IJIRR

International Journal of Information Research and Review
Vol. 10, Issue, 12, pp.7637-7639, December, 2023

RESEARCH ARTICLE

MYOEPITHELIAL TUMOR OF THE SOFT TISSUES OF THE THIGH: A CASE REPORT

***Ousama JELTI, Oussama El ALAOUI, Adnane LACHKAR, Abdeljaouad NAJIB and HichamYACOUBI**

Faculty of Medicine of Oujda, University Hospital Center, Department of Orthopedic Surgery B

ARTICLE INFO

Article History:
Received 16th September, 2023
Received in revised form
19th October, 2023
Accepted 26th November, 2023
Published online 15th December, 2023

Keywords:

Myoepithelioma, Soft Tissue, Thigh.Surgery,

ABSTRACT

Myoepithelial soft tissue tumors often pose diagnostic challenges due to their rarity. Their identification relies primarily on pathological analysis, although clinical and radiological features can guide the healthcare professional. Myoepithelial tumors, when benign, generallydo not invade bone. In this context, we present a new case of this rare pathology, observed inthe soft tissues of the thigh of a 32-year-old individual, with no bone involvement detected. Surgery was completely successful, and after one year, the patient showed a favorable evolution, with no signs of local recurrence. It should be noted that myoepitheliomas mainly affect the extremities. When they are deep-seated, it is essential to distinguish them from extraskeletal myxoid chondrosarcomas, parachordomas and synovialosarcomas. The majority of myoepitheliomas are benign in nature, although metastasis is a possibility.

INTRODUCTION

Myoepithelial tumors of the soft tissues occur at all ages, reaching a peak in incidence between the third and fifth decades, with a slight predominance in men. Around a third of cases reported in the literature involve the lower limb (1). Of these tumors, 60% are subcutaneous and 40% are deep-seated (intramuscular and subfascial). The risk of local recurrence is estimated at around 20%. A series of 101 myoepithelial soft-tissue tumors was published by Hornick and Fletcher, of which only 11 cases were localized to the thigh (2). The cases documented in the literature systematically describe the discovery, during medical consultations, of masses with classic features, progressive growth, indolence and absence of bone invasion.

Patient and observation

A young man aged 32, a former chronic smoker with an average consumption of 5 packs per year, who had quit 3 years ago, presented with a bulky soft-tissue enlargement in the right thigh for 9 months. This mass progressively increased in size, without the presence of other systemic symptoms such as fever, chills, night sweats, or unexplained weight loss.

Corresponding author: *Ousama JELTI,
Faculty of Medicine of Oujda, University Hospital Center, Department of Orthopedic Surgery B.

He had no personal or family history of cancer, and no previous surgeries. Physical examination revealed a round, soft subcutaneous mass in the lower third of the inner aspect of the right thigh, measuring 10 cm in length, well delimited, of soft consistency and not painful to the touch. The mass was mobile in relation to the deep and superficial planes. The adjacent skin was supple, without adherence to the mass, and showed no telangiectasia or ulceration, with joint amplitudes maintained. Magnetic resonance imaging (MRI) of the right thigh revealed a roughly oval mass within theright gracilis muscle. This mass was well circumscribed, presenting punctiform areas in hyposignal and showing marked intensification after gadolinium injection. Notably, the lesionremained clearly delimited by the gracilis muscle fascia, with no extension to other muscle compartments, and no evidence of bony abnormality (Figure 1). A biopsy of the mass was performed, suggesting the possibility of two diagnoses: either alow-grade fibromyxoid sarcoma, or a myoepithelial soft-tissue tumor (Figure 2). The mass was completely removed (Figure 3) and (Figure 4). Histopathological analysis confirmed the diagnosis of soft-tissue myoepithelial tumor. Excision was complete, with healthy margins, and post-operative recovery was straightforward. Following discussions in the medical team, it was recommended that the patient be monitored every six months by nuclear magnetic resonance imaging (MRI) and clinical examination, with no need for additional chemotherapy or radiotherapy. Six-month and one-year follow-ups revealed no local recurrence or metastatic lesions.

