CASE REPORT

Subungual Synovial Sarcoma. An Undescribed Localization Mimicking an Inflammatory Injury. A Case Report

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ABSTRACT

A case of poorly differentiated subungual monophasic synovial sarcoma, occurring in a woman 76 years old, clinically presented as an inflammatory process is described. The literature does not report other cases in this location which made it very difficult to diagnose and confirmed the ubiquity of this neoplasm.

Key words: Immunohistochrmistry, snovial sarcoma, subungual

INTRODUCTION

Synovial sarcoma (SS), although long known to the oncological nomenclature and well-defined in its clinical, its histomorphological, immunophenotypic, and biomolecular characteristics, still remains undefined in its histogenesis. Since the first reports, this tumor, in addition to the most common localizations at the level of the soft tissues of the limbs, has been found in the most various sites. The literature does not report any case with a subungual location. For this reason, the authors deemed appropriate reporting of this case whose diagnosis has presented considerable difficulties both clinically and histopathologically.

CASE REPORT

A 76-year-old woman had a left toenail injury, which was interpreted and treated as inflammatory. As it worsened despite the treatments, the patient underwent an onychectomy. Once the nail was removed, in the context of purulent necrotic material, a fleshy, pink-colored lump was highlighted. The material was sent for histological examination. Subsequently, the patient was subjected to the disarticulation of the big toe. Four years after the operation, the patient died of widespread metastases.

MATERIALS AND METHODS

The material consists of a few small fragmentst of soft consistency of grayish coloring and 1.3 cm size. The material was fixed in formalin and embedded in paraffin, and the sections were colored with HE. The immunohistochemical investigation was performed with a large panel of antibodies, as shown in Table 1.

Histology

The sample under examination is largely made up of fibrino-necrotic material and granulation tissues [Figure 1a]. In this context, there is a nodular area [Figure 1b] consisting of elements of greater volume – globular [Figure 1c] or spindle-shaped [Figure 1d] – with a voluminous and hyperchromatic nucleus – disordered [Figure 2a] or aggregate – sometimes, in morular formations [Figure 2b-d]. This proliferation tends to disperse into the surrounding phlogistic tissue.

Immunohistochemistry

Table 2 demonstrates the result of immunohistochemical investigations.

On the basis of the morphological pattern and the immunohistochemical profile, the diagnosis of Monophasic Poorly Differentiated SS was proposed.

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DISCUSSION

The possibility of an onset of SS outside the osteoarticular system, larynx, and at a distance from the joints was reported by Masson, "the starting point is any synovial bursa, more frequently one of the lower limbs, above all that of the knee.

Table 1: Panel of antibodies

Ab	Dilutions
CKAE1-AE3	Monoclonal 1:50 DAKO Agilent
EMA	Monoclonal 1:50 DAKO Agilent
SMACT	Monoclonal 1:50 DAKO Agilent
DESM	Monoclonal 1:50 DAKO Agilent
S100	Monoclonal 1:400 DAKO Agilent
CD10	Monoclonal 1:400 DAKO Agilent
CD34	Monoclonal 1:20 DAKO Agilent
CD117	Polyclonal 1:400 DAKO Agilent
KI67	Monoclonal 1:75 DAKO Agilent
Bcl2	Monoclonal 1:50 DAKO Agilent
CD68	Monoclonal 1:50 DAKO Agilent
VIM	Monoclonal 1:50 DAKO Agilent
MELA	Monoclonal 1:75 DAKO Agilent
Hmb45	Monoclonal 1:50 DAKO Agilent
CD56	Monoclonal 1:50 DAKO Agilent
CD68	Monoclonal 1:50 DAKO Agilent
CD138	Monoclonal 1:25 DAKO Agilent
ALK	Monoclonal 1:20 DAKO Agilent
TLE1	(M–101) 1:50 SANTA Cruz

