Image evaluation of the superficial soft-tissue tumors

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Learning objectives

The primary goal of this exhibit is to show that radiologists, when facing superficial soft-tissue lesions, by systematically using clinical history, lesion location and imaging characteristics on magnetic resonance, can help narrowing the differential diagnosis in an important subset of cases.
Background

Soft-tissue arises from the mesenchyme, which differentiates during development to become fat, skeletal muscle, peripheral nerves, blood vessels, and fibrous tissue. Soft-tissue tumors are histologically classified on the basis of the component that comprises the lesion, ranging from non-neoplastic conditions to benign and malignant tumors.

Presently, imaging shows limited ability to distinguish benign from malignant soft-tissue lesions reliably, given their variety and the overlap of their imaging characteristics.

Superficial soft-tissue masses can generally be categorized as mesenchymal tumors, skin appendage lesions, metastatic tumors, other tumors and tumor-like lesions, or inflammatory lesions.

They may also be divided into lesions that arise in association with the epidermis or dermis (cutaneous lesions), lesions that arise within the substance of the subcutaneous adipose tissue, or lesions that arise in intimate association with the fascia overlying the muscle.

Furthermore, characteristics such as the age of the patient, anatomic location of the lesion, specific imaging features and clinical manifestations can point towards the correct diagnosis.
Findings and procedure details

Presentation of a series of cases of benign and malignant superficial soft tissue tumors examined on our service, including cases of fibrosarcoma, liposarcoma, hidradenoma, merkel cell carcinoma, squamous cell carcinoma, hemangioma, neurofibroma and others, with analysis of the following characteristics: MRI imaging pattern, patient's age, site of the lesion, specific location of the lesion within the superficial tissue; along with a review of the recent literature pointing out the most important features that can help narrowing down the differential diagnosis.

1) HEMANGIOMA (Fig. 1 on page 10)

Hemangiomas are benign vascular lesions composed of various vessels by which they can be further histologically classified.

Lesions are usually identified in infancy or childhood but can occur in any age group. They can clinically manifest with bluish skin discoloration and a history of size fluctuation.

Cavernous hemangiomas are dilated blood-filled spaces and are frequently intramuscular in location. They may contain phleboliths and other calcifications that can be seen on radiographs. The most frequent subcutaneous type is a capillary hemangioma, which is composed of small vessels with flattened endothelium.

On images, classic cavernous hemangiomas have the generally characteristic imaging appearance of infiltrative lesions in which serpentine vessels interdigitate with fibroadipose tissue, whereas capillary hemangiomas often present as well-circumscribed masses.

On MR images, most hemangiomas show circular, linear, or serpentine high T2 signal caused by slow flow in vascular channels, while rapid flow can demonstrate a signal void on images. Areas of high T1 signal may also be found, representing fat tissue or hemorrhage. Identifying phleboliths on CT and radiographs can be helpful in characterization.

2) EPIDERMAL CYSTS (Fig. 2 on page 10)

Epidermal cysts are cysts filled with keratin debris and bounded by a wall of stratified squamous epithelium. It usually occurs in the hair-bearing areas of the body, such as the scalp, face, neck, trunk, and back [1].

They result from the proliferation of epidermal cells within a circumscribed dermal space and are probably formed by several mechanisms, such as remnant ectodermal tissues...
misplaced during embryogenesis, occlusion of the pilosebaceous unit, or traumatic or surgical implantation of epithelial elements [2].

On MR images, epidermal cysts are well-defined round or ovoid lesions of high signal intensity on T2-weighted images, low to intermediate signal intensity on T1-weighted images and no enhancement centrally but may present thin peripheral enhancement. In many cases, T2-weighted images show variable low-signal components within epidermal cysts.

3) NEUROFIBROMA (Fig. 3 on page 11)

Neurofibromas are benign peripheral nerve sheath tumors usually solitary and sporadic, however, there is a strong association with neurofibromatosis type 1 (NF1). Neurofibromas most frequently affect patients who are 20-30 years old and have no sex predilection [3]. Three types of neurofibromas are described: localized, diffuse, and plexiform.

The most common form of neurofibroma is the localized type, representing approximately 90% of these lesions, usually presenting in a solitary form and not related to NF1 [3]. Superficial neurofibromas may occur in the subcutaneous fat layer as well as the dermal and epidermal layers of the skin, although deep lesions with involvement of larger nerves may also occurs [4].