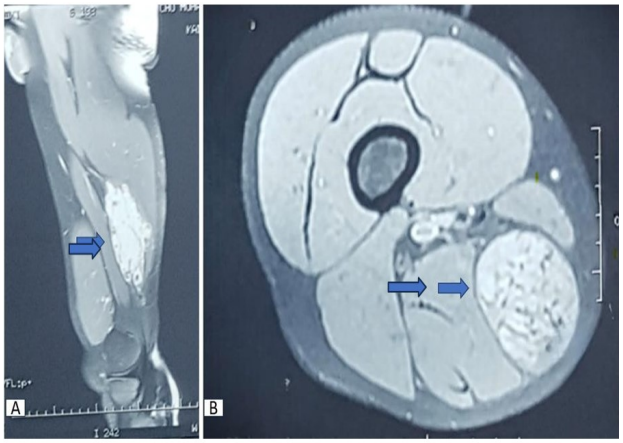

Figure 1. Magnetic resonance imaging scan (A): sagittal image showing a relatively well-defined oval-shaped tumour in the posteromedial side of the right thigh (B): coronal image also showing the mass in the subcutaneous fat layer

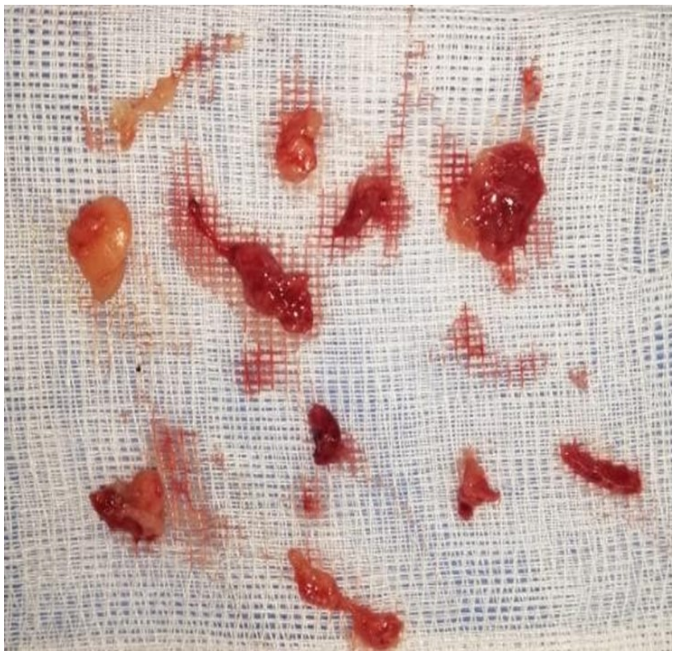

Figure 2. Intraoperative image of the biopsy taken for anatomopathologicalstudy

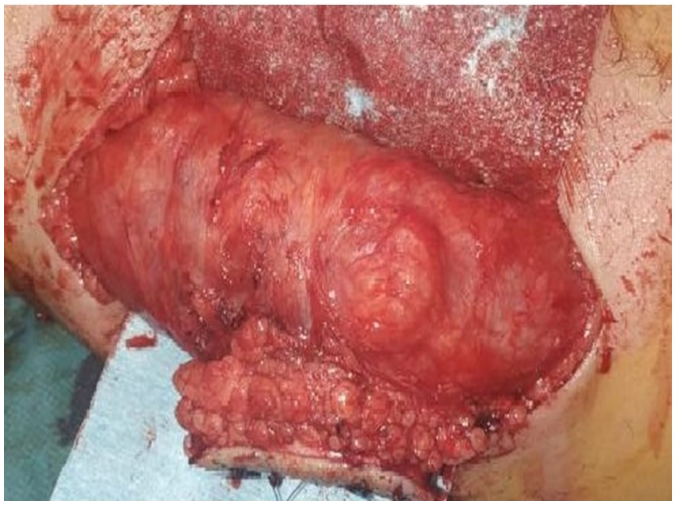

Figure 3. Intraoperative view of tumor excision

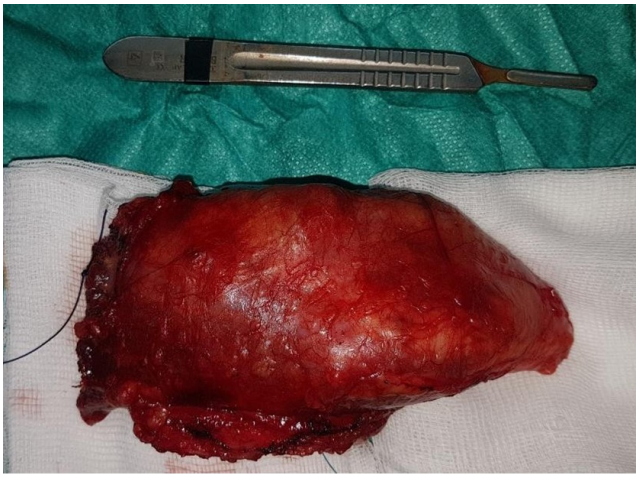

Figure 4. Intraoperative view of tumor excision

DISCUSSION

Soft tissue myoepitheliomas remain uncommon, but their characterization has improved considerably over the last decade (3). The very first report of this pathology was published in 1997 (4). Published case series indicate a distribution over a wide age range, with peak incidence between the third and fifth decades, and a slight male predominance (1). The majority of reported cases, nearly two-thirds, occur in the extremities (38% in the lower limbsand 27% in the upper limbs), with only a few cases localized to the thigh. Other sites include the head and neck region (16%), the trunk (13%), and visceral soft tissues (6%) (2). Average tumor size ranged from 1 to 7 cm (1). Observations have also revealed a correlation between bone involvement and a malignant form of the tumour. Interestingly, soft tissue myoepithelial tumors in children tend to be more malignant and aggressive than those observed in adults (5).

The diagnosis of myoepithelioma is based on the combined evaluation of morphological features and supportive immunohistochemical findings. Microscopically, the structure of this tumor is composed of lobular, multinodular, reticular, solid or mixed patterns, with or without very limited epithelial differentiation. Neoplastic myoepithelial cells may present a diversity of morphological features, including spindle-shaped, ovoid, plasmacytoid, epithelioid or clearcells. The composition of the stroma is also variable, and may present as hyalinized, myxoid, or with various forms of mesenchymal metaplasia (2-6-7-8). Once the diagnosis of myoepithelioma has been considered, confirmatory immunohistochemical analyses may prove useful. Tumor cells generally show positivity for epithelial markers such as cytokeratins, epithelial membrane antigen, and S100 protein, with variable expression of GFAP, P63, and myogenic markers such as smooth muscle actin, desmin, and h-caldesmon (2-10). On the molecular level, the detection of a reorganization of the EWSR1 gene, which can be easily identified by FISH analysis, can be a powerful diagnostic tool