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Systematic Review

Non-cutaneous soft tissue tumors and pseudotumors in the foot and ankle reported in Brazilian services

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Abstract

Objectives: Review the histological types of non-cutaneous soft tissue tumors and pseudotumors that affect the foot and ankle, reported in Brazilian health services.

Methods: An integrative review of the literature from the Scielo and PubMed platforms, based on the descriptors "Soft Tissue Tumors," "Pseudotumors," "Foot, "Ankle," and "Brazil." Those who answered the following research question: "What are the non-cutaneous soft tissue tumors and pseudotumors in the foot and ankle reported in Brazilian health services?" were included. The combination of each author's analysis contributed to reducing possible biases.

Results: Six articles were selected, yielding 23 reports of tumors and pseudotumors, which were subsequently categorized into malignant and benign tumors. There were five benign findings: three benign tumors, two giant cell tumors, one fibromatosis, and two pseudotumors classified as synovial chondromatosis. Eighteen malignant findings were reported: seven unspecified soft tissue sarcomas, four synovial sarcomas, three myxofibrosarcomas, one fibrosarcoma, one liposarcoma, one undifferentiated pleomorphic sarcoma and one papillary intralymphatic angioendothelioma.

Conclusion: In the last ten years, more reports of malignant than benign tumors have been reported. However, this finding does not necessarily reflect the Brazilian epidemiological reality, as it may represent a view in which resected benign tumors and pseudotumors are often not sent for anatomopathological study or reported in the literature.

Level of evidence I; Prognostic studies - investigating the effect of a patient characteristic on the outcome of disease

Keywords: Soft Tissue Tumors; Pseudotumors; Foot; Ankle; Brazil.

Introduction

The most common tumors reported in the foot and ankle are classified as primary and originate from bone tissue. Around 30% of tumors in this region originate from soft tissues, defined as tissues found under the skin, except bones[1,2]. Among them, the most prevalent benign tumors reported in the literature are lipoma, hemangioma, fibro histiocytoma, neurofibroma, schwannoma, and aggressive fibromatosis (desmoid tumor). Among malignant lesions, synovial sarcoma and myxofibrosarcoma stand out[2,3].

Pseudotumors represent a group of lesions often described as cysts, bursae, inflammatory masses, and adverse reactions to foreign bodies. However, the most accepted definition of pseudotumors is a non-neoplastic and non-infectious mass from an exudate surrounded by fibrous tissue originating from inflammatory processes[4]. Soft tissue pseudotumors in the foot and ankle comprise a diverse group of lesions, including synovial ganglia and cysts, intermetatarsal bursitis, epidermoid cysts, gouty tophi, rheumatoid nodules, Morton's neuroma and granuloma annulare[5].

Knowledge of the prevalence of benign and malignant neo formations in the foot and ankle in the Brazilian population more reliably supports the decision to investigate the masses identified in clinical practice further. Therefore, the objective of our study is to review the available literature on the histological types of non-cutaneous soft tissue tumors and pseudotumors that affect the foot and ankle, reported in Brazilian health services.

Study performed at the Universidade Federal de Alfenas, Alfenas, MG, Brazil.

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Methods

This study was elaborated based on the fundamental steps towards an integrative review. The primary research question that guided the literature search was: "What are the non-cutaneous soft tissue tumors and pseudotumors in the foot and ankle reported in Brazilian health services?" The descriptors utilized the controlled vocabulary DeCS/MeSH, using Portuguese, English, and Spanish keywords, including Soft Tissue Neoplasms or Soft Tissue Tumors, Pseudotumors, Foot, Ankle, and Brazil. The boolean operators OR and AND were employed during the literature search.

Inclusion criteria comprised publications featuring data or cases of non-cutaneous soft tissue tumors or pseudotumors affecting the foot and ankle in Brazil, published within the last decade on the PubMed and Scielo platforms, available in full and in Portuguese, English, or Spanish. Exclusion criteria included publications focusing solely on bone or skin tumors.

The search was conducted in January 2024 and resulted in 1048 studies. The search strategy followed the PRISMA guidelines, demonstrated in Figure 1.

Data extraction was performed on a validated data collection tool[6]. Article selection and data extraction were independently conducted by two researchers, with subsequent comparison of results to mitigate potential interpretation biases. Then, articles were synthesized and hierarchically ranked based on the level of scientific evidence, adhering to guidelines proposed by the Agency for Healthcare Research and Quality. No conflicts of interest were present during the execution of this research.

