

IMAGING APPROACH IN CHILDREN WITH SOFT TISSUE PALPABLE RESISTANCE

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A palpable resistance in a soft tissue is a common pathology in children. The detailed child's history should be taken and clinical examination should be performed before imaging evaluation. Ultrasound is a first-line screening modality of palpable soft-tissue resistance. The main role of US is to stratify the soft tissue lesion and determine can it be safely left alone or followed by ultrasound, does it require further imaging characterization (mostly magnetic resonance imaging), surgical excision, fine needle aspiration or core biopsy. Most soft tissue lesions are benign, but we should be careful and try to exclude any potential signs of malignancy. We should be aware of the overlap between malignant and benign tumours in imaging characteristics. Imaging is not and cannot replace pathohistology. The purpose of this paper is to review the diagnostic approach in children with palpable resistance in a soft tissue, with a focus on imaging algorithms. Most of the soft tissue lesions are nonspecific. However, we present the lesions with specific benign diagnosis and emphasize that the interpretation of imaging findings. We should be familiar with the strength and potential pitfalls of different imaging modalities. Radiological diagnostics also plays an important role in monitoring the treatment response. Multidisciplinary approach is mandatory in children with suspected malignant soft-tissue tumour.

Descriptors: RESISTANCE, SOFT TISSUE, IMAGING, ULTRASOUND, CHILDREN

INTRODUCTION

The palpable resistances within a soft tissue in children are common pathology. The diagnosis of soft-tissue resistances in children can be challenging because

there is a broad spectrum of entities that can present as such. The soft-tissue masses occurring during childhood are most frequently benign with a wide spectrum of different entities (1). Malignant soft tissue tumours constitute 6% to 8% of all cancers in children less than 15 years of age (2, 3). Among them approximately 50% to 60% are rhabdomyosarcoma, whereas the remainder are non-rhabdomyosarcoma soft tissue sarcomas (fibrosarcomas, synovial sarcomas, the extra-osseous Ewing's family of tumours, malignant peripheral nerve sheath tumours, and inflammatory myofibroblastic tumours) (4, 5).

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Although the malignant etiology of resistances is rare, it remains a primary diagnostic concern and should be ruled out in untypical lesions. On the other hand, we should avoid overdiagnosis because it might have unfavourable effects on child and child's family physical and psychological status. The majority of malignancies occurring in children are different diagnoses than those in adults, and for those which occur in children and adults, there are some differences in management due to patient age, comorbidities, and differences in disease natural history. Patients younger than 18 years at initial diagnosis should be imaged according to the Pediatric Oncology Imaging Guidelines (6, 7).

The primary purpose of imaging is to confirm whether the resistance is a normal anatomical structure or a true pathological mass (8-11). Ultrasound (US) is imaging modality of choice in evaluating soft tissue resistance. If the resistance could not be recognized as benign lesion, further diagnostic steps, usually magnetic resonance (MR), have been done to narrow the differential diagnoses. US guided fine-needle aspiration or core-needle biopsy has an important role in this stage of imaging. When diagnosis of malignant tumour is established, the imaging has an important role in tumour's staging and in follow-up (evaluation of treatment response, early recognition of complications related to tumour or treatment, and in evaluation of late effects of treatment). Jedrzejewski et al. showed that a routine screening of asymptomatic children for cancer is not justified (12). Neoplastic lesions were found in only 0.02% of the examined children. On the other hand, screening for cancer should be regularly performed in children with paediatric cancer predisposition syndromes, most commonly by whole body MR (13).

The purpose of this paper is to point out the role of imaging to determine whether a mass represents a true tumour

or not, but also to achieve a more correct diagnosis, and determine the extension of the mass and its relation with the nearby anatomic structures.