in complex cases (9). As far as imaging tests are concerned, the authors consider that radiological diagnosis relies mainly on MRI, which generally reveals (11), in T2 weighting, a globally increased and heterogeneous signal, with hyposignal in the lobular septa. The authors wish to review the histological diagnosis and assess severity, aware of the potential for concern for both patient and practitioner. Soft-

tissue myoepitheliomas generally exhibit benign behaviour, albeit with a relatively low and unpredictable risk of local recurrence (11). However, the presence of features such as necrosis and even moderate atypia on histology necessitates classifying these lesions as myoepithelial carcinomas (12). It is crucial to distinguish these malignant myoepithelial soft tissue tumours from myoepitheliomas present in the salivary glands and mammary gland (13), as they can be potentially fatal (14), although chemotherapy has been shown to be effective (15). Studies indicate that lesions smaller than 4 cm tend to be rather benign, in contrast to lesions largerthan 6 cm (12-13-14). Thus, in our case, the lesion was considered to have a low degree of malignancy. The criteria for malignancy in soft-tissue myoepithelioma are still a matter of debate; the mostimportant histological feature for predicting potentially aggressive behavior remains the presence of cytological atypia (6-10). Other clinicopathological parameters such as patient age, status of excisional margins, tumour depth, tumour size, infiltration into surrounding

tissues, presence of tumour necrosis, and mitosis rate do not correlate statistically significantly with recurrence or metastasis (7-10). In most situations, soft tissue myoepithelial neoplasms can be effectively treated by simple complete excision. Nevertheless, their prognosis remains ambiguous, although the majority appear to present with benign behavior. A case series by Michal and Miettinen (16) reported two patients with local recurrences and one death attributable to multiple pulmonary metastases. Notably, the recurrent tumors showed histological similarity to the original neoplasms. In our case, after 12 months of follow-up, no indication of tumor recurrence or metastasis was observed. It is plausible that the optimal approach, similar to that for salivary gland myoepitheliomas, is extensive local excision with healthy resection margins. In addition, various differential diagnoses, such as extraskeletal myxoid chondrosarcomas, parachordomas and synovialosarcomas, need to be considered. However, pathological examination confirmed the nature of the case as a soft tissue myoepithelioma.