Diffuse neurofibromas are lesions poorly delimited in the fat layer of the subcutaneous tissue, which is infiltrated along the septa of connective tissue. They primarily affects children and young adults and are associated with NF1 in about 10% of cases [5].

Plexiform neurofibromas are practically pathognomonic of NF1, usually arise in childhood and precede cutaneous neurofibromas. They represents a diffuse involvement of a long nerve segment and its branches with tortuous expansion, and its gross appearance has been described as a "bag of worms" [5].

On MRI neurofibromas show low to intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images. After the administration of paramagnetic agent inhomogeneous contrast enhancement is seen in two third of cases [3]. A characteristic of the neurogenic tumor is the "target sign" observed in the T2-weighted sequences and characterized by a low to intermediate signal in the central region and a peripheral high signal, which corresponds to central fibrous tissue and peripheral myxoid tissue. This imaging characteristic is most commonly observed in the neurofibroma, but can also be seen in the schwannomas [5].

4) LIPOMA (Fig. 4 on page 12)
Lipomas are the most common soft-tissue tumor and are composed of adipose tissue. The incidence of lipomas is up to 2.1 per 100 individuals [6]. Superficial lipomas account for approximately 16% to 50% of soft-tissue tumors [7]. Superficial lipomas mainly are found in the subcutaneous fat layer, but lipomas also may occur in deep locations beneath the superficial fascia. Deep lipomas may be larger and when found at the extremities they are usually intramuscular [7].

On MR images lipomas are isointense relative to subcutaneous fat in all MRI sequences. The classic lipoma is composed entirely of fat, without areas of nodularity or thickened septations. However, a substantial percentage of lipomas may have associated non-adipose components [8].

5) LIPOSARCOMA (Fig. 5 on page 13)

Liposarcoma is the second most common soft-tissue sarcoma, accounting for 16% to 18% of all malignant soft-tissue tumors [4]. The lesions usually are located in the extremities, particularly the thigh, and in the retroperitoneum in adults.

They are tumors that usually affect the deep soft tissues, being rarely found in the superficial tissues [9]. In case a superficial fatty lesion does not meet the imaging criteria for the diagnosis of lipoma, a lipoma variant should be considered as a diagnostic possibility, as should liposarcoma. It is important to remember that other subtypes of liposarcoma (dedifferentiated, myxoid, and pleomorphic) may contain minimal or no visible fat [8]. Another important characteristic of adipose tumors is that there is no malignant transformation of a lipoma [9].

In the imaging studies, the characteristics that may indicate the diagnosis of a well differentiated liposarcoma include lesion size greater than 10 cm, presence of thick (>2 mm) septa (diffuse or focal), presence of non-globular and / or nodular areas or masses and composition of lesions with less than 75% fat [8].

6) DERMATOFIBROSARCOMA PROTUBERANS (Fig. 6 on page 14)

Dermatofibrosarcoma protuberans is a superficial neoplasm that arises in the dermal and subcutaneous tissues, but large lesions may extend into the deeper soft tissues. Correspond to the most common mesenchymal superficial malignancy and accounts for about 6% of all soft-tissue sarcomas [4]. Males are more often affected than are females, and the tumor is most commonly seen in the 2nd through 5th decades of life [7]. Lesions are seen most commonly in the trunk but have been observed in both the upper and the lower extremities.

On MR images, the appearance of dermatofibrosarcoma is nonspecific, with signal often lot to isointense to that of muscle on T1-weighted images and signal intensity higher than that of fat on T2- weighted images [7], with moderate enhancement. Areas of hemorrhage
may be seen within the tumor and a linear extension along the skin surface also may be suggestive of the diagnosis.

7) FIBROSARCOMA (Fig. 7 on page 15)

Fibrosarcoma is defined as a malignant tumor, composed of fibroblasts with variable collagen and, in classical cases, a herringbone architecture. It is a diagnosis of exclusion [10]. It is part of the group of spindle cell sarcomas that have potential for locally destructive growth and recurrence, along with a significant risk of distant metastasis, and characterized by a fundamental spindled shape of neoplastic cells [11].

Tumors typically arise in middle aged or older adults, with no predominance by gender, and corresponds to 1% to 3% of sarcomas in adults [10]. It involves the deep soft tissues of the proximal extremity or trunk and clinically presents as a mass with or without pain.

On MR images, the solid elements of the lesion usually present isointense signal in T1-weighted images and hyperintense signal in T2-weighted images, being able to present areas of necrosis, hemorrhage or cysts or even pseudocapsule of low signal. The enhancement by the paramagnetic agent is variable.

