed of groups of poorly differentiated, small, round cells, which were separated and surrounded by dense fibrous septae. Multinucleated giant cells were also present within the tumor (**Fig. 5a**). Immunohistochemistry showed that the tumor cells were strongly positive for myoglobin, desmin, vimentin, MyOD1 and Myl-4 and focally positive for CD99, CD56, INI-1 and S-100p (**Fig. 5b**). A diagnosis of alveolar rhabdomyosarcoma

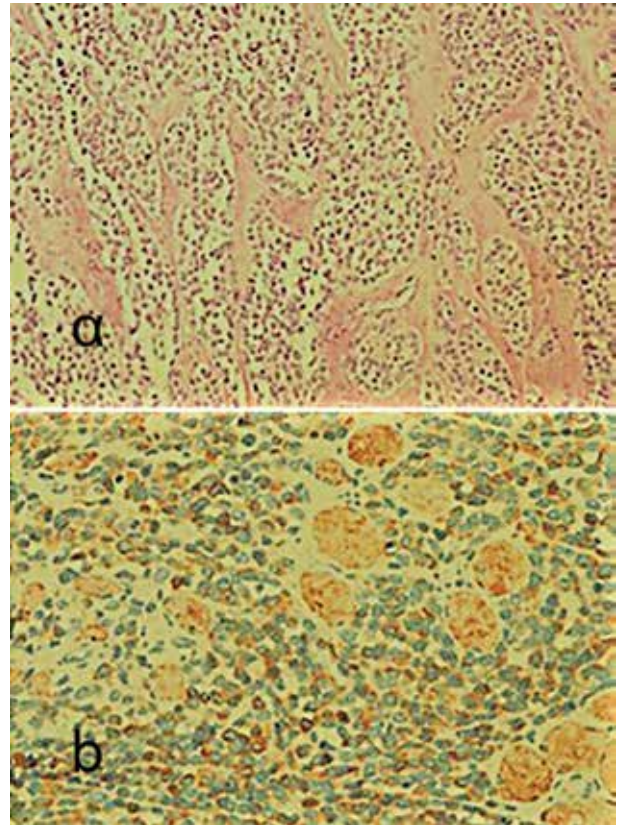


Fig. 5: (a) Diffusely infiltrating tumor cells loosely attached to fibrous septa peripherally with central loss of cohesion. **(b)** Diffuse desmin positivity within the tumor cells and the entrapped normal muscle fibers.

was confirmed.

Because of negative surgical margins, there was no need for reoperation and radical surgical clearance. A CT scan of the abdomen and chest, as well as a whole body bone scan, were also performed and there was no sign of metastasis. On consultation with an oncologist, it was decided the application of six cycles of external beam radiotherapy (RT) and four cycles of chemotherapy with vincristine and actinomycin.

Sixteen months postoperatively, a full clinical and imaging workup was done and no local recurrence or metastases were found (**Fig. 6a-b**). Seven years after the operation, the boy remains tumor-free, with a full range of motion of his right

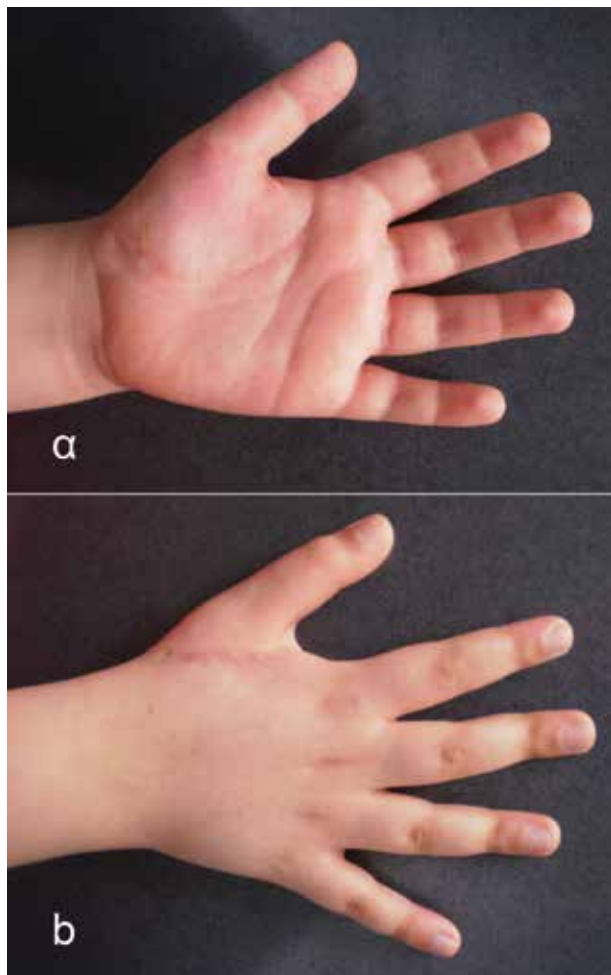


Fig. 6: (a-b) Pictures of the right hand and thenar eminence sixteen months postoperatively.

thumb and no other local disturbances.

