applied sciences

MDPI

Review

Myoepithelioma of the Hand: A Systematic Review

Guido Bocchino [1], Giacomo Capece [1], Silvia Pietramala [1,*], Giuseppe Rovere [1,2], Lorenzo Rocchi [1,3], Pasquale Farsetti [2], Giulio Maccauro [1] and Camillo Fulchignoni [1,3]

[1] Department of Orthopaedics and Traumatology, Fondazione Policlinico A. Gemelli IRCCS-Università Cattolica del Sacro Cuore, 00136 Rome, Italy; guido.bocchino@hotmail.it (G.B.); giacomocapece97@gmail.com (G.C.); giuseppe.rovere02@icatt.it (G.R.); lorenzo.rocchi@policlinicogemelli.it (L.R.); giulio.maccauro@policlinicogemelli.it (G.M.); camillo.fulchignoni@gmail.com (C.F.)
[2] Section of Orthopaedics and Traumatology, Department of Clinical Science and Translational Medicine, University of Rome "Tor Vergata", 00133 Rome, Italy; farsetti@uniroma2.it
[3] Orthopaedics and Hand Surgery Unit, IRCSS Policlinico Gemelli, 00136 Rome, Italy
* Correspondence: silvia.pietramala01@gmail.com; Tel.: +39-3498910713

Abstract: Myoepithelioma is an exceptionally rare tumor, primarily arising in glandular tissues but occasionally found in soft tissues, including the hand. Its occurrence in the hand is particularly uncommon, presenting unique clinical challenges due to the limited number of documented cases and the unusual location. We conducted a literature review in June 2024, with the aim to evaluate the current understanding of hand myoepithelioma, recent diagnostic advances, treatment options, and the diverse presentations of this neoplasm. Articles confirmed that patients present with a painless, slow-growing mass in the hand, often misdiagnosed as more common soft tissue tumors like lipomas or fibromas. Imaging, particularly MRI and ultrasound, aids in assessing the tumor, but definitive diagnosis relies on histopathology, including immunophenotyping. Managing spindle cell myoepithelioma in the hand requires a multidisciplinary approach, with surgical excision being the primary treatment. Achieving clear margins is critical yet challenging due to the hand's complex anatomy. In some cases, adjuvant therapies such as radiation or chemotherapy may be necessary. The prognosis depends on factors like tumor size, location, and the success of surgical removal, with complete excision typically leading to a favorable outcome.

Keywords: myoepithelioma; hand; skin; tumor; soft tissue

Citation: Bocchino, G.; Capece, G.; Pietramala, S.; Rovere, G.; Rocchi, L.; Farsetti, P.; Maccauro, G.; Fulchignoni, C. Myoepithelioma of the Hand: A Systematic Review. *Appl. Sci.* **2024**, *14*, 9149. https://doi.org/10.3390/app14199149

Academic Editors: Francesco Cappello and Stefano Burgio

Received: 20 August 2024
Revised: 3 October 2024
Accepted: 4 October 2024
Published: 9 October 2024

1. Introduction

Myoepithelioma is a benign neoplasm that originates from the salivary glands, and it is composed entirely of myoepithelial cells. These cells can show different morphologies, such as spindle, epithelioid, plasmacytoid, or clear cells. Considering their glandular origin, myoepitheliomas share characteristics of both epithelial and smooth muscle cells [1]. Myoepithelial cells are found in various secretory organs, including the salivary glands [2,3]. The neoplastic behavior of salivary gland tumors, like myoepitheliomas, is unpredictable, and despite being classified as benign, malignant counterparts do exist. There is ongoing debate among researchers regarding the nature of myoepitheliomas. Some suggest that these tumors are a monomorphic, composed exclusively of myoepithelial cells [3,4]. Others describe a spectrum of salivary gland tumors, ranging from monomorphic adenomas to myoepitheliomas [5–7]. When myoepitheliomas arise in soft tissues, like those in the hand, they retain some characteristics of glandular tissue; however, their unusual location can sometimes lead to a misdiagnosis. This rare manifestation in the hand requires careful consideration in diagnosis and treatment, highlighting the importance of distinguishing these tumors from other soft tissue neoplasms. Understanding the complex behavior and characteristics of myoepithelial cells is crucial for the accurate diagnosis and effective management of myoepitheliomas in both typical and atypical locations. Clinicians might

find it hard to differentiate these tumors from more common soft tissue lesions, such as ganglions or lipomas, which may present similarly but require different management strategies. The final diagnosis relies on histologic examination but physicians might proceed with simple excision, thinking of a more obvious diagnosis. The aim of this study is to provide a comprehensive review of the clinical presentation, diagnostic approaches, management strategies, and prognosis of spindle cell myoepithelioma in the hand, drawing on the latest research and clinical case studies to offer valuable insights for practitioners dealing with this uncommon and challenging condition.

2. Materials and Methods

We conducted a systematic literature review according to PRISMA guidelines. A literature search was performed on MEDLINE through PubMed, Google Scholar, and Web of Knowledge. Searches were conducted in June 2024 using the terms "myoepithelioma AND hand OR finger." No restriction was applied on the date of publication to ensure comprehensive coverage. To avoid missing studies, no filters were applied to the search strategies.

