ID Design Press, Skopje, Republic of Macedonia
Open Access Macedonian Journal of Medical Sciences. 2018 Jan 25; 6(1):76-78.
**Special Issue: Global Dermatology-2**
https://doi.org/10.3889/oamjms.2018.039
eISSN: 1857-9655
***Case Report***

# A "Yellow Submarine" in Dermoscopy

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**Citation:** Satolli F, Rovesti M, Zucchi A, Gandolfi M, Feliciani C, Tchernev G, Wollina U, Gianfaldoni S, Lotti T. A "Yellow Submarine" in Dermoscopy. Open Access Maced J Med Sci. 2018 Jan 25; 6(1):76-78. https://doi.org/10.3889/oamjms.2018.039

**Keywords:** histiocytic sarcoma; dermoscopy; yellow colour; CD68; WHO classification lymphomas

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**Received:** 13-Sep-2017; **Revised:** 12-Oct-2017; **Accepted:** 14-Oct-2017; **Online first:** 07-Jan-2018



**Funding:** This research did not receive any financial support

**Competing Interests:** The authors have declared that no competing interests exist

**Abstract**

**BACKGROUND:** Histiocytic sarcoma (HS) is an extremely rare, non-Langerhans cell tumor. HS affects especially adults, its etiology is unknown yet. Skin could be interested by papules or nodules, single or multiple.

**CASE REPORT:** A Caucasian man in his late 40s came to our clinic for a naevi evaluation. During the visit, a rose papulonodular lesion was observed in the lumbar region. This lesion was completely asymptomatic, and it had been there for an indefinite period. The clinical evaluation revealed that the lesion appeared elevated, of 9 x 15 mm in dimension, symmetrical and of a homogeneous pinkish colour. The videodermoscopical evaluation revealed a homogeneous yellow central pattern, polymorphic vessels, an eccentric peripheral pigmentation and a white collar. An excisional biopsy was performed. The morphology and the expression of CD163, CD68 and/or lysozyme to the immunophenotypic analysis, revealed the true nature of the lesion.

**CONCLUSION:** HS is usually diagnosed at an already advanced clinical stage and it has a high mortality rate even today. Dermoscopy, showing a yellow and distributed homogeneously colour, can facilitate its hard diagnosis.

## Introduction

Histiocytic sarcoma (HS) is an extremely rare, non-Langerhans cell disorder with morphologic and immunophenotypic evidence of histiocytic disorders [1]. HS affects all ages, but it is more common in adults (46–55 years). The aetiology of this disorder is unknown [2]. Systemic symptoms (such as fever, night sweats, and weakness) and other skin, hepatosplenic or intestinal manifestations could appear. For some years there has been confusion about this type of a tumour and its terminology. Nonetheless, thanks to the advances in knowledge in the field of genetics and biology in recent years, we are today able to classify and recognize these kinds of hematologic neoplasms.

The latest WHO classification of lymphomas, published in 2008 [3] and updated in 2016 [4], describes the HS within the histiocytic and dendritic cells neoplasms; this group includes pathologies which share the same functional properties of their normal counterpart (phagocytosis and/or processing and presentation of antigens) rather than their cell of origin [4].

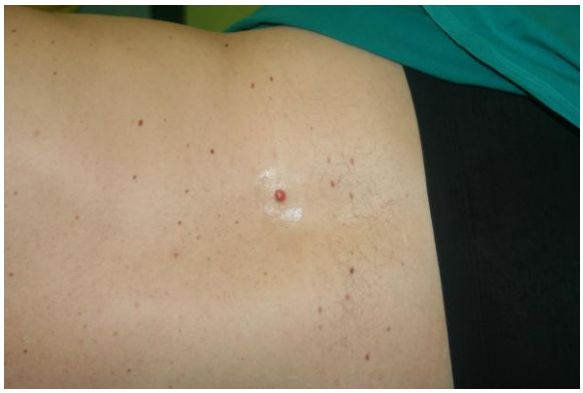
## Case Report

A Caucasian man in his late 40s came to our clinic for a naevi evaluation.