Discussion

Rhabdomyosarcoma is the most common soft tissue tumor in children that accounts for more than 50% of all soft tissue sarcomas [1]. It is a highly malignant neoplasm which can invade the surrounding tissues, as well as to disseminate via lymph and blood flow and metastasize to the lymph nodes, lungs, bones, bone marrow, liver, breast etc [5]. According to the literature, about 15% of all RMS occur in the extremities and have poor prognosis irrespective of the kind of treatment applied. Involvement of the hand seems to be extremely rare and is reported only in a few

cases [2]. By the way, despite the fact that the majority of soft tissue sarcomas about the hand are painless, sometimes early detection of RMS primarily occurring in the hand can lead to a good prognosis [4]. Synchronous appearance of multifocal RMS has also been reported [6]. Many of the limb RMS in previous studies, had metastases at the time of initial presentation [5].


Histopathologically RMS is classified in four subtypes: embryonal, alveolar, botryoid, and pleomorphic. This classification is correlated with prognosis [2]. Alveolar RMS is more common in older children and adolescents and accounts for 32% of all rhabdomyosarcomas. It displays a more aggressive clinical course and has worse prognosis than the other subtypes [7]. Microscopically, RMS must be differentiated from other small round blue cell malignant tumors such as neuroblastoma, lymphoma, leukemia, Ewing's sarcoma and metastatic disease [8]. In the differential diagnosis of RMS must be also considered benign tumors like lipoma, neurofibroma and rhabdomyoma and other lesions like hematoma, pyogenic myositis, and myositis ossificans. The gold standard in the differential diagnosis of soft tissue tumors is always the histopathological examination [8].

Despite the fact that in some studies authors insist to the radical excision of RMS and partial or complete limb amputation, many other reports claim that patients who were treated primary with amputation had lower survival rates than those treated primary with wide local tumor excision. Therefore, wide local RMS resection is the primary treatment of choice in most of the cases and amputation must be applied only in case of positive surgical margins, or in wide tumor excision failure and recurrence of the tumor. Radical RMS excision may lead to significant functional and cosmetic impairment of the limb. Sometimes, reconstructive surgery can improve limb function at a later stage of treatment [9,10]. Surgical technique is very important. In case of biopsy, a longitudinal excision must be done and be the one which will be used during the final opera-

tion. Exsanguination of the limb with tourniquet is prohibited, because there is a chance of tumor dissemination [10].

Chemotherapy must be applied in all RMS cases, because it was proven that improves the overall survival. Response to chemotherapy can be predicted by the identification of gene fusions and chromosomal rearrangement. There is a significantly increased risk of failure and death in patients with metastatic disease if their tumors express PAX3-FKHR [10].

Most authors consider that there is no need of radiotherapy application, in case where radical surgical excision or amputation was done. RT must be used when local wide excision is done, or in cases of close/positive surgical margins and unresectable tumors, in order to enhance local tumor control [10].

In conclusion, RMS is a malignant soft tissue tumor which rarely occurs in the hand, and often presents with disseminated disease at its initial diagnosis. Alveolar RMS is the most common histological subtype seen, with the worst prognosis. In most patients wide tumor excision is the treatment of choice. Radical surgical excision significantly impairs limb function and cosmesis and must be kept in mind in case of wide excision failure or in recurrence of the tumor. Advances in chemotherapy and radiotherapy protocols in selected cases have improved the prognosis of the disease. 

Conflict of interest:

The authors declared no conflicts of interest.

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ΠΕΡΙΛΗΨΗ

Το ραβδομυοσάρκωμα αντιπροσωπεύει έναν υψηλής κακοήθειας όγκο μαλακών μορίων ο οποίος εξορμάται από τα μυϊκά κύτταρα. Πρόκειται για το συχνότερο σάρκωμα μαλακών μορίων σε παιδιά και εφήβους ενώ σπανίως εντοπίζεται στο χέρι. Το κυψελιδικό ραβδομυοσάρκωμα είναι ο συνηθέστερος ιστολογικός τύπος με τη χειρότερη πρόγνωση. Περιγράφεται η περίπτωση ενός επτάχρονου αγοριού με κυψελιδικό ραβδομυοσάρκωμα στο δεξιό θέναρ, το οποίο αντιμετωπίστηκε με ευρεία χειρουργική εκτομή, συνεπικουρούμενη από χημειοθεραπεία και ακτινοβολίες. Επτά χρόνια μετά την επέμβαση δεν παρατηρήθηκε υποτροπή ή μετάσταση.

ΛΕΞΕΙΣ ΚΛΕΙΔΙΑ: όγκος, μαλακοί ιστοί, ραβδομυοσάρκωμα, χέρι, θέναρ