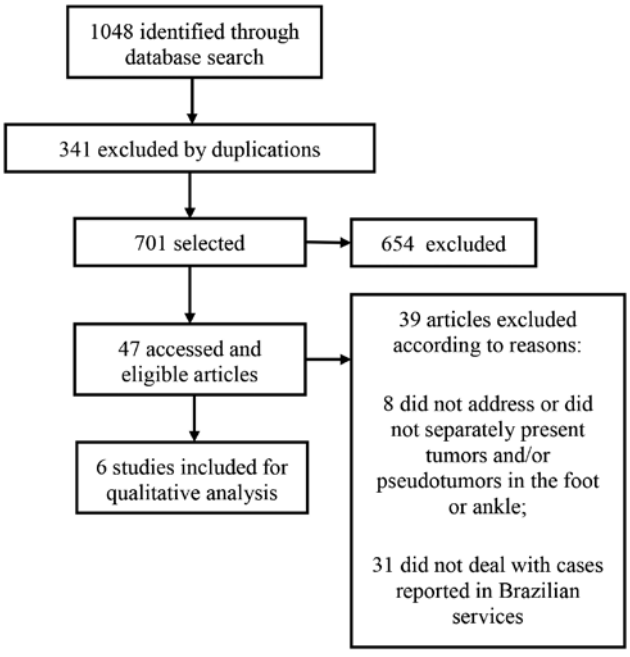

Figure 1. Flowchart, according to PRISMA, describing the literature search and study selection.

Results

This study included six articles, all conducted in Brazil. Regarding the year of publication, one (16.67%) was published in 2023, two (33.34%) in 2022, one (16.67%) in 2020, one (16.67%) in 2018 and one (16.67%) in 2016. Regarding study design, one (16.67%) was a retrospective study, three (50.00%) case reports, and two (33.34%) case series (Table 1).

The analysis of the selected articles resulted in 23 reports of neoplasms that affected the foot and/or ankle in Brazilian health services. Among the tumors and pseudotumors reported, 18 (78.26%) were malignant neoplasms, and five (21.74%) were pseudotumors or benign neoplasms.

Concerning the histological origin of the malignant tumors, seven were classified broadly as soft tissue sarcomas[7]. Four were categorized as undetermined origin tumors, all classified as synovial sarcomas[8]. Four were fibroblastic sarcomas, three myxofibrosarcomas[9] and one fibrosarcoma[10]. One was lipomatous sarcoma, which is a liposarcoma[8]. A fibrohistiocytic sarcoma is an undifferentiated pleomorphic sarcoma[8]. A tumor of lymphatic origin, in the case of a papillary intralymphatic angioendothelioma[11].

Regarding the pseudotumors and benign tumors classification, three were classified as fibrous tumors and two as pseudotumors. Among the fibrous tumors, one case of fibromatosis and two giant cell tumors were reported. All reported pseudotumors were synovial chondromatosis.

Discussion

In our study, most of the soft tissue tumors in the foot and ankle in Brazilian health services were classified as malignant, representing 78.26% of the neoplasms. This data differs from the findings of international studies that demonstrate a higher prevalence of benignity among tumors and pseudotumors derived from soft tissues in the foot and ankle, with proportions ranging from 70.7% to 97.1%[12,13]. Regarding the overall malignant tumors, a study shows that soft tissue sarcomas account for about 1% of all adult malignancies[14]. Only one epidemiological study was selected in this review, conducted in an oncology center in Brazil, covered musculoskeletal tumors at the ankle and showed a slight prevalence of malignant tumors of soft tissues (8%) compared to benign tumors (5%)[8].

One possible explanation for the lowest rate of benign tumors obtained in our study is due to underestimating the incidence of these tumors. Benign soft tissue tumors in the foot and ankle are often resected and not sent for anatomopathological examination[3]. Another hypothesis is the publication bias, in which reports of malignant findings are preferred over benign ones since the publication of malignancies would have a pronounced impact, as they are more aggressive and rare lesions.

Table 1. Results extracted from the studies included in the integrative review regarding authors, study design, level of evidence, sample, and histological types

Authors	Study design	Level of evidence	Sample	Histological types (number of cases) - Malignancy
Buscharino et al. (2023)	Retrospective study	3	121 participants (Middle age = 45.4 years old) 63 women 58 male	Soft tissue sarcoma (7) - Malignant
Kondo et al. (2016)	Case report	4	1 participant (47 years old) Male	Giant cell tumor (1) - Benign
Oliveira et al. (2022)	Case series	4	70 participants (Middle age = 21.6 years old) 41 male 29 women	Synovial chondromatosis (2) - Benign Giant cell tumor (1) - Benign Fibromatosis (1) - Benign Synovial sarcoma (4) - Malignant Liposarcoma (1) - Malignant Undifferentiated pleomorphic sarcoma (1) - Malignant
Pereira et al. (2022)	Case report	4	1 participant (7 months) Women	Infantile congenital fibrosarcoma (1) - Malignant
Silva et al. (2020)	Case report	4	1 participant (35 years old) Women	Papillary intralymphatic angioendothelioma (1) - Malignant
Zumárraga et al. (2018)	Case series	4	75 participants (Middle age = 49.7 years old) 44 women 31 male	Myxofibrosarcoma (3) - Malignant

The giant cell tumor of tendon sheath (GCT-TS) was reported in two selected articles[8,12], and knowledge about this tumor is crucial for an accurate diagnostic approach. GCT-TS is a benign tumor rarely located in the foot and ankle and most commonly found in the hands. It manifests as a painful or painless subcutaneous nodule with slow growth, resembling a synovial cyst[12,15]. Another benign tumor, synovial chondromatosis, involves the proliferation of cartilage tissue in the synovial membrane, tendons, and/or bursae. While there is no consensus on whether the disease is neoplastic or metaplastic, it is benign, rarely developing into malignancy such as chondrosarcoma[16]. Some international studies highlight the low incidence of this disease, especially in the foot and ankle, and the hands are identified as the most affected region[17,18].