CLINICAL APPROACH

The child's history and clinical examination are first and indispensable steps in the initial diagnosis of soft-tissue resistance. It is necessary to obtain relevant clinical information and perform a clinical examination in order to narrow down the diagnostic possibilities. The history must contain information about family and child's history, the age of the child at the soft tissue resistance presentation, the location and the duration of the presence of the palpable resistance (was it present at birth?), growth dynamic, whether there are any associated systemic symptoms, and whether any pre-existing conditions exist that may be associated with specific tumours (5). During the clinical examination it is important to note the location of the mass, the overlying skin abnormalities, the consistency of the soft-tissue mass, its relation to the surrounding tissues (mobility), look for the presence of pulsations or variations of volume on crying or Valsalva manoeuvre, the presence of systematized muscle atrophy, enlarged draining lymph nodes, and the size of the tumour (14).

Clinical risk factors for malignancy of soft tissue resistance include: onset of mass during the neonatal period, progressive or rapid growth, known primary tumour, firm mass greater than 3 to 5 cm, location underneath the fascia, and skin ulceration (9, 15). Any child with a soft tissue mass increasing in size, with size larger than 5 cm or localized underneath the deep fascia, whether or not painful, should be referred to a tertiary health care centre under suspicion of a soft tissue malignant tumour (5, 16).

IMAGING APPROACH

Diagnostic imaging

Superficial lumps and bumps may arise in soft tissue (skin, fat, muscle) or in the underlying bone. Further evaluation with imaging studies is required when the presence of the mass is not entirely clear on the physical examination, when the clinical findings are not completely contributory for a final diagnosis, and when there is uncertainty as to whether the mass is arising from soft tissue or underlying bones (10). Imaging is used to increase the diagnostic specificity and assess the local extension of the lesion and its relation to adjacent anatomic structures.

Ultrasound with colour/duplex Doppler, as most accessible and children-friendly method, is usually the first choice imaging modality to evaluate palpable soft-tissue mass. US with Doppler give information including size, shape, location, internal content and vascularity (15, 17-19). Different sonographic patterns of soft tissue masses are described that can be encountered when dealing with differential diagnosis of soft tissue masses (18). On the base of US imaging we can stratify the soft tissue lesion and determine, if the palpable lesion, a) can safely be left alone - "watchful waiting" (clinical observation of follow-up US), b) require further imaging characterization in cases where it can't be fully assessed (too deep or too large) or if malignancy is suspected, c) require surgical excision or biopsy (16). US-guided tumour sampling, fine-needle aspiration biopsy or core-needle biopsy, is usually performed. The area of sampling should be carefully chosen on the bases of imaging data to get representative sample. Contrast-enhanced ultrasound is a new diagnostic tool. Different perfusion patterns of soft-tissue lesions are better displayed by using second-generation US contrast agents. They serve as additional tool and help to differentiate

the benign from malignant lesion (23, 24). There are no dedicated studies included children with soft tissue tumours.

Magnetic resonance imaging is the reference modality for further evaluation of soft-tissue tumours due to its excellent tissue contrast. It is the diagnostic modality of choice for large and deep lesions and for some lesions for which US has not been diagnostic, when the clinical and US presentation is nonspecific, when the lesion is situated close to the central nervous system (paraspinal, facial mass) or when the mass is located on the midline (systematic search for and associated dysraphism) (5). MR angiography is useful method in delineating vessels of the vascular lesions, particularly in arterio-venous malformations to determine feeding artery and drainage vein.

The role of other imaging modalities is more limited. Plain radiographs may be useful in cases when the resistance of bone (osseous origin) or calcification of soft tissue (myositis ossificans) is suspected, and to assess bone involvement by a soft tissue mass. Computer tomography (CT) is rarely used due to radiation exposure.

Interpretation of the imaging findings

Interpretation of imaging findings has to be done by cautious because there is an overlap between benign and malignant lesions. The radiologist should be familiar with the strengths and potential pitfalls of imaging method when evaluates soft-tissue masses. Imaging is not pathohistology!