During the visit, a rose papulo-nodular lesion was observed in the lumbar region (Figure 1: A, B). This lesion was completely asymptomatic, and it had been there for an indefinite period. The clinical evaluation showed that the lesion appeared elevated,

of 9x15 mm in dimension, symmetrical and a homogeneous pinkish colour.

A)



B)

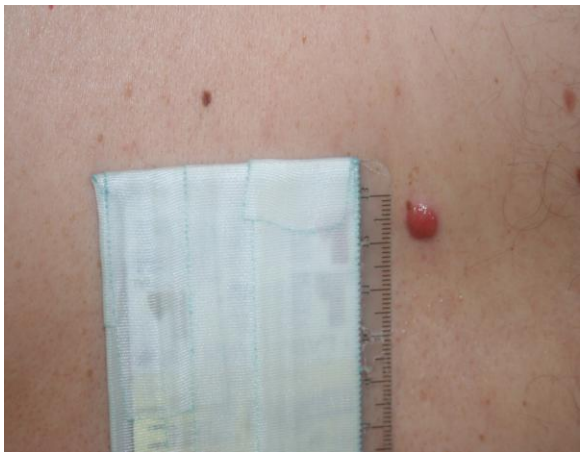


*Figure 1: Clinical evaluation (A, B)*

Suspecting a malignant lesion, also a videodermoscopy of the lesion was also immediately performed, and this revealed a homogeneous yellow central pattern, polymorphic (arborizing, dotted and glomerular) vessels, an eccentric peripheral pigmentation and a white collar (Figure 2).

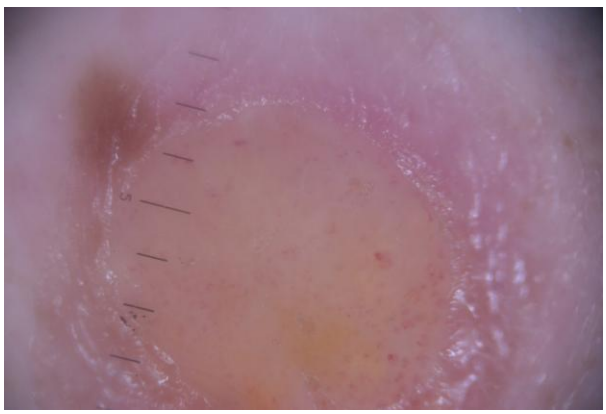


*Figure 2: Dermoscopic evaluation*

An excisional biopsy was performed. The morphology showed a diffuse proliferation of large cells, with grooved and indented nuclei and abundant eosinophilic cytoplasm (Figure 3: A, B). The immunochemistry of the surgical specimens showed cells with positivity for CD68, CD163 and CD4, with negativity for S100, CD34 and HMB45 (Figure 4). The Ki67 index was about 18% (Figure 4).

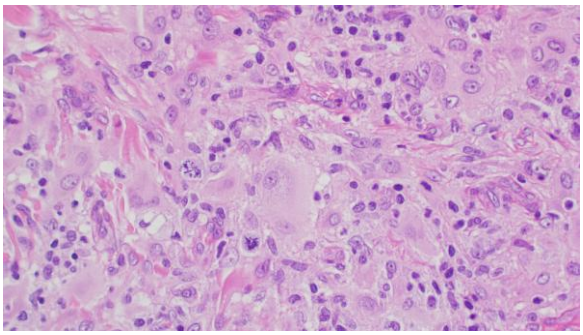


*Figure 3: Morphology*

The patient has always been in good health and was not taking any drugs. He had a skin type II according to Fitzpatrick's classification; he did not suffer from any skin diseases and did not have melanoma familiarity. In the previous check-ups of neavi no atypical lesions had been observed.