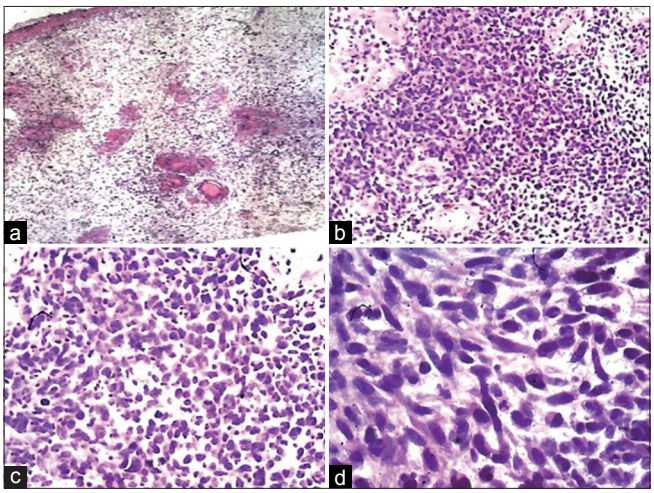

Figure 1: (a) Necrotic-phlogistic tissue that constitutes the great part of the material; - ×HE 100X; (b) hypercellular nodule in the context of necrotic tissue-HE ×100; (c) the same area higher magnification-HE ×120; (d) a mixture of globular and spindle elements-HE ×150

Personally, I have observed an indisputable case of laryngeal load due probably from the cricoarytenoid bursa. Some seem to come from aberrant synovial structures".[1]

In addition to the classic localizations of the upper and lower limbs, the abundant subsequent literature reported primitive onset in a wide range of localizations such as the head-neck area (mouth, tongue, pharynx, larynx, and thyroid), the gastroenteric tract (liver, stomach, and colon), the urogenital tract (kidney, prostate, penis, ovary, vagina, and vulva), skin, bone, spine, and spinal meninges.[2]

Although by convention we continue to call this neoplasm as SS, its derivation from synovial structures is strongly questioned or even denied.[3] To date, however, no plausible histogenesis is proposed so much that the WHO Classification of Tumors of Soft Tissue and Bone in its latest edition (2013) places this neoplasm in the group of tumors of uncertain differentiation.[4]

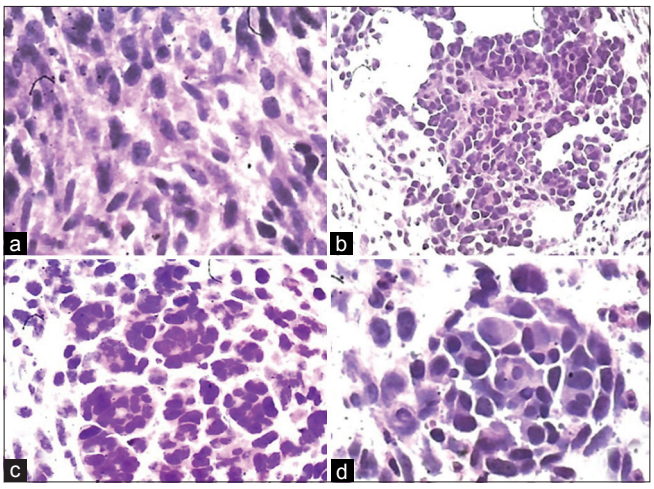

Figure 2: (a) Globose and spindle elements at higher magnification HE ×175; (b-d) Morular aggregates at various magnifications HE ×100, ×!50, ×200

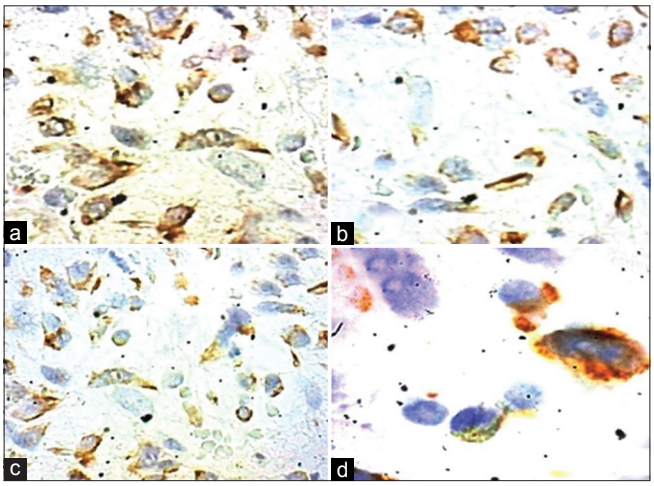

Figure 3: (a-d) CK AE1-AE3 – ×150, ×200

Table 2: Results of the immunohistochemical investigation

AEI–AE3+	EMA+		CD99+	BCL2+	CD10+	SMA+–	DESM–	VIM+	S100+–	KI67+ >20%	TLE1+
Figure 3a-d	Figure 4a and b		Figure 5a and b	Figure 4c and d	Figure 6c and d	Figure 6a	Figure 6b	Figure 5c and d	Figure 7a	Figure 7b	Figure 7c and d
CD34–	CD68–	MEL A-	Hmb45–	CD56–	CD68–	CD138–	ALK–				