On US, echogenicity and homogeneity of the masses are nonspecific. Study of Hung et al. assessed the diagnostic accuracy of US in the assessment of malignant superficial musculoskeletal soft-tissue tumours - sensitivity was 94.1% and specificity 99.7%, respectively (25). US/Doppler risk factors for malignancy are

described (26, 28). The size of the lesion is not as important as in adults. However, when the soft-tissue lesion is bigger than 3 to 5 cm and has a heterogeneous structure, has to be further investigated. The borders of lesion can be well or ill-defined, with or without invasion of adjacent structures (Figure 1). Vascularity is not necessarily equated with malignancy and we should be careful in evaluating it. Complete absence of any detectable flow within a solid mass is a good criterion in favour of a benign lesion (5). Carra et al. systematically described the potential pitfalls at US evaluation of soft tissue masses: a) infected fluid cannot be differentiated from non-infected fluid; b) lipoma cannot be accurately diagnosed in some cases; c) hematoma cannot be reliably differentiated from a hemorrhagic solid soft-tissue neoplasm; d) some solid tumours can be mischaracterized as cystic; e) whether a palpable mass is truly of soft tissue origin or an extra-osseous component of an underlying bone lesion can be difficult to determine (17).

MR criteria in favour of malignancy are: absence of low signal intensity on T2-W sequences, signal heterogeneity on T1-W sequences, peripheral and centripetal contrast enhancement, contiguous invasion of bone and/or neurovascular bundles, and lower apparent diffusion coefficient (29, 30). However, there are overlaps between malignant and benign masses regarding vascularization (a predominantly peripheral enhancement pattern can also be observed in benign lesion), signs of marked aggressiveness (f.e. desmoid fibromatosis), and also in apparent diffusion coefficient values. The presence of oedema around the mass is not predictive criteria. Therefore, we should be careful when we interpret the MR imaging and should be aware that all MR criteria can be misleading (30-32).

When malignant soft-tissue tumour is diagnosed, the staging should be determined to plan proper treatment. For sta-

ging cross section imaging is required (CT and MR), nuclear or hybrid imaging (PET CT, PET MRI, SPECT CT).

Follow-up imaging

Follow-up imaging is frequently required in both benign and malignant lesions. Particularly nonspecific, but according to all criteria, benign lesion and lesions with specific diagnosis should be followed-up. Paediatric patients diagnosed with soft tissue malignant tumours often require systemic routine serial imaging during and after treatment. Imaging has an important role in evaluating the response to the treatment and in detecting the treatment complications (33). Familiarity with the expected posttreatment imaging findings in children with soft-tissue tumours can aid in the detection of complications. On the other hand, Vaarwerk et al. in their study found no survival advantage for patients whose relapse of rhabdomyosarcoma was detected before the emergence of clinical symptoms (34). The majority of relapses were detected as a result of clinical symptoms. These results show that the value of off-therapy surveillance is controversial, particularly because repeated imaging may also entail potential harm.

Imaging of late treatment consequences

Children with malignant tumours could experience long-term side effects of treatment. As the physical, mental and sexual development of children is incomplete at the time of treatment, they may suffer from growth or intellectual impairment, hormonal problems, infertility, and may develop secondary cancers.

CHARACTERISTIC IMAGING APPEARANCES OF SOFT TISSUE LESIONS FOR DEFINITIVE DIAGNOSIS

Most imaging findings are nonspecific and further diagnosis is necessary. There are certain clinical and imaging