Using titles and abstracts, three authors independently selected studies for inclusion.

Inclusion criteria: (1) level I-V evidence; (2) complete demographic data; (3) documentation of imaging used for diagnosis, histologic features, surgical techniques used and eventual further treatments applied; (4) documentation of surrounding tissue involvement; (5) and documentation of surgical or post-surgical complications and recurrence rate.

Exclusion criteria: (1) any language not known by the authors (English, Italian and French); (2) no full-text available; (3) reviews; (4) and insufficient documentation regarding the pathology. Any discordances were solved by consensus with a senior author (C.F.)

To avoid bias determined by articles with missing data, we excluded any article with incomplete data. All abstracts were reviewed to determine adherence to inclusion and exclusion criteria of our study. If no abstract was published or if the abstract did not have sufficient information to determine eligibility, the full-length manuscript was reviewed.

All the relevant data, including demographics, location, clinical presentation, diagnostics, type of surgery, recurrence rates, follow-up, and involvement of bone or surrounded tissues were collected. Primary aim of the study was to assess the state of the art regarding myoepithelioma located into the hand, the recent developments in diagnostics and treatment options, and the different presentations of this neoplasm.

3. Results

Fifty abstracts met our inclusion criteria. Among those, 35 articles were finally excluded, and 15 were reviewed (Figure 1).

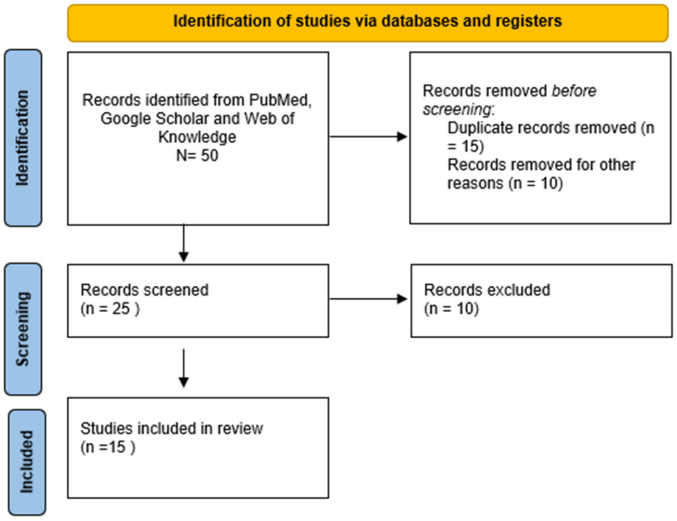

Figure 1. PRISMA flowchart of articles selection.

All the reviewed articles were case reports, each describing a single case of myoepithelioma in the hand or fingers (Table 1).

Table 1. Included articles and main characteristics.

Author	Year	Age	Gender	Localization	Clinical Presentation	Differential Diagnosis
Jo, V.Y. [8]	2013	52	M	3D	Firm, painless popular lesion	/
Clark, J.C. [9]	2009	36	M	TE	Firm mass attached to the FDP/CTS	Ganglion/PVNS
Fulchignoni, C. [10]	2024	12	F	2D	Violet, carnose lump	Dermoid Cyst
Pietramala, S. [11]	2023	15	M	3D	Firm mass adhered to the bone	Lipoma
Patton, A. [12]	2022	52	M	2D	Firm painless mass	Vascular tumor emboli (pleomorphic adenoma)
Ansari, M.T. [13]	2017	33	M	3D	Soft tissue swelling	Enchondroma
Nambirajan, A. [14]	2016	37	F	3D	Hard swelling	Enchondroma
Itintenag, T. [15]	2014	69	M	2D	Enlarging mass	GCT
Montalvo, N. [16]	2020	52	M	HE	Painless growing mass	/
Yeoh, D. [17]	2015	28	F	2D	Slowly enlarging tender mass	Chondromyxoid fibroma
Choi, Y.J. [18]	2011	51	M	HE	Painless growing mass	/
Zamora, C.A. [19]	2011	35	F	TE	Firm mass + CTS	Chondromyxoid fibroma
Chateau, F. [20]	2011	29	M	4D	Painful post-traumatic mass	/
Pilavaki, M. [21]	2006	50	F	HE	Slow growing mass	/
Hamada, K. [22]	2006	69	F	TE	Slow growing mass	Malignant neoplasm

D = digit, HE = hypothenar eminence, TE = thenar eminence, M = male, F = female, CTS = carpal tunnel syndrome, PVNS = pigmented villonodular synovitis, GCT = giant cells tumor.

The 15 cases comprised nine males (60%) and six females (40%), with an average age of 41.3 years (range 12–69 years). In nine cases [8–15,17,20], the lesion was located on a finger. Of these, five were on the third finger [8,11,13,14], with the second most affected being the second finger, followed by the fourth. No cases were described on the first or fifth fingers. For the five cases involving the hand [9,16,18,21,22], three were located in the hypothenar region [16,18,21] and two in the thenar area [9,22].

All patients, except one, reported no pain. The single patient who reported pain had a history of trauma to the same digit [20]. The mass was consistently described as firm and solid with no distinctive characteristics, except in one case where it was described as a carnose, violet-ish lump [10]. Two cases involved carpal tunnel syndrome [9,19].