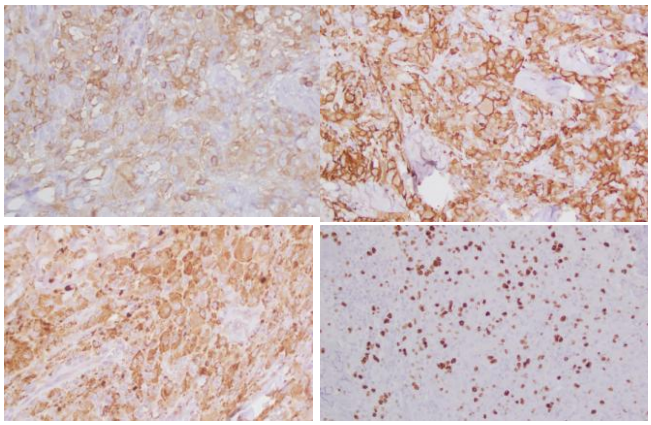


*Figure 4: Immunophenotype (first line, from left to right: CD4, CD68; second line, from left to right: CD163, Ki67). [Courtesy of Dr Mancini C., Unità Operativa di Anatomia e Istologia Patologica, Dipartimento Diagnostico, Università degli Studi di Parma]*

## Discussion

Even though the lymph nodes are the most common site of HS presentation, the gastrointestinal tract, the soft tissue and the skin could also be affected. Skin manifestations can be isolated or associated with systemic involvement. A cutaneous HS could appear as single or multiple papules and nodules of a light colour. The clinical differential diagnosis must take into account other cutaneous lesions: amelanotic melanoma, B or T cell lymphoma, dermatofibroma, dermatofibrosarcoma protuberans, juvenile xanthogranuloma, etc.

With dermoscopy, the cutaneous HS is predominantly yellow, distributed homogeneously.

This colour, in dermoscopy, was investigated by Cavacchini et al. some years ago [5]. In the case of HS, the yellow may appear in various shades and is the background to some polymorphic vessels (arborising, dotted or glomerular) and a white-pinkish collar. In fact, the presence of polymorphic vessels represents a highly risky dermoscopic pattern that can be seen not only in amelanotic melanoma or Spitz naevi [6] but also in other malignant lesions, such as an HS. When confronted with this kind of lesion, excisional biopsy is the most accurate diagnostic method. The morphology is characterized by large atypical pleomorphic cells with eosinophilic cytoplasm and large, round to oval, irregular nuclei [1].

The immunophenotype is characterized by the expression of one or more histiocytic markers, such as CD163, CD68 and lysozyme [1]. CD163, a haemoglobin scavenger receptor whose expression is limited to neoplasms of macrophage/histiocytic lineage, is more specific than CD68, and a strong immunoreactivity for this antigen is suggestive of histiocytic differentiation [2]. The Ki-67 index is variable. Staging studies, including imaging studies such as computed tomography (CT) or a combined positron emission tomography (PET/CT), should be performed after the diagnosis to determine the extent of the disease.

Unfortunately, HS is usually diagnosed at an already advanced clinical stage, and it does not have a good response to chemotherapy. This tumour has a high mortality rate and, in fact, most patients die from the progressive disease within two years [7].

Due to the aggressive course and the limited treatment options of this tumour, performing a careful clinical and dermoscopic evaluation is essential to help the final histological diagnosis. Over the last ten years, there has been various case reports in the literature regarding patients with cutaneous or subcutaneous histiocytic sarcoma. Only one of them [8] focused on the use of dermoscopy on this kind of lesion.

Dermoscopy, a non-invasive analysis technique, used especially for the early detection of melanoma, an instrument that is becoming increasingly widespread and more commonly used in dermatology clinics; it represents a tool which helps increase the low clinical sensitivity of the naked eye [9].

In conclusion, the early diagnosis of a rare and malignant tumour-like HS currently remains hard. However, dermoscopy can facilitate it and lead to quickly performing an excisional biopsy, thus improving the prognosis for patients.

When dermoscopy notices the colours pink or black in a lesion, it should indicate to dermatologists that something may be wrong. Nevertheless, yellow too could be suspicious, and it is a wake-up call in dermoscopy… beware of the “yellow submarine”!

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