

Histiocytomas and Fibromas, For Surgeons

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ABSTRACT

In regard to soft tissue tumors, surgeons are often confronted with final pathological diagnosis and terms that might be unfamiliar and alarming, such as Histiocytoma and its variants. Pathologic versus clinical titles to Histiocytomas do differ however a simplified description and classification for surgeons is being discussed and recommended. The common Dermato-fibroma is the cutaneous benign version of Histiocytoma. Atypical fibrous histiocytoma and the Deeply located histiocytoma categories can be benign or malignant.

Keywords

Histiocytoma, Fibrous, Dermato-fibroma, Calcification.

Abbreviations

STT: Soft tissue tumor, DF: Dermato-fibroma, NF: Neuro-fibroma, BFH: Benign fibrous histiocytoma, AFH: Atypical fibrous histiocytomas, MFH: Malignant fibrous histiocytoma.

Introduction

Soft tissue tumors (STT) when vague in presentation, are frequently being referred to plastic and reconstructive surgeons. The obvious reason being, those could be of any tissue origin. Hence, depending on the work up, differential diagnosis and eventually the intraoperative findings; each situation might need specific intraoperative strategy. This might involve nerve reconstruction for example in case of Neurofibroma (NF) when arising from a specific motor nerve, or the need for skin grafting or flap procedure in other situations. Exclusion of a sarcomatous lesion with the appropriate biopsy technique and strategy also requires plastic and or oncologic surgical expertise. Apart from the typically and clinically obvious benign tumors such as superficial lipomas or otherwise known childhood vascular lesions like hemangiomas, all others bear some worrisome or alarming concerns unless proven otherwise [1]. Patients presenting with systemic manifestations combined with a mass are also being evaluated by medical

oncologists from the beginning, as a multidisciplinary approach.

Cutaneous “Fibromas” are very common and are often being referred for excisional biopsy. Pigmentation, and a firm-hard nodule fixed to the skin can be quite worrisome to patients and often dermatologists for obvious reasons. However, such signs are classic of the commonly benign tumor, Dermatofibroma (DF) (Figure 1). Likewise, for instance seborrheic keratosis happens to be the most commonly biopsied skin lesion due to its darkly pigmented appearance and resemblance to malignant melanoma to the novice eye. Pedunculated rubbery skin nodule is typical of cutaneous NF, which may present either as a skin tumor or a deeper lesion arising from a significant nerve or both (Figure 2 a,b,c). A specific form of NF is the “plexiform” kind of lesion, classically described to have the consistency of “a bag of worms”, exclusively described in Neurofibromatosis type 1 patients (Figure 3).

“Histiocytoma” in humans, which is the main subject here, pathologically indicates a tumor consisting of histiocytes, which are part of mononuclear phagocytic system. They comprise a wide group of lesions ranging from benign to malignant. In fact, the benign common DF is a subtype of histiocytoma. A case of an upper arm solid tumor in a young girl, was pathologically diagnosed as a benign fibrous histiocytoma (BFH) is presented.



Figure 1: A classic Dermato-fibroma, nodular fibrotic cutaneous lesion with pigmentation.



Figure 2: Neuro-fibroma: Cutaneous form, may occur as a single lesion or often as multiple in Neurofibromatosis type 1 (a). Pairing a neurofibroma gives a gritty feel, the fibrous hard core could be seen (b). A Neurofibroma arising from the Median nerve, has been dissected in a patient with Neurofibromatosis type 2 (c).



***Figure 3:** Young adult with Neurofibromatosis type 1, with typical

“plexiform neurofibroma” of orbito-temporal region.

Case Presentation

A 10 years old, healthy girl presented with a history of right upper arm swelling of 1year duration, otherwise asymptomatic (Figure 4). The mass was incidentally noted by the parents, slowly growing in size and they denied any history of trauma. It was initially diagnosed in another facility as vascular malformation based on ultrasound findings of a 4.6 x 9.7 cm vascular soft tissue lesion with calcifications in its proximal aspect (Figure 5). Hence, the patient was referred to vascular surgery clinic at first. As per vascular surgery team and based on MRI findings; the mass of 2x3.2x4.6 cm, displacing the biceps muscle postero-medially and separate from muscle fibers, was suspected to be a nerve related tumor (Figure 6 a,b) and hence referred to plastic surgery.