We also included five cases of bony myoepithelioma as the clinical presentation and histological features suggest the same origin.

In four cases [8,9,16,18], no additional imaging examination was requested. X-ray was the first diagnostic exam requested in the other cases, complemented by MRI in six cases [11,14,15,17,21,22] and ultrasound (US) in three cases [10,11,19]. A CT scan was requested in two cases [11,21] to assess bone involvement. A PET scan was used in one case [22], showing a mass with malignant-like features.

The S-100 protein was always positive, followed by cytokeratin AE1/AE3 and EMA, which were negative in three cases only [14,17,21]. Other markers that tested positive included vimentin, calponin, and p63.

In one case, an excisional biopsy was performed before complete excision due to the suspicion of malignancy [17]. In all other cases, clear margin excision was performed. For the four cases involving bones [13,14,17,20], complete bone resection was performed in two cases, simple curettage in one case, and curettage followed by bone grafting in another. The average follow-up was 10.2 months, with one case [17] of recurrence observed, which resulted in ray amputation (Table 2).

Table 2. Demographic and diagnostic data.

N° of Patients	15
Gender	
Male	9 (60%)
Female	6 (40%)
Age	41.3 y.o. (12–69)
Location	
Third finger	5 (33.3%)
Second finger	3 (20%)
Fourth finger	1 (6.7%)
Thenar eminence	3 (20%)
Hypothenar eminence	3 (20%)
Imaging	
None	4
XRAY	6
MRI	1
CT	2
PET	3
US	3
Recurrence	
Yes	1 (6.7%)
No	14 (93.3%)

y.o. = years old; MRI = magnetic resonance imaging; CT = computer tomography; PET = positron emission tomography; US = ultrasound.

4. Discussion

Myoepitheliomas are infrequent occurrences, comprising merely 1–1.5% of all salivary gland tumors [23,24]. They were described in soft tissues in 1995 and, ever since, more and more cases have been described in the literature and, although very rare, they can also occur in bones [13,14,17,19,20]. Although these tumors commonly emerge around the age of 44 on average, instances have been documented across a broad spectrum ranging from as young as 9 to as old as 85 years [25]. As regards hands, we reported a mean age of 41.3 years old, which is comparable to what is described in the literature. While instances among pediatric populations are documented, they remain exceptionally rare [26]. Furthermore, we found two cases of spindle cell myoepithelioma in the fingers of a 15- and 12-year-old and so far; these are the only cases described in children [27]. Our research highlighted 15 cases of hand myoepithelioma—both on soft tissues and bones—but considering that data were not specified for location in some articles, we had to exclude them, so the number is surely higher [28]. Moreover, it has to be considered that since sometimes the clinical presentation suggest a common lesion such as fibromas, many cases could be missing as no histological examination is conducted. In fact, in the majority of cases discussed, the diagnosis of myoepithelioma is made after histological examination as different differential diagnosis are possible, as also reported in Table 1. The rarity of myoepitheliomas in this location necessitates careful consideration in differential diagnosis, often involving imaging studies such as MRI and histopathological examination for definitive diagnosis.

Cases of myoepitheliomas in the hand and wrist more often present as painless, slow-growing masses, mimicking more common soft tissue tumors. Despite that, as already said, sometimes the presentation can be extremely variable, leading the physician to other differential diagnosis such as enchondroma, lipoma, and even malignant tumors. Primary intra-osseous myoepithelioma are rare and occur predominantly in the axial and proximal appendicular skeleton in middle-aged patients [14].

Considering such variety of features, the focus of our discussion is to highlight the main and characteristic features of myoepithelioma as emerged from our review.

4.1. Clinical Features and Gross Findings

The clinical features of myoepitheliomas vary depending on their location. The size of the mass is variable, and while these tumors are typically painless, some patients may experience discomfort, especially if the tumor compresses nearby structures. The average size for benign tumors is 4 cm, whereas malignant myoepitheliomas have been reported to have an average size of at least 6 cm [28]. However, in our study, we did not collect data on the dimensions of hand masses, as the authors did not provide this information. Given the small anatomical area, the size of the mass should not be considered a definitive indicator of malignancy. Generally, these tumors present as slowly growing, painless masses [16], and patients often seek medical advice due to aesthetic or functional impairments, such as when the mass is located between two fingers or near a joint. We described two cases [9,19] of myoepitheliomas associated with carpal tunnel syndrome due to nerve compression, which is rare but should be considered when evaluating a patient with a mass in the thenar region, as it could be misdiagnosed as a nerve tumor.

On the other hand, when occurring in bones, these tumors can be completely painless and emerge following a simple X-ray or associated with local soft tissue swelling and pain.

4.2. Microscopic Findings

Microscopically, myoepithelial tumors of soft tissue have a lobulated appearance. Most cases are well-circumscribed, but in certain cases they can infiltrate local soft tissues. Pietramala et al. [11] described a case in which the main mass partially infiltrated the collateral radial nerve of the third finger and the flexor tendon pulley (Figure 2).