8) NODULAR HIDRADENOMA (Fig. 8 on page 16)

Nodular hidradenoma is a benign skin adnexal neoplasm located in the dermal layer with no connection with the overlying epidermis. Recently a subdivision in two groups has been suggested: tumors with eccrine differentiation and tumors with apocrine differentiation [12].

It is found in middle-aged adults, being more prevalent in females [13]. The most affected sites are the scalp, face, trunk and proximal extremities. Clinically, nodular hidradenoma manifests as a solid or cystic nodule, well delimited, with a variable diameter of 0.5 to 3 cm, slow-growing and endophytic. Skin changes such as changes in colour and thickness often accompany the lesion and may also be associated with serous or bloody discharge. This tumour has been reported to rarely undergo malignant transformation [14].

The presentation on MRI is varied and may present as hypointense to isointense signal in T1-weighted images and hyperintense signal in T2-weighted images depending on their composition. It usually presents as a superficial dermal lesion and hence is diagnosed early and often excised without much investigation [14].

9) MELANOMA (Fig. 9 on page 17)

Primary cutaneous melanoma is the most common subtype of malignant melanoma, a malignant neoplasm arising from melanocytes, found predominantly in the basal layer of the epidermis, but also found in other parts of the body. An incidence rate of 2.8 -
3.1 cases per 100,000 individuals is estimated [15] and accounts for the majority of skin cancer-related deaths because of their tendency to metastasize.

The typical lesion of cutaneous melanoma is an asymmetric macule or nodule with irregular borders, frequently with variations in color within the lesion [16]. Metastatic melanoma may be manifested with a similar pattern of multiple subcutaneous nodular lesions, and its presence must be considered in a patient who presents with multiple subcutaneous nodules [4].

MR imaging may be useful for differentiating melanoma from benign pigmented skin tumors such as blue nevi and intradermal nevi [17]. Lesions with a higher melanin content have been shown to have higher or mixed signal intensity on T1-weighted images. In general, the T2-weighted signal intensity of malignant tumors was higher than that of subcutaneous fat, whereas that of benign lesions was lower than that of subcutaneous fat. The higher T2-weighted signal intensity of malignant lesions may be secondary to the presence of hemorrhage [7].

10) MERKEL CELL CARCINOMA (Fig. 10 on page 18)

Merkel cell carcinoma is a rare aggressive neuroendocrine tumor that usually presents in the elderly population, with most cases occurring after 65 years of age [7]. Studies have reported an incidence of 0.15 to 0.6 cases per 100,000 individuals worldwide [18], with an increased incidence of this neoplasm in immunosuppressed patients with leukemia, organ transplantation or rheumatoid arthritis [19].

Merkel cells are thought to arise from the neural crest and are believed to be primary neural cells found within the basal layer of the epidermis [20]. Patients may present with painless red or violaceous nodules on sun-exposed skin. The Merkel cell carcinoma follows an aggressive clinical course and at diagnosis approximately 66% of patients present local disease, 27% have nodal involvement and 7% have distant metastasis [20].

On MR images, Merkel cell carcinoma is characterized by nodules in the superficial planes affecting the skin and subcutaneous tissue associated with adjacent fat stranding due to the associated inflammatory component. The lesion shows iso to high signal intensity on T1-weighted images and high signal intensity on T2-weighted images, in comparison to the signal of the muscle. Larger lesions may show heterogeneous signal in T2-weighted images [7].

11) CUTANEOUS SQUAMOUS CELL CARCINOMA (Fig. 11 on page 19)

Cutaneous squamous cell carcinoma is the second most common cutaneous malignancy, and is frequently associated with chronic sun damage that affect mainly sun-exposed areas such as the face, scalp, neck, arms, and hands [21]. The current estimated
incidence of cutaneous squamous cell carcinoma is 15 to 35/100,000 people per year [22]. Exposure to ultraviolet radiation is the most common cause of this type of cancer.

The principal precursor of cutaneous squamous cell carcinoma is actinic keratosis, which is characterized by scaly lesions, typically 2 to 6 mm in diameter, that are more easily felt than seen; they may be the same color as the skin, pink, or brown. They can involute or persist, and affected persons usually have many lesions, some of which may evolve into squamous cell carcinoma [23].