Figure 4: 10 years old female with a soft tissue mass, upper arm anteromedially of 1year duration, with differential diagnosis of a vascular mass and a tumor of nerve origin.

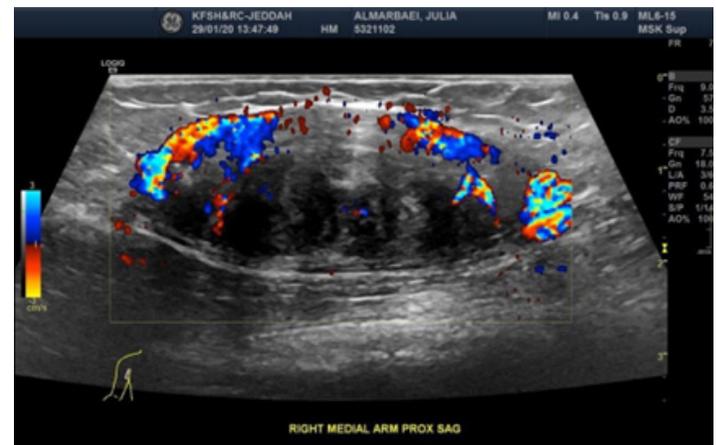


Figure 5: Ultrasound findings suggestive of a solid lesion of high vascularity and proximal calcification.

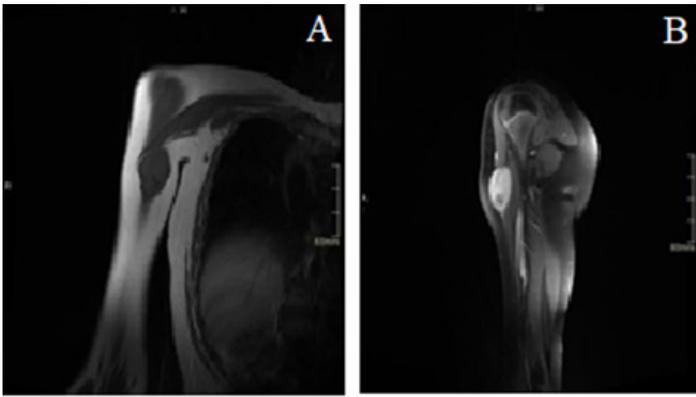


Figure 6: MRI findings; a mass with high T2 and low T1 signal intensity (a), avid enhancement post contrast administration. No vessels noted within the lesion, suggesting unlikely to be a vascular malformation, and more of a nerve related (b).

Clinical examination revealed a firm, non-mobile mass about 4x3 cm, not well circumscribed, non-pulsatile, located in the antero-medial aspect of the right arm. The overlying skin was free of any tethering or changes. Distal neuro-vascular exam was normal. A plain radiograph showed an oval soft tissue mass containing punctate calcifications (Figure 7). The Patient was taken for exploration, an encapsulated mass was entirely excised (Figure 8 a,b). Final histopathology came as benign spindle cell tumor of fibroblasts and histocytes with no malignant features, favoring the diagnosis of Benign fibrous Histiocytoma (BFH). The Patient has been followed for over 2 years or until this publication, with complete healing and no recurrence.



Figure 7: The Plain radiograph, oval soft tissue shadow with punctate calcification.

Discussion

The term “Fibrous Histiocytoma” could be alarming on a first instance due to the historically heard term of Malignant fibrous histiocytoma (MFH). Pathologic classifications do evolve with time, MFH is now considered as an undifferentiated pleomorphic sarcoma. Fibrous histiocytoma is infrequent in patients under 20 years of age. It more commonly affects adults, the mean age

of presentation is 40 years, and it is rarely being heard among the differential diagnosis of a child with STT. For clinicians or surgeons, a “Benign” fibrous histiocytoma (BFH) seems to be a generally less recognized term. When involving skin only, BFH is in fact merely the common “Dermatofibroma” (DF) [2].