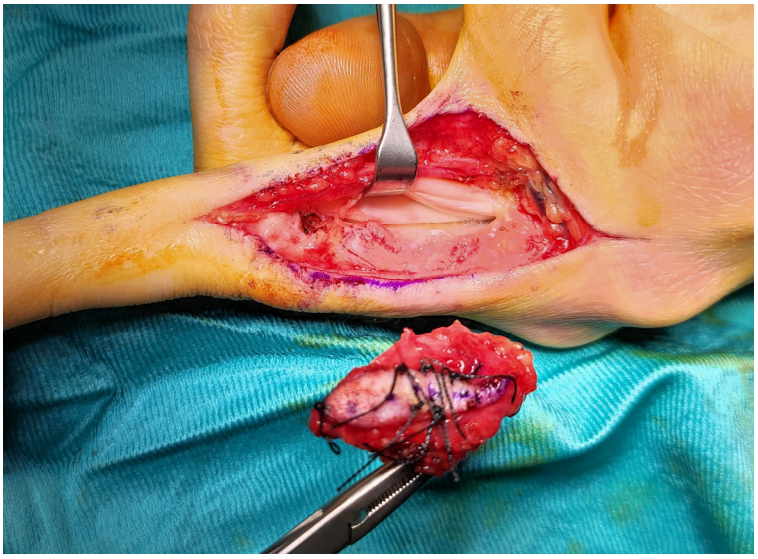

Figure 2. Tumor excision showing macroscopical features. The lesion has been excised including the overlying skin and marked with stiches for histologic analysis. The tendons are retraced and the underneath bone is visible [11].

In cases like this, despite the benign nature of the tumor, its excision could cause some degree of impairment. One of the main characteristics of myoepithelial tumors is their variety, which refers to architectural, cytological, and stromal features. Cells can show trabecular or retinacular arrangement patterns. Neoplastic myoepithelial cells can display different morphological appearances, such as epithelioid, clear cell, plasmacytoid, and spindle cell phenotypes [29].

Necrosis can be found, but it should be investigated for malignancy if highly represented. When occurring in bones, the mass shows a chondroid-like aspect with a chondromyxoid stroma-rich tumor composed of polygonal or spindle cells arranged in cords without significant cellular atypia [14,17].

4.3. Immunohistochemistry

Histologic examination is crucial to make our diagnosis. Different cell morphologies within myoepitheliomas show varying immunoreactivity to smooth muscle markers. Spindle cells exhibit the strongest immunoreactivity, followed by epithelioid cells, with plasmacytoid and clear cells showing less or no activity [30].

Myoepithelioma cells typically express cytokeratin (AE1/AE3, CK 5/6, Cam 5.2, CK-7 and CK-14) and vimentin (positive in neoplastic myoepithelial cells but negative in normal myoepithelial cells).

Neoplastic transformation can modify the typical smooth muscle phenotype of myoepithelial cells, showing positivity for the following markers [31]: S-100 (positive in neoplastic myoepithelial cells but not in normal myoepithelial cells), calponin (the most sensitive myogenic marker [31,32], smooth muscle actin (SMA), muscle-specific actin (MSA), smooth muscle myosin, P63 protein, glial fibrillary acidic protein (GFAP), and SOX10 (expressed in 80% of myoepitheliomas but are only a third of myoepithelial carcinomas [33]. E-cadherin is also expressed in myoepitheliomas [34]. Myoepithelial cells are typically negative for carcinoembryonic antigen (CEA), indicating no tubular differentiation [34].

As myoepitheliomas can show focal areas similar to parachordomas, it is becoming a common idea that parachordoma should be considered in the same spectrum of myoepithelioma of soft tissue, as also described in the new WHO classification [1]. One element that differentiates myoepitheliomas from parachordomas is that the latter is GFAP- and SMA-negative. Nevertheless, since very few cases of parachordomas have been described, we cannot consider this distinction.

From our review, it emerges that the consistently positive immunohistochemical markers in myoepitheliomas include AE1/AE3, S-100, vimentin, calponin, and EMA. These markers are essential for identifying the neoplasm and may vary depending on the cell morphology present.

Histologic examination is the primary method to assess malignancy.

Malignancy should be suspected in presence of an infiltrative growth pattern. In a study of 101 myoepithelial tumors of soft tissue [28], the only parameter related to recurrence and metastasis was the presence of at least moderate cytological atypia, defined as nuclear pleomorphism, vesicular or coarse chromatin, and/or prominent nucleoli. It must be pointed out that, when occurring in pediatric population, myoepithelial tumors are more prone to be malignant. When exclusively talking about cutaneous myoepithelioma, the main features of malignancy are represented by cytological atypia and high mitotic rate and the presence of infiltrative borders, but occasional cases showed no histological features of malignancy [35].

When talking about mitotic rate, there is no cutoff although Nagao et al. [32] reported that finding >7 mitoses per 10 HPF was diagnostic of malignancy. Despite being quite reliable, cytologic atypia is not pathognomonic of malignancy as some low-grade myoepithelial carcinomas behave aggressively [29], and the single most useful criterion remains the invasive growth pattern. Nevertheless, this is not reliable in soft tissues, as nearly half of the cases described by Hornick et al. [28] showed microscopically infiltrative margins, and this feature was associated with neither recurrence nor metastasis. Although experience is limited, the presence of at least moderate cytologic atypia should be sufficient to classify the neoplasm as myoepithelial carcinoma with risk for aggressive behavior and propensity for metastasis.

