CLINICAL CASE

# SPLENECTOMY FOR SPLENIC HISTIOCYTOSIS - CASE PRESENTATION AND REVIEW OF LITERATURE

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**ABSTRACT**

*Langerhans cell histiocytosis (HCL) is a pathological condition defined by the excess production of histiocytes, a type of dendritic cells involved in the immune response, which is sometimes associated with hematological diseases such as leukemias. In terms of incidence, this condition does not occur by the routine in medical practice, most cases being discovered accidentally. The presented case describes a 47-year-old woman with isolated splenic HCL who manifested clinically repeated pain in the left hypochondrium. The computer tomography described nodular lesions with multiple localization in the spleen. The histopathological and immunohistochemical results, however, provided the diagnosis of certainty of isolated splenic HCL. The surgical intervention performed was splenectomy, with favorable early and late postoperative evolution. The patient does not show clinical and paraclinical changes during the periodical follow up during the 7 years since the surgical intervention. The early diagnosis and the surgical intervention represent complete solution of these types of cases.*

**KEYWORDS**: *surgery, histiocytes, Langerhans, treatment, splenectomy*

## INTRODUCTION

Histiocytosis of Langerhans cells (HCL), is defined by the increase in the number of histiocytes, these representing the cells involved in the presentation of antigen to T-type lymphocytes, being characterized by a low incidence, namely 1 case per 200,000 people if we are talking about children under 5 years old, unlike adults, where the number of cases is significantly lower, 1 case in 500,000 people [1].

Few cases of HCL are mentioned in the specialty literature and most of them are discovered accidentally. Therapeutic resources in this case were quite limited due to the incompletely elucidated ethiopathogenic mechanism, but the latest advances that led to the determination of new genetic mutations of the patients opened the way to new treatment options [2,3].

Initially, all cases were treated as a benign condition, but the discovery of the BRAF – V600E gene mutation brings oncological treatments into question [4,5].

From the point of view of symptomatology, it can set in slowly, with mild pain syndrome at the abdominal level, until the installation of the occlusive syndrome, by compression on the intestinal loops, or it can start with an acute form, such as Letterer - Siwe

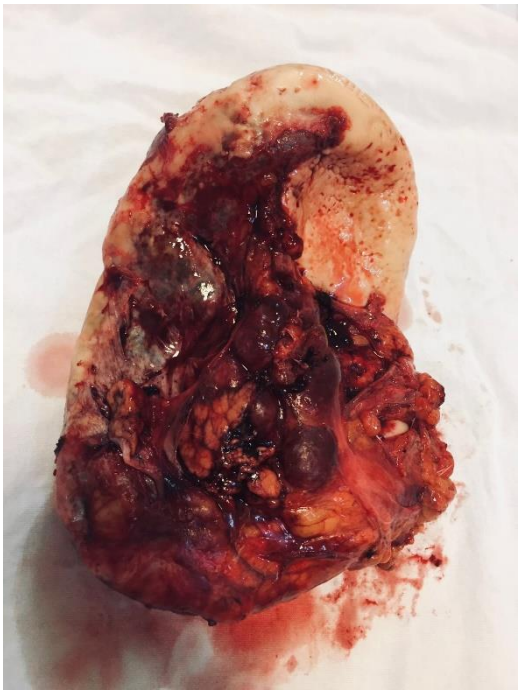
disease, with the proliferation of histiocytes at plurivisceral level, which is characterized by an unfavorable prognosis [6,7].

In what follows, we present a case of HCL with isolated splenic involvement in a 47-year-old patient.

## CASE PRESENTATION

The 47-year-old patient presented to our hospital with repeated pain in the left hypochondrium for 5 weeks. No pathological changes were observed after the clinical examination. From a biohumoral point of view, the patient was within normal limits, including a hemoglobin level of 13.1 g / dL, a leukocyte count (WBC) of 7.0 × 103 / uL, and a platelet count of 243 × 103 / uL. Liver and coagulation tests were within normal limits. Abdominal ultrasound revealed splenomegaly and hypoechoic masses measuring approximately 1.2 × 1.25 cm located along the spleen, and computed tomography (CT) described multiple nodular lesions at the splenic level. Examination of the bone marrow revealed no pathological changes. Hematological evaluation recommended splenectomy, which was performed openly (Figure 1). On the resection piece were found nodules with a diameter of approximately 1.2 cm.