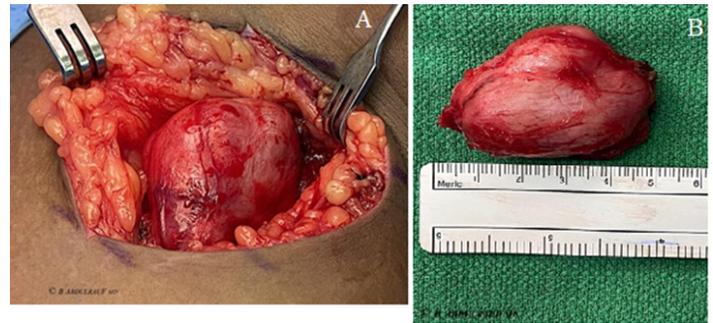


Figure 8: Intraoperatively, a multilobulated mass with a pseudo-capsule surfaced with a net of vessels, and unrelated to major nerves or significant vascular pedicles (a). Fully excised with maximum dimensions of 5.0x 3.0 cm (b).

There are few not uncommon STTs that are considered locally aggressive and may even simulate or resemble an aggressive sarcoma, these include Dermatofibrosarcoma protuberans, abdominal Desmoid tumor and less commonly Nodular Fasciitis [3]. Dermatofibrosarcoma protuberans (commonly abbreviated as DFSP) is another misnomer. In fact, it has no relationship to DF and it is not a sarcoma in the first place. However, it can transform into a fibrosarcoma’s variant or rarely into pleomorphic sarcoma [2]. Such locally aggressive tumors are supposedly curable with wide surgical resection and reconstruction, often with a single well-planned stage.

DF and NF are the 2 most common kinds of fibromas arising in the skin. When solitary, both may appear similar (dome shape or button like). DF is usually pigmented, and when it is hard and dark, often reported as “sclerosing hemangioma”. DF is more common in women and often a history of insect bite or a minor trauma is a very common scenario. Synonyms to DF are: “BFH” and “Nodular sub-epidermal fibrosis”. It is the most common mesenchymal or spindle cell origin tumor of the skin.

Fibromas in general including the cutaneous form of NF do share 3 characteristic features: being sub-epidermal lesions; nodular and obviously fibrotic. NF have both neural (ectodermal) and fibrous (mesodermal) origins. It is important to realize that NF could be a pure “cutaneous” lesion(s) or otherwise, arising along course of a peripheral motor or sensory nerve and even as a skull base tumor, as part of Neurofibromatosis type 2 (figure 9 a,b). Histologically, NF is composed of Schwann cells, fibroblasts, peri-neural cells, mast cells, macrophages with intermixed axons. The cutaneous NF would have skin adnexal cells as well. Schwannomas or Neurilemmomas are different since they are composed of Schwann cells only and arise from a single axon or fascicle.

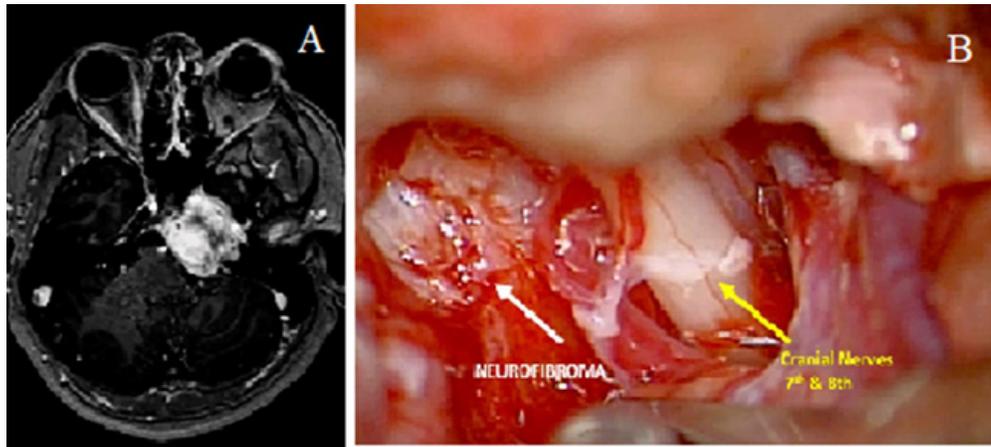


Figure 9: Intracranial invasive large vestibular Neurofibroma (much less common than a schwannoma), CT scan (a) and a microscopic intraoperative-view, white arrow indicates the tumor and the yellow indicates cranial nerves 7th and 8th (b), *Courtesy of Prof. Saleem Abdulrauf.