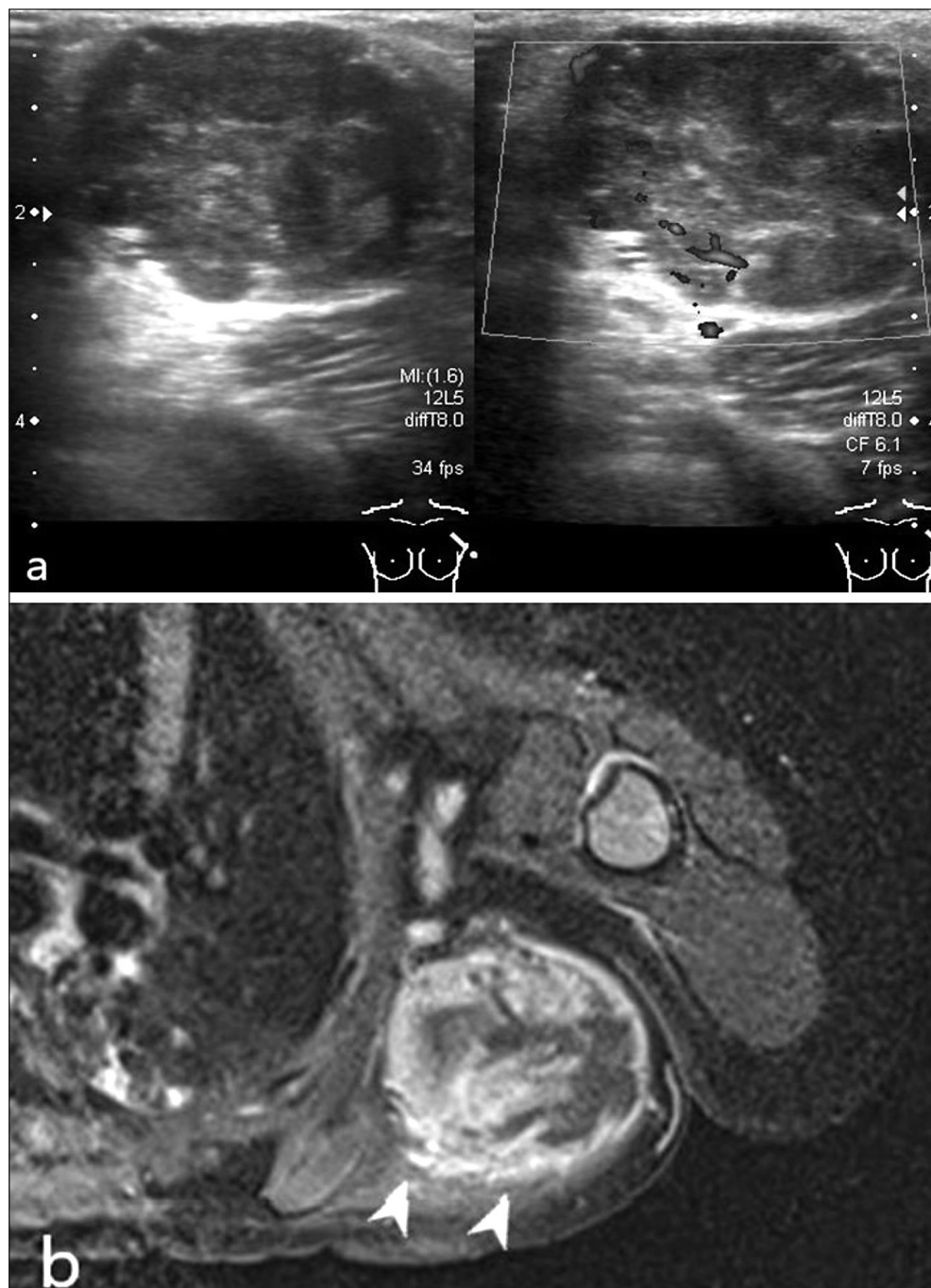


Figure 1.

A 1-year-old girl with palpable mass on the back side near the left shoulder. a) Ultrasound showed a heterogeneous soft-tissue mass, which was ill-defined at its posterior border, moderate vascularized on colour Doppler. b) Magnetic resonance imaging (contrast-enhanced T1 TSE fat sat sequence) showed relatively well-defined mass, except posterior border (arrowheads), intense and inhomogeneous contrast-enhanced mass. Imaging findings suggest the diagnosis of rhabdomyosarcoma and diagnosis was confirmed by US guided biopsy.

findings which allow diagnosis of selected soft-tissue masses with high diagnostic accuracy. We described specific imaging characteristics of some of the most common soft-tissue lesions in children (8, 10, 11, 15, 19, 21).