Regarding cytogenetic and genomic findings, genomic fusions are prominent features. The most common genomic fusions are ESWR1-POU5F1 and ESWR1-PBX3. Chromosomal translocations and complex chromosomal aberrations are frequent, but specific patterns may vary from tumor to tumor. Chromosomal aberrations are especially frequent in the case of malignant behavior [12,28].

4.4. Diagnosis

Accurately diagnosing myoepithelioma in the hand and wrist region involves a combination of imaging techniques and histopathological analysis.

Initially, radiographs are employed to rule out any bone involvement, providing an initial assessment of adjacent skeletal structures and confirming if the tumor interacts with bone. As described in our results, four authors [13,14,17,20] described a bony myoepithelioma and one author described bony erosion [Figure 3] [11].

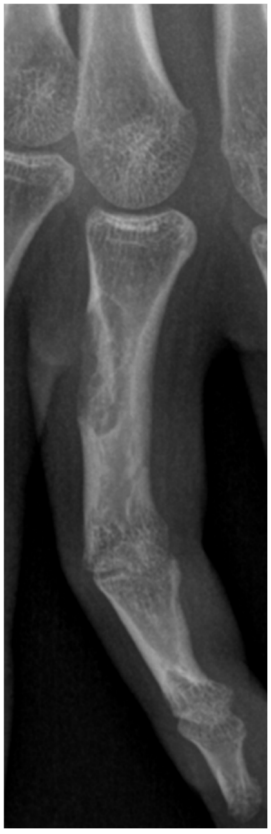

Figure 3. X-ray of a 15-year-old patient with a firm mass on his third finger, initially identified as a lipoma. Here, it is the X-ray showing bone erosion of the proximal phalanx [11].

Subsequently, ultrasound becomes invaluable for evaluating the soft tissue characteristics of the mass. This technique allows for a detailed examination of superficial structures, aiding in assessing the size, shape, and internal features of the tumor in the hand and wrist [19]. Despite this, only three authors used ultrasound imaging, underlying how the unknown nature of the mass might let the physician ask for a more costly and invasive imaging such as the MRI.

MRI typically reveals a well-defined mass with a heterogeneous enhancement pattern, providing crucial information for treatment planning and the accurate assessment of the tumor extent in the hand and wrist [35].

Positron emission tomography (PET) scans have been mentioned in some articles regarding spindle cell myoepithelioma diagnosis; however, their utility may be limited. PET scans could lead to misinterpretation [22], suggesting malignancy when it may not be present, and therefore might be discouraged in favor of more reliable imaging techniques [22].

For a definitive diagnosis, histopathological analysis following biopsy remains essential.

This can be achieved through core needle biopsy or excisional biopsy to obtain tissue samples necessary for detailed microscopic evaluation. All authors except for Yeoh, D. et al. [17] described the excision of the mass without previous needle biopsy.

In one case [11], the previously planned excision was converted into incisional biopsy due to the odd macroscopic aspect during surgery.

The differential diagnosis includes many entities such as epithelioid schwannoma, epithelioid sarcoma, and, rarely, benign and malignant ossifying fibromyxoid tumors. Specifically, in the hands, other diagnoses should be ruled out. As emerged from our review, before reaching the final diagnosis, authors suspected ganglion, PVNS [8], lipoma [11], dermoid cyst [10], and GCT [15]. The ultrasound and MRI features of these neoplasms are almost indistinguishable from those of myoepithelioma. Given the high recurrence rate of some entities, such as GCT [36] and PVNS, accurately diagnosing these conditions is crucial for determining the prognosis (Table 3).

Table 3. Differential diagnosis.

Myoepithelioma Differential Diagnosis			
	Clinical Features	**Histologic Features**	**Imaging**
Myoepithelioma	Slow growing	S100 protein, EMA, CK7, AE1/AE3, calponin	X-ray: may show bone erosion MRI: low contrast T1, high contrast T2. May show tendon involvement
Schwannoma	Alongside nerve, +/− sensitive or motor impairments	SOX10, S100, NF, GFAP	MRI: isointense to intermediate signals on T1WI, heterogeneously hyperintense signals on T2WI, and STIR with no suppression on fat saturated sequences
Lipoma	Slippage sign	S-100, CD34, Leptin	MRI: well-defined margins, hyperintense in T1WI and T2WI
Synovial Sarcoma	Slow growing single or lobulated	CK-pan, EMA, vimentin, BCL-2	MRI: infiltrative margins, isointense to muscle belly in T1WI and hyperintense in T2WI. Typical "triple signal"
Myoepithelial carcinoma	Slow growing firm mass	CK-14, CK-7, CK-pan, EMA, S100, calponin, HMW-CK, p63	MRI: ill-defined margins, isointense in T1, hyperintense in T2

EMA = epithelial membrane antigen; CK7 = cytokeratin 7; AE = cytokeratin antibody; NF = neurofilament; GFAP = glial fibrillary acidic protein; SOX-10 = SRY-box transcription factor 10; HMW-CK = high-molecular-weight cytokeratin; MRI = magnetic resonance imaging; WI = weighed images.