Fibromatosis is classified as benign fibrous tumors of soft tissue[3]. In the foot, superficial fibromatosis manifests itself as plantar fibromatosis or Ledderhose syndrome, a hyperproliferation of the fibrous tissue of the plantar fascia. It is a rare and benign condition, locally infiltrative and rarely metastatic, with no well-defined etiology, resulting from the hyperactivity of mature fibroblasts[19]. Fibromatosis is associated with palmar fibromatosis, also called Dupuytren contracture, involving the fourth and fifth fingers, causing characteristic contractions[20].

Unlike synovial chondromatosis calcifications, synovial sarcoma calcifications are predominantly extra-articular, along with tendons and bursae, and have an irregular contour[21]. In the foot and ankle, the tumor initially affects the extra-articular tissue of a joint, slowly progressing towards the adjacent bone, and can generate ganglionary and pulmonary metastases[22]. Liposarcoma, a lipomatic sarcoma, corresponds to 9.8% to 16% of soft tissue sarcomas, the most common type among these[3]. This neoformation acquires large dimensions, has slow growth, and usually does not manifest painful symptoms. Due to poor symptoms, the diagnosis of liposarcoma is delayed[23].

An epidemiological study analyzing 623 tumors and pseudotumors of the foot and ankle reported undifferentiated pleomorphic sarcoma as the most prevalent malignant tumor, accounting for 22.2% of the malignancies studied[13]. Undifferentiated pleomorphic sarcoma, previously identified as malignant fibrous histiocytoma, is a highly aggressive soft tissue sarcoma that can metastasize to various organs[24]. Usually, this sarcoma affects deep soft tissues but can also manifest in skin and subcutaneous tissue[25].

Among soft tissue sarcomas in one-year-old children, infantile congenital fibrosarcoma is the most described, despite being a rare malignant neoplasm. It is a tumor histologically defined as a proliferation of dense mesenchymal fusiform cells with

hypervascularized areas, showing considerable clinical similarity to tumors and vascular malformations. Clinically, they manifest themselves as fast-growing masses that occupy soft tissues and evolve with bleeding[26].

Myxofibrosarcoma is the myxoid variant of malignant fibrous histiocytoma, described as a rare malignant mesenchymal tumor of soft tissues. Clinically, it presents as a pleomorphic mass of myxoid stroma, painless and slow-growing[14]. The histopathological study of the lesion allows diagnosis and staging by visualizing the proliferation of the myxoid stroma with pleomorphic cells and curvilinear vessels[27]. Another rare tumor, which has around 40 cases described in the literature, the papillary intralymphatic angioendothelioma, is a vascular neoplasm observed in soft tissues, frequently in the derme, but also deep tissues such as the spleen, testicle, and tongue[28].

Our study has compiled the findings of six publications that have addressed the topic of interest over the last ten years, resulting in one retrospective study, three case reports, and two case series. From this number of findings, it is evident that there is a limited amount of publications with a high level of evidence related to the prevalence of non-cutaneous soft tissue tumors and pseudotumors in the feet or ankle in Brazilian health services. The stimulation to study these neoplasms is fundamental to improving the knowledge that helps clinical practice, accelerates diagnosis, and promotes early treatment, aiming for greater chances of cure.

Conclusion

In the last ten years, in Brazilian health services, the following non-cutaneous soft tissue tumors and pseudotumors in the foot or ankle have been reported: unspecified soft parts sarcomas, giant cell tumors, synovial chondromatosis, fibromatosis, synovial sarcoma, liposarcoma, undifferentiated pleomorphic sarcoma, congenital infantile fibrosarcoma, papillary intralymphatic angioendothelioma and myxofibrosarcoma. There have been more reports of malignant tumors than benign tumors. However, this finding does not necessarily reflect the Brazilian epidemiological reality, but it may represent a view in which resected benign tumors and pseudotumors are often not sent for anatomopathological study or reported in the literature. Moreover, there is a notable scarcity of Brazilian publications addressing the topic. International literature guides the epidemiological, diagnostic, and therapeutic understanding of these neoplasms in the feet and ankle. However, Brazilian epidemiologic studies with higher levels of evidence are crucial for grounding the Brazilian orthopedist's approach and clinical decisions.

Authors' contributions: Each author contributed individually and significantly to the development of this article: GSB *(https://orcid.org/0000-0003-1753-2655), and VHMR *(https://orcid.org/0000-0003-0154-7385) Conceptualization, data curation, formal analysis, investigation, methodology, validation, visualization, writing – original draft, writing – review & editing; EASJ *(https://orcid.org/0000-0002-5054-874X) Conceptualization, data curation, formal analysis, investigation, methodology, project administration, supervision, validation, visualization, writing – original draft. All authors read and approved the final manuscript. *ORCID (Open Researcher and Contributor ID).

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