The vascular anomalies are common soft tissue lesions in small children and could be determined with high sensitivity and specificity because they have specific colour and also duplex Doppler patterns (14, 18-20). Vičič et al. suggested the diagnostic imaging algorithm in suspected avascular anomalies in children (14). US with Doppler is used for the initial diagnosis. At first, high and low flow lesions have to be separated on the bases on spectral Doppler US patterns. High flow lesions include haemangiomas, arterio-venous malformations, and arteriovenous fistulas. Haemangiomas are the most common benign tumour in children (infantile haemangioma starts to growth in a few weeks after birth) with its characteristic two phase natural history - proliferative and involution phase. Arterio-venous malformation starts to grow during puberty or after trauma in older children. Direct shunting between arteries and veins presents as shunts on spectral Doppler US together with high resistance arterial and venous flow (Figure 2). Low flow lesion is usually venous malformation and a cystic lesion without flow is lymphatic malformation. Vascular lesions could be part of various congenital syndromes.

Epidermod/dermoid cyst is usually congenital (most common diagnosing during the first year of life) and is located supraorbital, frontal or temporal quadrant. US examination shows a subcutaneous simple cystic lesion, well encapsulated, easy compressible, and with intact bone below the cyst (8).

Ganglion cysts are usually located on hand (wrist) and foot. Etiology is unknown, but they are related to trauma,

repetitive micro-trauma or degenerative changes. An anechoic lesion usually with the tail extending to the tendon sheath or joint is seen on US. More complex ganglion cysts have internal echoes, septations or loculations, even solid components and internal vascularity. In this cases MR is helpful.

Fibromatosis colli is a benign scar-like fibroblastic proliferation specific to the sternocleidomastoid muscle in children age 2 to 8 weeks. It presents with torticollis. On US a focal or diffuse enlargement (usually fusiform) of the sternocleidomastoid muscle is seen. On colour Doppler increased vascularity could be identified. It resolves spontaneously after physiotherapy (35).

Pilomatrixoma is a benign skin tumour associated with hair follicles. It presents as a small, hard lump under the skin, grows relatively slowly, without pain or other symptoms. It is located in head or neck. On US, it classically appears as target-shaped nodules with well-defined hypoechoic rim and hyperechoic centre (correspond to calcium deposits within nodule in 70 to 85%) (36).

Subcutaneous and soft tissue lymph nodes are in most cases reactive with preserved hyperechoic centre and hypoechoic cortex, ovoid shape, and well seen hilar artery (35). However, radiologists have to be aware of classic US criteria used in differentiating benign vs. malignant lymph nodes including size, shape, hilum, echogenicity, margins, structural changes, soft tissue oedema, flow, vessel location, vascular pedicles, vascular pattern, and impedance values (37).

Fat containing lesions have less characteristic appearance (22). Combination of different MRI sequences, including fat sat sequences, are helpful in assessing a fat-containing masses. In children the most common adipocytic tumours are lipomas