DF have been histologically classified as cellular, aneurysmal, and atypical. Immunohistochemistry might have a limited role in differentiating these subtypes [2]. Xantho-Fibroma or (Fibro-Xanthoma) is another type of a fibroma, which may involve skin or other tissues including bone. Although it can be a benign lesion however, the atypical and malignant forms of it (xantho-sarcoma) are highly aggressive (Figure 10).



Figure 10: Atypical Xantho-Fibroma (or Fibroxanthoma), affecting the scalp in an elderly patient, and quite an aggressive course of disease.

All Fibrous histiocytomas are characterized histologically by proliferation of polymorph-nuclear, spindle shaped, or histiocytoid cells and or multinucleated cells, admixed with inflammatory cells [4].

The Atypical fibrous histiocytoma (AFH) is a well-established entity in pathology literature. It was previously known as a Dermatofibroma- with “monster cells”. The wide range of these tumors are grouped under the term Fibro-histiocytic tumors, histologically they contain histiocytes and fibroblasts. Due to a

degree of nuclear atypia, AFH is often histologically mistaken for sarcoma [2,4].

Adding the affix of “Atypical” to any common and essentially benign lesion gives the liberty of reporting findings that seemed unfamiliar, furthermore even though the resection was total but the tumor recurs afterwards or behaves unusually, then the description “Atypical” would have been proven to be invaluable in retrospect. As an example, same applies to the concept of “Atypical nevi”.

Kaddu et al. analyzed 59 cases, which were pathologically diagnosed as AFH aiming to better identification of this category and its implications. Median age was 38, as a solitary lesion in all cases. Most common site was in extremities. A combination of histologic signs in terms of pleomorphism, mitotic activity, atypical mitoses were reported, three cases had local recurrence. Two of the reviewed cases were reported to have developed distant metastases, whose histological features were not distinct from rest of the group. The authors stressed on the importance of identifying this category of (AFH), not to confuse it with a pleomorphic sarcoma and exposing the patient to an aggressive treatment [5].

The third main category of histiocytomas would be the “Deeply located” one. Gleason and Fletcher reviewed 69 of such apparently “benign” BFHs that were found at deeper anatomical planes. Most were in extremities but few occurred as head and neck tumors as well visceral. Four of the cases had histologic features of AFH. The authors indicated, when numerous foam and giant cells are present, the main differential diagnosis would be giant cell tumor of soft tissue.

Their cases with local recurrence were not in fact totally excised in the first place. However, two of the patients had distant metastases, both ultimately died of the disease. Those two cases had large size tumors on presentation ranging 6-9 cm [6]. Our patient who was presented above seemed belonged to this (Deeply located histiocytoma) category however, it did not have any