Hamada, K. et al. [22] reported a mass located in the thenar eminence that showed high F-FDG accumulation, leading to the initial suspicion of a malignant lesion. In cases of bony lesions, the first suspicion is usually enchondroma or chondromyxoid fibroma. Finger bony myoepithelioma typically presents as a well-defined, expansive, geographic lytic lesion, which usually does not raise suspicion of malignancy. The first diagnosis can also occur after trauma, as is sometimes seen with enchondromas [20].

4.5. Management

Surgical excision is the cornerstone in treating myoepithelioma of the hand, aiming for complete removal to prevent recurrence, which accounts for almost 30% if the mass is not excised completely [28,36]. This typically involves a wide local excision procedure, meticulously ensuring clear margins around the tumor. Given the hand's intricate anatomy and functional importance, achieving these negative margins can be particularly challenging but is crucial for long-term outcomes.

Reconstructive surgery may follow depending on the tumor's size and location. This step not only focuses on removing the tumor but also aims to restore optimal function and the appearance of the affected hand. Techniques such as skin grafts or flap reconstruction may be employed to achieve these goals effectively.

When spindle cell myoepithelioma involves bone, complete excision of the affected bone segment may be necessary, followed by reconstructive procedures, such as bone

grafting or flap reconstruction (e.g., toe-to-hand transfer) to restore function and aesthetics. This approach is crucial in maintaining the structural integrity of the hand while ensuring effective treatment [13]

In cases where surgical margins are positive or in recurrent disease, adjuvant therapies are considered to manage residual disease or reduce the risk of recurrence. Radiotherapy is often utilized for its ability to target remaining cancerous cells [37], while chemotherapy remains reserved for cases demonstrating malignant behavior or metastatic potential [36].

A novel approach gaining attention involves a multidisciplinary strategy combining surgical excision with custom brachytherapy. This approach integrates precise radiation therapy directly into the surgical site, offering localized treatment while sparing healthy tissues. Such innovations are pivotal in enhancing treatment efficacy and minimizing potential side effects, thereby optimizing patient outcomes [11].

Adjuvant therapies, such as radiotherapy or chemotherapy, may be indicated in cases with positive margins or higher-risk features. Long-term follow-up is essential to monitor for any signs of recurrence or potential malignant transformation, allowing for timely intervention if necessary.

4.6. Prognosis and Predictive Factors

According to Sciubba and Brannon [38], myoepitheliomas have lower recurrence rates post-complete surgical resection compared to pleomorphic adenomas, with only one recurrence in sixteen cases observed over 1 month to 7 years [4]. Nayak et al. found that 93% of myoepithelial tumors followed a benign course, while 7% exhibited malignant behavior [38]. We found only one recurrence described [17], accounting for 13.3%.

The progression of cutaneous myoepithelioma does not always align with its histological features. While most metastatic myoepitheliomas in the skin and soft tissue exhibit cellular abnormalities, a high rate of cell division, and invasive growth, there are a few cases that behave malignantly despite lacking these features [39]. A high rate of cell division is often linked to the risk of recurrence or spread in tumors with otherwise benign characteristics [40]. Cellular atypia is the primary indicator of malignancy and the best predictor of aggressive behavior. However, even without atypia, factors such as a high mitotic rate, dense chromatin, prominent nucleoli, nuclear pleomorphism, and necrosis can indicate an increased risk of malignancy [41]. The recurrence rate for cutaneous myoepitheliomas is about 20%, but the prognosis is generally favorable even after recurrence [42]. Therefore, the recommended treatment is complete surgical removal of the lesion with clear margins [28].

Spindle cell myoepithelioma of the hand generally shows a favorable prognosis following complete surgical excision; low recurrence rates are typical with adequate surgical margins, necessitating close follow-up to monitor for any signs of recurrence [16]. Although rare, there is a potential for malignant transformation, emphasizing the importance of long-term follow-up. Patients typically achieve good functional recovery and quality of life post-treatment; most patients regain normal hand function with appropriate surgical and rehabilitative care. This highlights the importance of comprehensive management and ongoing surveillance to optimize outcomes for patients with myoepitheliomas in the hand and wrist.

Additionally, the rarity of myoepitheliomas in the hand adds complexity to their diagnosis and management, making it crucial for clinicians to be well-versed in their clinical and histological features [43]. Early and accurate diagnosis, coupled with a clear understanding of the tumor's behavior, is essential for ensuring successful treatment outcomes. Moreover, multidisciplinary care involving surgeons, pathologists, and radiologists can significantly improve the management and prognosis of these patients. Regular follow-ups are necessary to detect any early signs of recurrence, and patients should be counseled on the importance of monitoring and reporting new symptoms. Overall, a proactive approach in the diagnosis, treatment, and follow-up care of myoepitheliomas in the hand can greatly enhance patient outcomes and reduce the risk of recurrence or complications.

5. Conclusions

Myoepithelioma of the hand presents as a rare and diagnostically challenging condition, necessitating a thorough clinical approach. Accurate diagnosis relies on comprehensive imaging modalities and detailed histopathological assessment to distinguish it from other hand tumors. Despite this, we believe that being aware of the most typical clinical presentations and radiological findings can help physicians to obtain the right diagnosis even before histological examination

In terms of treatment, a multidisciplinary approach is crucial for achieving optimal outcomes. Continued research is pivotal to further elucidate the behavior and optimal management strategies of myoepitheliomas in uncommon locations like the hand. This ongoing exploration will enhance our ability to refine treatment approaches and improve outcomes for affected patients.