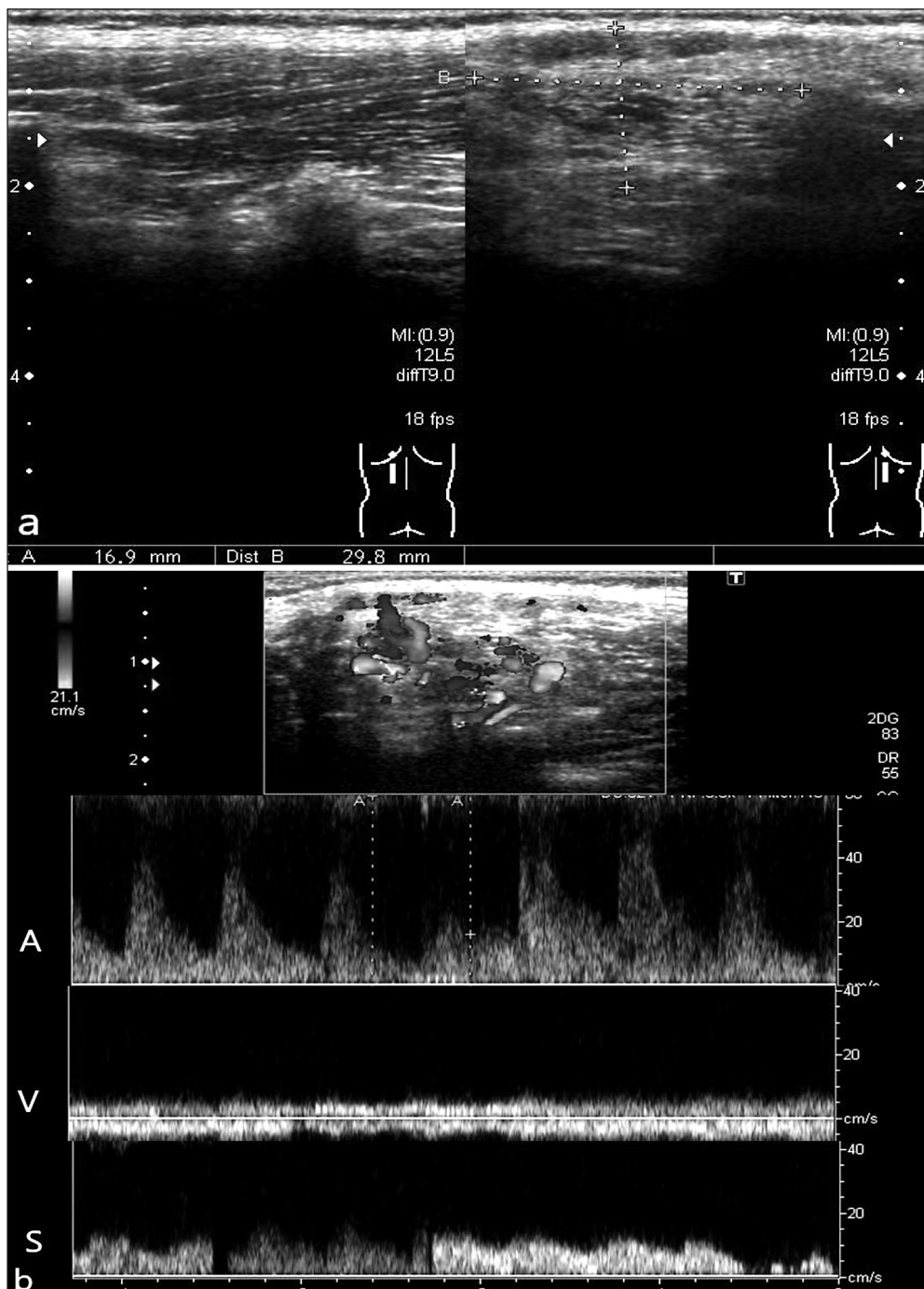


Figure 2.

Ultrasound (US) examination of a palpable paravertebral soft tissue resistance noticed in an 11-year-old girl 3 months ago. Painless, no skin changes, but pulsations were filled under tip of the fingers. a) grey-scale US showed inhomogeneous ill-defined isoechogenic lesion within the right paravertebral muscle (measure marks). b) Colour Doppler showed high vascularity of the lesion with high resistant arterial signals (A), low venous signals (V) and arteriovenous shunts (S) which is consistent with arterio-venous malformation.

(4% of all soft tissue tumours and 2/3 of all adipocytic tumours) (29). Lipoblastoma (30% of adipocytic tumours) is a benign neoplasm that is capable of invading surrounding soft tissue but lacks metastatic potential. It is usually diagnosed within the

first 3 years of life. The fat cells are of various stages of maturation, the more immature cells having less fat and, therefore, less "fat" signal intensity at MR imaging (Figure 3). Liposarcoma is extremely rare in children, occurring mostly in teenagers.