Histiocytomas

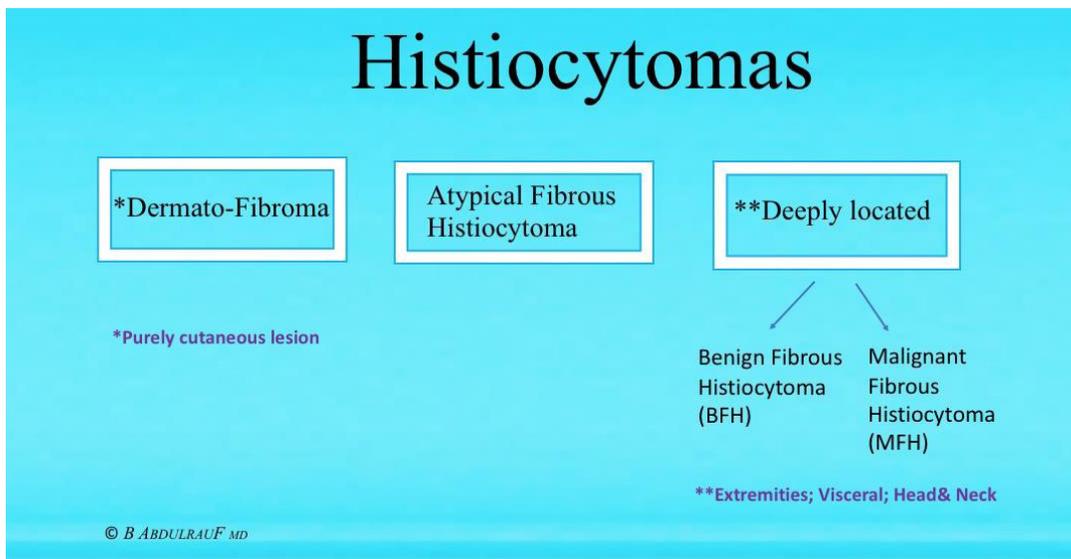


Figure 11: Histiocytomas, categories and a practical guide.

atypical features and was reported to be a BFH. A calcification (or mineralized densities) was seen on the plain radiograph and reported as well on ultrasound in our case, this made it more interesting, concerning and challenging in terms of differential diagnosis. Ossification generally indicates new bone formation in ectopic sites such as in the case of myositis ossificans. Calcification versus ossification is still not a straightforward distinction for nonradiologists. However, the distinction could narrow down the differential diagnosis. Such radiologic findings raise the suspicion or necessitate exclusion of a sarcomatous lesion.

Kwee & Kwee have thoroughly discussed various soft tissue lesions with calcification or ossification, this included a wide list of true neoplasms and other metabolic or inflammatory processes. Some of the notable lesions, which might be of special interest, and significance to surgeons included: Venous malformation; Calcific tendinopathy; Lipoma with metaplasia; Calcifying aponeurotic fibroma; Ancient schwannoma [7]. In our case of BFH, calcification was associated and to our knowledge, this has not been previously reported.

Speaking of calcification in STTs, it would be important to mention and add “Pilomatrixoma” or “Calcifying epithelioma of Malherbe”, a well-known childhood STT. It arises from hair follicles, usually presents as a hard subcutaneous red to blue mass, overall size of 1 cm or less. More commonly occurs on hair bearing areas (preauricular is most common site) but may also occur on extremities or trunk. It is commonly being misdiagnosed as epidermal cyst or dermoid. Some cases might even undergo rapid growth. If clinically recognized and diagnosed meanwhile symptomatic, it could be observed otherwise excisional biopsy has been the usual approach [8,9].

For simplicity, Histiocytomas may present as one of 3 main categories: 1. Dermato-fibroma, a purely cutaneous lesion

recognized clinically and confirmed histologically or 2. Atypical FH as a pathological diagnosis of a STT, and 3. A deeply located STT which is pathologically diagnosed either as a BFH or a MFH (Figure 11).

Summary

1. A clarification on histiocytomas and fibromas has been presented, from the perspective of surgeons who deal with STTs.
2. Final Pathological diagnoses and terms can sometimes sound sophisticated and worrisome on first instance to surgeons. This is often the case in STTs.
3. The term Histiocytoma can be alarming; however, it is evident that it has benign variants too.
4. One of the most common cutaneous lesions clinically diagnosed as DF, must be routinely and carefully examined pathologically, due to its reported atypical type or other variants.
5. Fibromas other than DF, which include Neuro-fibromas and Xantho-fibromas, are not Histiocytomas; however, they are quite diverse with variable presentations.
6. Radiological heterotopic calcification in our presented case of BFH, has not been previously reported. This means BFH should be included in the differential diagnosis of STT with calcification.
7. The final diagnosis for the variants; benign, atypical or a malignant type of Histiocytoma (BFH, AFH & MFH) and their synonyms is based purely and obviously on pathological details, which in turn would dictate the course of management to be considered by surgeons or clinicians.

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