The great morphological variability and its practically ubiquity make this neoplasm a thorny diagnostic problem whose differential diagnosis must be made not only with morphologically similar forms but also in relation to the site of onset of the neoplasm.

For these reasons, the diagnostic work-up of this case has been very tortuous and complicated by the scarcity of available material and the concomitant necrotic-phlogistic phenomena.

The morphological pattern both on the cytological and architectural level was difficult to interpret because of contamination by the elements of inflammation and necrosis. In the best readable areas, the elements showed globular or spindle morphology and a disordered arrangement, sometimes moulood.

This morphology could be compatible both with a melanocytic neoplasm, with a sarcomatoid carcinoma, and with a poorly differentiated sarcoma.

Immunohistochemistry was addressed primarily to document the tumor more likely eventuality at that site: A melanoma. The negativity to specific antibodies (HMB45 and MELA) made this event unlikely. However, in that context, the possibility of squamous sarcomatoid carcinoma, given the positivity to epithelial antibodies (CK, EMA), which did not conflict with the concomitant positivity for Vimentin, could also not be excluded. The presence of positivity for Bcl2 and CD99, however, made this diagnosis unlikely. Concurrent positivity for Vimentin and epithelial antibodies opened diagnostic options to two non-epithelial neoplasia: Epithelioid sarcoma and SS. The subungual site would favor the former, while the negativity for CD34, the positivity for CD99, and Bcl2, the latter. The paucity of material required a choice or search INI1, the loss of which would have supported the diagnosis of Epithelioid Sarcoma or search TLE1, whose positivity would have made plausible the diagnosis of SS. The authors opted for the latter taking into account the positivity for CD99 and Bcl2. In fact, the test with the TLE1 antibody gave a clearly positive result. The translocation of the SYT gene could not be sought due to the necrotic state and the small amount of residual material. Nevertheless, the combination of the various immunophenotypic positivity makes this diagnostic interpretation more than plausible. The following Table 3 shows the significant antibodies for the diagnosis of SS reported in the literature.[5,6]

Table 3: Antibdies for the diagnosis of SS

Ab		%
TLE1	+	100
EMA	+	90
CK	+	88
Bcl2	+	93
CK7	+	73
CK19	+	100
Calp	+	?
NY-ESO-1	+	?
CD99	+	48
AE!-AE3	+ or –	88
E-Cadherin	+ or –	?
βcatenin	– or +	100
S100	– or +	39
SMA	– or +	0
CD117	– or +	rare
SOX10	– or +	?
DESM	–	1
CD34	–	0
h-Caldesmon	–	0

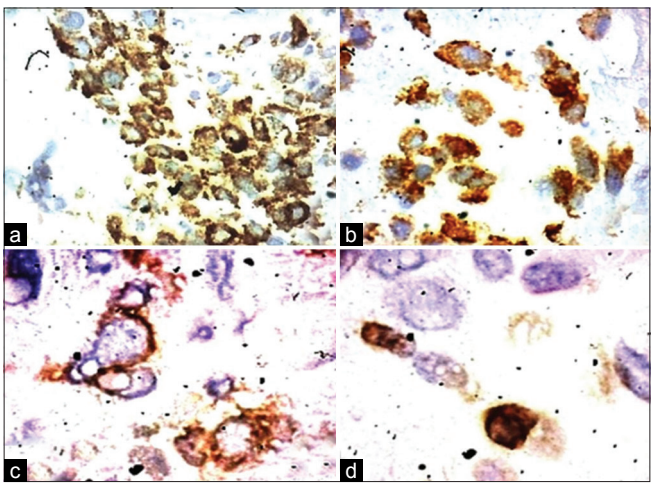

Figure: 4 (a and b) EMA ×150; (c and d) BCl2, ×200

Positives for SMA, desmin, and CD10 that are not part of SS's phenotypic immune profile are expressions of the