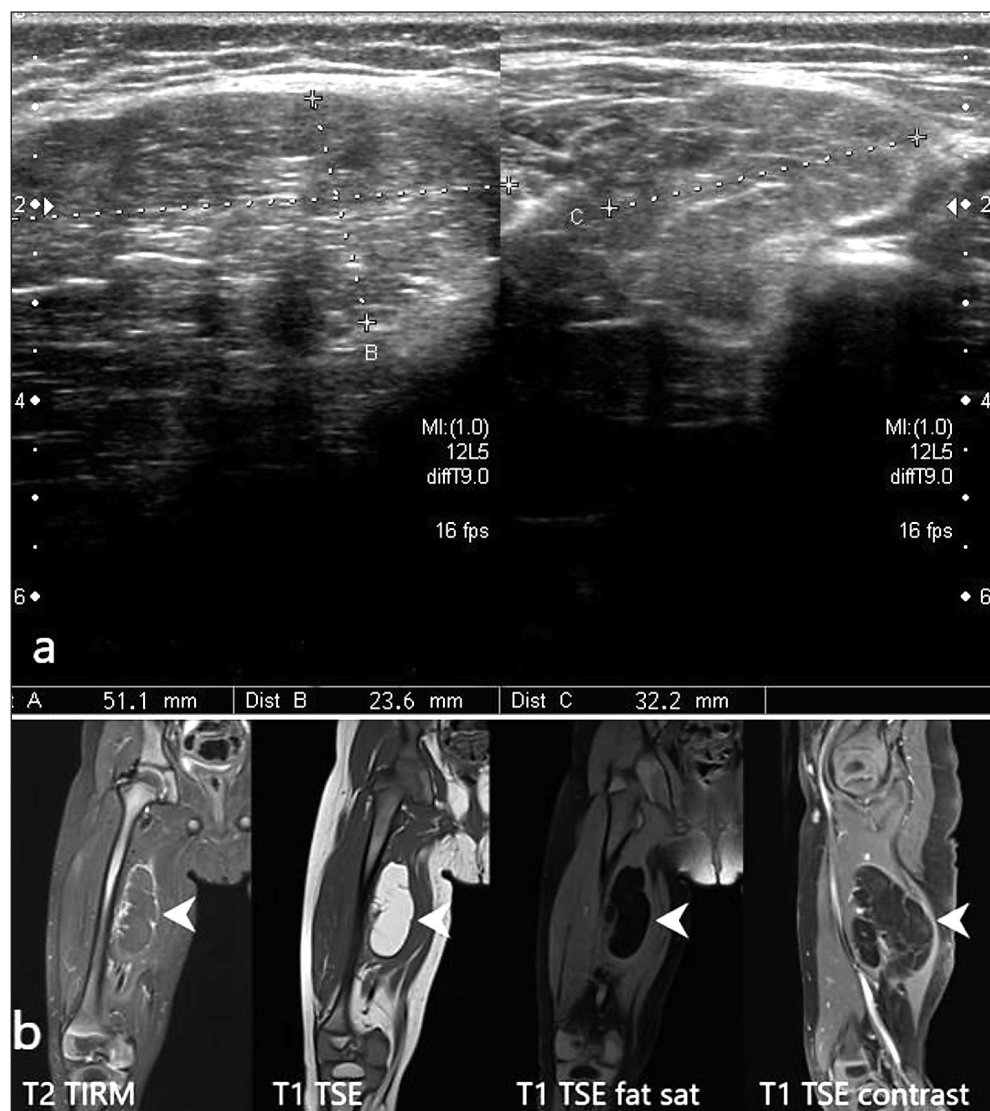


Figure 3.