CONCLUSION

We encourage authors and ourselves to document cases of limb myoepitheliomas. The aim isto expand the references available on this complexly diagnosed tumour, and to facilitate, through patient follow-up, a better understanding of its local evolution, as well as the documentation of any recurrences.

Conflict of interest: The authors declare that they have no conflict of interest.

REFERENCES

1. Rastrelli M, Del Fiore P, Damiani GB, Mocellin S, Tropea S, Spina R, Costa A, CavallinF, Rossi CR. Myoepithelioma of the soft tissue: A systematic review of clinical reports. Eur JSurg Oncol. 2019 sept; 45 (9): 1520-1526.
2. Hornick JL, Fletcher CD. Myoepithelial tumors of soft tissue: a clinicopathologic and immunohistochemical study of 101 cases with evaluation of prognostic parameters. Am J Surg Pathol 2003;27:1183e96.
3. Jo VY and Fletcher CD: Myoepithelial neoplasms of soft tissue:an updated review of the clinicopathologic, immunophenotypic, and genetic features. Head Neck Pathol (2015) 1:32- 38.
4. Kilpatrick SE, Hitchcock MG, Kraus MD, Calonje E and Fletcher CD:Mixed tumors and myoepitheliomas of soft tissue: a clinicopatho-logic study of 19cases with a unifying concept.Am J Surg Pathol (1997) 1:13-22.
5. Gleason BC, Fletcher CD. Myoepithelial carcinoma of soft tissue in children: An aggressive neoplasm analyzed in a series of 29 cases. Am J Surg Pathol 2007;31:1813-24.
6. Park S, Kim A, Gu M, Choi J, Shin D. Imprint cytology of soft tissue myoepithelioma: a case study. Korean J Pathol. 2013;47:299-303
7. Go J. A case of soft tissue myoepithelial tumor arising in masticator space. Yonsei Med J.2005;46:710-4.
8. Robert E. Le Blanc, Janis T: myofibrome, myopéricytome, myoépithéliome et myofibroblastome de la peau et des tissus mous. Pathologie chirurgicale 2011, 4: 745-759. (9).
9. Antonescu C, Zhang L, Chang N, Pawel B, Travis W, Katabi N, et al. Fusion EWSR1- POU5F1 dans les tumeurs myoépithéliales des tissus mous. Une analyse moléculaire de soixante-six cas, y compris des lésions des tissus mous, des os et des viscères, montrant une implication commune du gène EWSR1. Gènes Chromosomes Cancer. 2010; 49: 1114-24.
10. Min K, Seo I, Pitha J. Ossifying fibromyxoid tumor: modified myoepithelial cell tumor? Report of three cases with immunohistochemical and electron microscopic studies. Ultrastruct Pathol. 2005;29:535-48immunohistochemical and electron microscopic studies.
11. Pivalaki M, Givissis P, Tzarou V. Soft tissue myoepithelioma of the hypothenar region: a case report. J Hand Surg 2007;32(5):674–6. Roentgenol 2006;186,270–271.
12. Gleason BC, Hornick JL. Myoepithlial tumours of skin and soft tissue: an update. Diagn Histopathol 2008;14(11):552–62.
13. Balogh Z, Deak L, Sapi Z. Malignant myoepithelioma of soft tissue: a case report with cytogenetic findings. Cancer Genet Cytogenet 2008;183(2):121–4.
14. Noronha V, Cooper DL, Higgins SA, Murren JR, Kluger HM. Metastatic myoépithélial carcinoma of the vulva treated with carboplatin and paclitaxel. Lancet Oncol 2006;7(3):270–1.
15. Lee JR, Georgi DE, Wang YW. Malignant myoepithelial tumor of soft tissue: a report of two cases of the lower extremity and a review of the littérature. Ann Diagn Pathol 2007;11(3):190–8.
16. Michal M, Miettinen M. Myoepitheliomas of the skin and soft tissues.Virchows Arch 1999; 434: 393–400.

****