On MR images, cutaneous squamous cell carcinoma is characterized by hypointense signal in T1-weighted images, intermediate signal in T2-weighted images and heterogeneous contrast enhancement. Imaging tests play a key role in the investigation of distant metastases.
Fig. 1: 13-year old male with a lobulated lesion in the subcutaneous of the inferior abdominal wall. Unenhanced Axial CT (A) shows calcifications inside the lesion, suggesting phleboliths (arrows). MRI study shows a T2 hyperintense lesion (B) with T1 isointensity (C). Dynamic contrast imaging (D - Arterial; E - Portal; and F - Late Phases) demonstrates progressive enhancement. Biopsy revealed a hemangioma.

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**Fig. 2:** 63 year-old female with a heterogeneous and well-defined mass anteriorly to the clavicular portion of the deltoid. MRI shows a T1 hypointensity (A) with a heterogenous intermediate T2 intensity with interior septa (B) and no significant enhancement (C). Biopsy revealed an epidermoid cyst.

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**Fig. 3:** 67 year-old male with a well-defined nodule in the subcutaneous of the posteromedial region of the forearm. It also has apparent contact with the peripheral neurovascular bundle. The MRI study shows intermediate T1 intensity (A) with T2 hyperintensity (B) and periferical enhancement (C). Biopsy revealed a neurofibroma.

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Fig. 4: 56 year-old female with an encapsulated and well-defined lesion in the subcutaneous adjacent to the medial epicondyle of the right elbow. MRI shows a T1 hyperintensity (a) and T2FS hypointensity (B) and no significant enhancement (C). Biopsy revealed a lipoma.

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**Fig. 5:** 75 year-old female with a multisseptated intramuscular lesion in the distal third of the vastus medialis, invading the adjacent soft tissue. MRI shows T1 (A) and T2 (B) hyperinsity with signal loss in T1FS (C). it also shows peripheral and septal enhancement, with an enhancing nodule in its medial portion (D and E). Biopsy revealed a lipossarcoma.

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Fig. 6: 58 year-old female with an exophytic tumor in the dorsal thoracic region with slow and progressive growth. MRI shows a lesion with well-defined margins and in the skin and subcutaneous of the reported region. It has a heterogeneous and intermediate signal intensity in T1 (A) and T2 (C), with some interior areas of T1 and T2 hyperintensity, suggesting hemorrhagic content. It has a heterogeneous enhancement by Gd (B). There are no signs of invasion of the adjacent paravertebral muscles. Biopsy revealed a dermatofibrossarcoma.

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Fig. 7: 44 year-old female with a vegetating, lobulated and ulcerated lesion in the superficial and deep soft tissues of the anterior portion of the left leg. MRI shows an intermediate signal intensity in T1 (A) and T2 (B) and intense contrast enhancement (C). Biopsy revealed a fusiform cell fibrossarcoma.
**Fig. 8:** 56 year-old female with a soft tumor in the dorsal side of the left forefoot (A - lateral forefoot x-ray). MRI demonstrates a well-defined and multiloculated lesion with T2 hyperintensity (B and D) T1 hypointensity (C) with heterogeneous enhancement (E). Biopsy revealed a hidradenoma.

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**Fig. 9:** 58 year-old male multiple nodules in the dorsal region. CT scan showed confluent isoattenuating nodules (A) with intense and heterogeneous enhancement (B) in the skin and subcutaneous of the left paramedian dorsal region. There were also axillary enlarged and enhancing lymph nodes at the same side by the level of the cervicothoracic transition (C). Biopsy revealed a melanoma.

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Fig. 10: 74 year-old male with a tumor in the medial part of the left leg with associated inguinal lymphnode enlargement. MRI showed a lesion in the skin and subcutaneous of the reported region with T1 intermediate signal intensity (A), T2 and STIR heterogeneous hyperintensity (B and C) and periferal enhacement by Gd (D). Biopsy revealed a Merkel cell carcinoma.

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Fig. 11: 74 year-old male with a skin lesion in the first finger of the right hand. Patient also presented movement and sensibility impairment in this finger. MRI showed a nodular skin lesion in the posterolateral part of the first finger that was invading the subcutaneous, as well as the underlying ligaments and bone. It has a discrete hyperintense signal in PD (A), intermediate T1 intensity (B and E) with heterogeneous contrast enhancement (C, D and F). Biopsy revealed a squamous cell carcinoma.

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Conclusion

Despite superficial soft-tissue tumor`s wide variety and their overlapping imaging characteristics, by considering some aspects like patient's age, anatomic site of the mass and the specific location of the mass within the superficial tissue, the radiologist can help narrowing the list of differential diagnosis, and in some cases determine the correct one, therefore avoiding unnecessary invasive procedures.
References