A 2.5-year-old boy was admitted to the hospital because of painless soft tissue resistance in right thigh. a) On ultrasound an iso- to hypoechogenic formation was found in thigh muscle b) At magnetic resonance imaging a well-defined lobulated mass with mostly high signal intensity on T1 TSE sequence and hypointense on fat sat sequences (T2 TIRM and T1 TSE fat sat) was found in the area of musculus adductor magnus. Contrast-enhanced MR showed poor tumour vascularity and only septa enhanced. Intramuscular lipoblastoma was suspected and was confirmed by core needle biopsy. It was surgically removed.

The differentiation of lipoblastoma from liposarcoma is not possible at MR imaging. Even at histologic analysis, the diagnosis might be difficult and inconclusive.

Clinical history of injury or infection can help in differentiating soft tissue hematoma from abscess. US findings depend on the stage of haematoma or abscess. At the time of first presentation can look similar as well-circumscribed, heterogeneous solid mass with alternating hypo- and hyperechogenicity inside, hypervascularity in the surrounding soft tissue. The echointensity of blood clots in a hematoma changes with the age and usually reduces. A classic sonographic appearance of abscess is an anechoic or hypoechoic complex fluid collection. However, a soft tissue mass larger than 5 cm, with internal vascular Doppler flow, presenting without a clear mechanism of injury or with constitutional symptoms should be considered as suspicious for malignancy (Figure 1) (38). US has an important role in distinction between cellulitis and abscess what is crucial to choose the appropriate therapy (systematic antibiotics or incision and drainage).

CONCLUSION

Most soft tissue resistances are of benign origin. Certain clinical and imaging findings allow diagnosis of selected soft tissue masses with high diagnostic accuracy. However, most imaging findings are nonspecific and further evaluation is necessary. Particularly children with atypical or suspected malignant soft-tissue lesion should be managed by multidisciplinary team. Radiologist should be aware of the imaging overlap between benign and malignant lesions. Therefore, an interpretation of the imaging findings has to be done by cautious in atypical lesions. Imaging is not pathohistology.

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Sažetak

PRISTUP RADIOLOŠKOJ DIJAGNOSTICI U DJECE S MEKOTKIVNOM PALPABILNOM TVORBOM

Damjana Ključevšek, Lidija Kitanovski

Opipljive kvržice i ispupčenja u mekim tkivima su česta patologija kod djece. Prije slikovnih pretraga potrebno je uzeti detaljnu anamnezu djeteta i obaviti dobar klinički pregled. Prva izborna pretraga je ultrazvuk (UZ), koji nam daje slojevit pregled mekog tkiva. Nakon obavljene pretrage se možemo odlučiti o potrebi redovite kontrole bez dodatnih pretraga ili UZ slijeđenju. Često je potrebna pomoć drugih slikovnih pretraga (magnetska rezonanca) radi preciznije dijagnostike traženih promjena, a ponekad je potrebno napraviti i kiruršku intervenciju (eksciziju) ili biopsiju. Većina lezija mekih tkiva je benigna, ali je potreban oprez i potrebno je pokušati isključiti sve potencijalne znakove malignosti. Moramo biti svjesni preklapanja zloćudnih i dobroćudnih tumora u slikovnim karakteristikama. Svrha ovog rada je napraviti pregled dijagnostičkog pristupa kod djece s palpabilnim kvržicama u mekim tkivima, fokusiranom na slikovne dijagnostičke algoritme. Većina lezija mekih tkiva su nespecifične. Međutim, predstavljamo lezije s karakterističnim izgledom kod slikovnih pretraga i kako interpretirati nalaz snimanja. Trebali bismo biti upoznati s jačinama i potencijalnim zamkama različitih metoda snimanja. Slikovna dijagnostika je veoma bitna u praćenju odgovora na liječenje. Multidisciplinarni pristup je obavezan kod djece sa sumnjom na zloćudni tumor mekih tkiva.

Deskriptori: KVRŽICE, MEKO TKIVO, SLIKOVNA DIJAGNOSTIKA, ULTRAZVUK, DJECA