**Figure 1 - Postoperative piece – Splenectomy**



Histopathological examination described proliferation of Langerhans cells with oval-shaped nucleus, with a longitudinal groove at the level of the nucleus, a "coffee bean" appearance and eosinophil infiltration.

The immunohistochemical examination identified the presence of S-100 protein, CD1a and CD45RO markers, respectively Vimentin. The rest of the markers were negative. Following the results of the clinical and paraclinical examinations, the definite diagnosis of HCL was confirmed.

To specify that HCL was isolated only at the splenic level, the patient was transferred to the hematology clinic and then reevaluated in the endocrinology clinic where additional investigations were performed, where no pathological values were determined for thyroid hormones, gonadotropes .

Chest CT and MRI (Magnetic Resonance Imaging) of the cephalic extremity, whole body bone scintigraphy did not describe pathological lesions. Finally, There were no evidence for other organs involvement, the only affected organ being the spleen - isolated diagnosis of HCL of the spleen. The patient did not benefit from adjuvant treatment, but only from monitoring.

## DISCUSSION

HCL has cells that are very similar to those in the bone marrow, which often makes the diagnosis difficult, requiring further investigations for a clear diagnosis. In general, this disease affects the skin tissue, the bone tissue, the ganglia and quite rarely ends up invading other important organs.

The involvement in the pathogenic mechanism of cytokines includes this condition in the framework of immune-type disorders, but the recent discoveries related to gene mutations lead to the inclusion of HCL in the group of neoplasms, so it can be considered that we can talk about a myeloid neoplasia with an inflammatory character [8,9].

The clinical manifestations of HCL differ depending on the organ affected and can range from asymptomatic variants to painful syndromes with repeated exacerbations. Particular to the case were the pain syndrome that occupied the left hemiabdomen, but also the imaging results that described splenomegaly, but

also multiple nodular lesions at the splenic level determined by ultrasound.

On the surgical specimen, the specific histopathological tests, respectively the immunohistochemical tests, were performed. Those tests provided the diagnosis of certainty.

At the present time, based on the modified Lahey criteria, the forms of the disease are divided according to the degree of risk into those with single organ damage (SS-LCH) and plurivisceral damage (MS-LCH), such as liver and lung dysfunction or hematopoietic system. Imaging investigations are the ones that can help us in this classification of the disease, by highlighting possible lesions detected at plurivisceral level or their absence [10]. Considering this stratification, the presented case can fit into the SS-LCH subtype without the secondary damage at the systemic level.

The BRAF V600E oncogenic mutation was detected in the majority of HCL patients, which places our HCL into an oncological pathology [11]. Although research has been conducted into the pathophysiological mechanisms of the disease and studies with various types of treatment, no consensus has been reached regarding the standardization of therapy. Currently, the therapeutic resources used are either surgical, or oncological treatments, chemotherapeutic medication or radiotherapy [12]. The main objective of the treatment is to prevent complications or relapses, The treatment is chosen according to the initial stage of the disease [13,14].

Therefore, prognosis, 5-year overall survival were the basis of HCL classification. The prognosis of patients with HCL is influenced by the stage of the disease, this being more reserved in the case of the multivisceral form, and pulmonary damage or poor response to the initial treatment represents an additional risk factor. Likewise, if only one organ is affected, the prognosis is significantly improved [15].

We stress that isolated HCL of the spleen is a condition that is not often encountered in medical practice. The standard in the diagnosis of splenic histiocytosis is entirely histopathological examination and immunohistochemistry, requiring the exclusion of the presence of MS-HCL of the spleen. As a final observation, splenectomy represents the indicated treatment for isolated HCL of the spleen.

## CONCLUSION

HCL is a pathological condition with a low incidence, which requires a certain diagnosis and a correct staging in order to opt for a personalized treatment. The isolated form of spleen damage benefits from first-line surgical intervention and has a good prognosis, and in the case of multivisceral damage, oncological treatments such as chemotherapy or radiotherapy are used, followed by follow-up.

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