Author Contributions: Conceptualization: G.B. and G.C.; data collection S.P.; writing—review and editing, G.B. and G.C.; visualization, G.R. and C.F. supervision, P.F., L.R. and G.M. All authors have read and agreed to the published version of the manuscript.

Funding: This research received no external funding.

Conflicts of Interest: The authors declare no conflicts of interest.

References

1. Seifert, G.; Sobin, L.H. Myoepithelioma. In *World Health Organization International Histological Classification of Tumours: Histological*.
2. Erlandson, R.A.; Cardon-Cardo, C. Histogenesis of benign pleomorphic adenoma (mixed tumor) of the major salivary glands. An ultrastructural and immunohistochemical study. *Am. J. Surg. Pathol.* **1984**, *8*, 803–820. [CrossRef] [PubMed]
3. Batsakis, J.G. Myoepithelioma. *Ann. Otol. Rhinol. Laryngol.* **1985**, *94*, 523–524. [CrossRef] [PubMed]
4. Sciubba, J.J.; Brannon, R.B. Myoepithelioma of salivary glands: Report of 23 cases. *Cancer* **1982**, *49*, 562–572. [CrossRef]
5. Sciubba, J.J.; Foldstein, B.H. Myoepithelioma. Review of the literature and report of a case with ultrastructural confirmation. *Oral Surg. Oral Med. Oral Pathol.* **1976**, *42*, 328–338. [CrossRef] [PubMed]
6. Miliauskas, J.R.; Orell, S.R. Fine-needle aspiration cytological findings in five cases of epithelial-myoepithelial carcinoma of salivary glands. *Diagn. Cytopathol.* **2003**, *28*, 163–167. [CrossRef]
7. Matsuyama, A.; Hisaoka, M. PLAG1 expression in mesenchymal tumors: An immunohistochemical study with special emphasis on the pathogenetical distinction between soft tissue myoepithelioma and pleomorphic adenoma of the salivary gland. *Pathol. Int.* **2012**, *62*, 1–7. [CrossRef]
8. Jo, V.Y.; Antonescu, C.R. Cutaneous syncytial myoepithelioma: Clinicopathologic characterization in a series of 38 cases. *Am. J. Surg. Pathol.* **2013**, *37*, 710–718. [CrossRef]
9. Clark, J.C.; Galloway, S.J. Myoepithelioma within the carpal tunnel: A case report and review of the literature. *Int. Semin. Surg. Oncol.* **2009**, *6*, 15. [CrossRef]
10. Fulchignoni, C.; Pietramala, S. Spindle-cell myoepithelioma, a rare neoplasm with various clinical presentations that can affect fingers. *Hand Surg. Rehabil.* **2024**, *43*, 101668. [CrossRef]
11. Pietramala, S.; Tulli, A. Spindle-cell myoepithelioma: A rare case in a finger of a 15-year-old boy. *Hand Surg. Rehabil.* **2023**, *42*, 549–552. [CrossRef]
12. Patton, A.; Speeckaert, A. A novel IRF2BP2::CDX2 Gene fusion in digital intravascular myoepithelioma of soft tissue: An enigma! *Genes. Chromosomes Cancer* **2023**, *62*, 176–183. [CrossRef] [PubMed]
13. Ansari, M.T.; Kotwal, P.P. Intraosseous myoepithelioma: A rare tumour in the hand. *J. Hand Surg. Eur.* **2017**, *42*, 530–531. [CrossRef] [PubMed]
14. Nambirajan, A.; Mridha, A.R. Primary intra-osseous myoepithelioma of phalanx mimicking an enchondroma. *Skelet. Radiol.* **2016**, *45*, 1453–1458. [CrossRef]
15. Itinteang, T.; Morine, A.E. Finger soft tissue myoepithelioma: A rare entity. *ANZ J. Surg.* **2015**, *85*, 883–884. [CrossRef]
16. Montalvo, N.; Redrobán, L. Cutaneous myoepithelioma: An unusual tumor in the hand. *Case Rep. Pathol.* **2020**, *2020*, 3747013. [CrossRef]
17. Yeoh, D.; Symmans, P. Bony myoepithelioma: A rare condition presenting in the hand. *J. Hand Surg. Eur. Vol.* **2017**, *42*, 94–95. [CrossRef]
18. Choi, Y.-J. Cutaneous myoepithelioma on the palm. *J. Dermatol.* **2011**, *38*, 1210–1213. [CrossRef]
19. Zamora, C.A.; Zamora, M.A. Myoepithelioma of the hand and carpal tunnel: An unusual cause of median nerve compression. *J. Clin. Ultrasound* **2011**, *39*, 44–47. [CrossRef] [PubMed]
20. Chateau, F.; Nguyen, P. Tumeur myoépithéliale des tissus mous avec extension intraosseuse dans la première phalange de l'annulaire: À propos d'un cas Clinique. *Chir. Main.* **2011**, *30*, 144–147. [CrossRef]