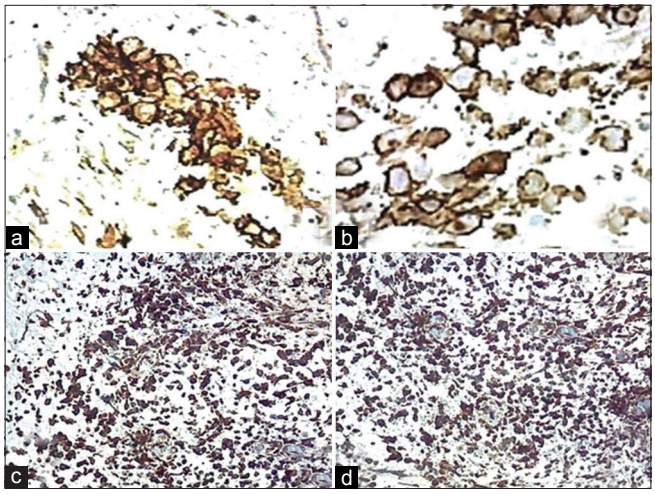

Figure 5: (a and b) CD 99 ×150; (d and e) Vimentin ×150

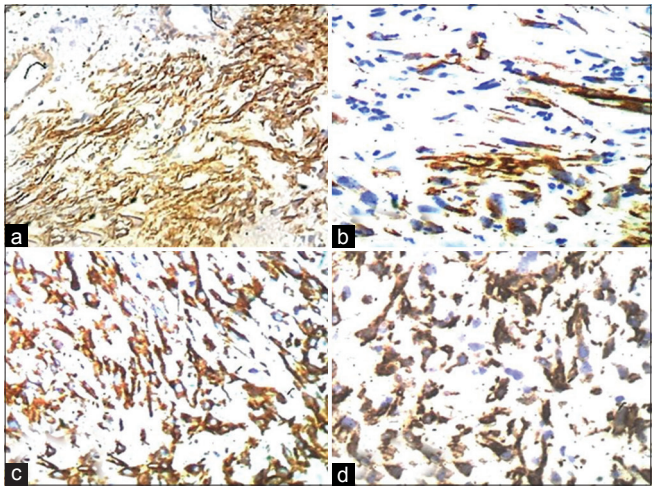

Figure 6: (a) SMA ×100¸ Desm ×100; (c and d) CD10 ×!00, ×150

myofibroblastic fraction reactive to the inflammatory process and neoplastic tissue.[7]

Scrolling through the literature, we have found a unique report of SS of the distal phalanx of a toe. From the photos, the lesion would appear on the medial side of the finger and not in the subungual area.[8]

CONCLUSIONS

This case, beyond its uniqueness, confirms the ubiquitous onset of the SS, the occurrence of which must be kept in mind even in the least probable sites and can represent a dangerous diagnostic pitfall.

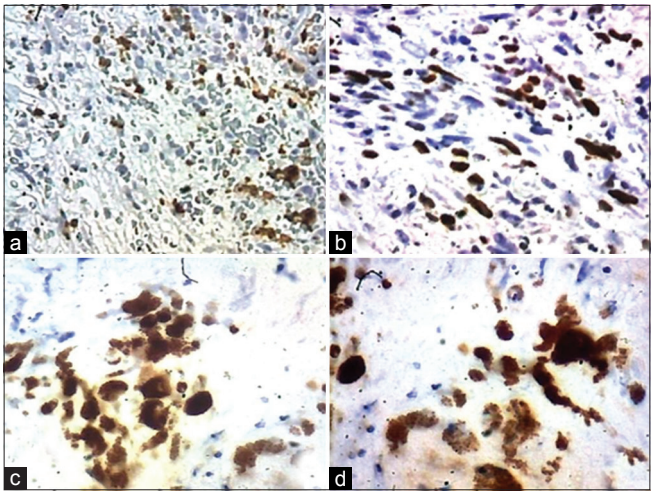

Figure 7: (a) S100 ×100; (b) Ki67 ×2150; (c and d) TLE1 ×175

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