21. Pilavaki, M.; Givissis, P. Soft-tissue myoepithelioma of the hypothenar region: A case report. *J. Hand Surg. Am.* **2007**, *32*, 674–676. [CrossRef]
22. Hamada, K.; Ueda, T. Myoepithelioma of soft tissue originating from the hand: 18F-FDG PET features. *AJR Am. J. Roentgenol.* **2006**, *186*, 270–271. [CrossRef] [PubMed]
23. Dardick, I.; Thomas, M.J. Myoepithelioma--new concepts of histology and classification: A light and electron microscopic study. *Ultrastruct. Pathol.* **1989**, *13*, 187–224. [CrossRef]
24. Kawashima, Y.; Kobayashi, D. A case of myoepithelioma arising in an accessory parotid gland. *J. Laryngol. Otol.* **2002**, *116*, 474–476. [CrossRef] [PubMed]
25. Dardick, I.; van Nostrand, A.W. Myoepithelial cells in salivary gland tumors–revisited. *Head. Neck Surg.* **1985**, *7*, 395–408. [CrossRef]
26. Lins, J.E.; Gnepp, D.R. Myoepithelioma of the palate in a child. *Int. J. Pediatr. Otorhinolaryngol.* **1986**, *11*, 5–13. [CrossRef] [PubMed]
27. Mahdi, Y.; Zouaidia, F. Combined myoepithelial carcinoma and myoepithelioma in soft tissue: A case report and review of the literature. *J. Med. Case Rep.* **2014**, *8*, 317. [CrossRef]
28. Hornick, J.L.; Fletcher, C.D. Myoepithelial tumors of soft tissue: A clinicopathologic and immunohistochemical study of 101 cases with evaluation of prognostic parameters. *Am. J. Surg. Pathol.* **2003**, *27*, 1183–1196. [CrossRef]
29. Savera, A.T.; Zarbo, R.J. Defining the role of myoepithelium in salivary gland neoplasia. *Adv. Anat. Pathol.* **2004**, *11*, 69–85. [CrossRef] [PubMed]
30. Cardesa, A.; Alos, L. Myoepithelioma. In *World Health Organization Classification of Tumours: Pathology and Genetics of Head and Neck Tumours*; Barnes, L., Eveson, J.W., Eds.; IARC Press: Lyon, France, 2005; pp. 259–260.
31. Savera, A.T.; Gown, A.M. Immunolocalization of three novel smooth muscle-specific proteins in salivary gland pleomorphic adenoma: Assessment of the morphogenetic role of myoepithelium. *Mod. Pathol.* **1997**, *10*, 1093–1100.
32. Sugano, N.T. Salivary gland malignant myoepithelioma: A clinicopathologic and immunohistochemical study of ten cases. *Cancer* **1998**, *83*, 1292–1299.
33. Miettinen, M.; McCue, P.A. Sox10—A marker for not only schwannian and melanocytic neoplasms but also myoepithelial cell tumors of soft tissue: A systematic analysis of 5134 tumors. *Am. J. Surg. Pathol.* **2015**, *39*, 826–835. [CrossRef] [PubMed]
34. Prabhu, S.; Kaveri, H. Benign; malignant salivary gland tumors: Comparison of immunohistochemical expression of e-cadherin. *Oral Oncol.* **2009**, *45*, 594–599. [CrossRef]
35. Barnett, M.D.; Wallack, M.K. Recurrent malignant chondroid syringoma of the foot: A case report and review of the literature. *Am. J. Clin. Oncol.* **2000**, *23*, 227–232. [CrossRef]
36. Al-Qattan, M.M. Giant cell tumours of tendon sheath: Classification and recurrence rate. *J. Hand Surg. Br.* **2001**, *26*, 72–75. [CrossRef] [PubMed]
37. Shimosawa, H.; Susa, M. Soft tissue mixed tumor of the hand. *Rare Tumors* **2009**, *1*, e30. [CrossRef] [PubMed]
38. Jo, V.Y.; Fletcher, C.D. Myoepithelial neoplasms of soft tissue: An updated review of the clinicopathologic, immunophenotypic, and genetic features. *Head. Neck Pathol.* **2015**, *9*, 32–38. [CrossRef]
39. Kabarriti, R.; Quinn, T.J. Neoadjuvant radiation therapy for the management of myoepithelial carcinoma of the upper extremity. *Int. J. Cancer* **2018**, *142*, 854–862. [CrossRef]
40. Nayak, J.V.; Molina, J.T. Myoepithelial neoplasia of the submandibular gland: Case report and therapeutic considerations. *Arch. Otolaryngol. Head. Neck Surg.* **2003**, *129*, 359–362. [CrossRef]
41. Miller, T.D.; McCalmont, T. Recurrent cutaneous myoepithelioma treated using Mohs micrographic surgery: Case report and review of the literature. *Dermatol. Surg.* **2009**, *35*, 139–143. [CrossRef]
42. Hornick, J.L.; Fletcher CD, M. Cutaneous myoepithelioma: A clinicopathologic and immunohistochemical study of 14 cases. *Hum. Pathol.* **2004**, *35*, 14–24. [CrossRef]
43. Flucke, U.; Palmedo, G. _EWSR1_ gene rearrangement occurs in a subset of cutaneous myoepithelial tumors: A study of 18 cases. *Mod. Pathol.* **2011**, *24*, 1444–1450. [CrossRef] [PubMed]