ABSTRACT

Background: Hamartoma is a slow-growing, rare mixed benign tumor. In general, it does not produce symptoms, so it is more commonly found as an incidentaloma during autopsies or laparotomies. Incidence of splenic hamartomas is low, representing 0.001% of the general population.

Clinical case: We report the case of a 39-year-old male without relevant antecedents. The evolution of his condition began 2 months prior with pyrosis and occasional pain in the upper hemi-abdomen. Abdominal ultrasound demonstrated a pseudocystic tumor in the spleen. Magnetic resonance showed four lesions in the spleen, predominating with a large, bilobulated lesion in the inferior pole of 12 × 10 × 9 cm. Splenectomy was done without complications. The patient was discharged to home on the third postoperative day. Pathological report showed a splenic hamartoma.

Conclusions: Hamartomas of the spleen as in other localizations are benign lesions found as incidentalomas because only few produce symptoms. The final diagnosis is made histopathologically. Definitive treatment is splenectomy and treatment of choice is complete laparoscopic intraabdominal splenectomy. Although there are few incidences in Mexico, it is of vital importance that the physician considers it among the differential diagnoses when evaluating a tumor.

Key words: Hamartoma, incidentaloma, pseudocyst, splenectomy.
ABSTRACT

Hamartomas are rare benign, slow-growing mixed tumors that do not generally cause symptoms; therefore, it is common to find them as an incidental finding during autopsies or laparotomies.\textsuperscript{1,2}

The incidence of hamartomas of the spleen is so low that it is present in ~0.001\% of the population in general. They are one or two times more common in males than in females and very rare in children. They are typically diagnosed during adulthood, especially from the sixth decade of life. Due to the low frequency of reported cases, we decided to present this case from the Hospital San José Tec of Monterrey, Monterrey, Nuevo Leon. To the author’s knowledge this is the only case reported in Mexico.

CLINICAL CASE

We present the case of a 39-year-old male without any significant medical history. The patient developed heartburn and occasional pain in the upper hemi-abdomen 2 months prior without other symptoms being present. Abdominal ultrasound revealed a pseudocystic tumor of the spleen. Magnetic resonance demonstrated four lesions in the spleen, one large bilobulated lesion on the inferior pole of 12 × 10 × 9 cm with a thin septum directed to the center of the lesion (Figure 1).

The lesion was surgically resected. Massive splenomegaly was found on entering the cavity (Figure 2). Splenectomy was carried out without complications and the patient was discharged on the third postoperative day (Figure 3). The pathology report was splenic hamartoma.

DISCUSSION

According to Yusul and Assaf,\textsuperscript{4} the first description of a splenic hemangioma was made by Rokitanski in 1861. Since that time there have been ~150 cases reported, of which only 20 were
in children. Primary splenic tumors are not common, with hemangioma being the most common and hamartoma the rarest. Its origin is linked to genetic disorders, ionizing radiation or maternal viral infections during early stages of pregnancy but in the majority of cases is idiopathic.

There are two types of splenic hamartomas: 1) follicular and 2) pulpous and fibrous, which is the most common.

The majority of cases are asymptomatic. Only in the case of red pulp hamartomas are there manifestations such as thrombocytopenia, anemia, splenomegaly and pancytopenia. Few cases with spontaneous rupture, especially in adults, have been reported.

Hamartoma is also known as splenoma, which is a biologically benign lesion composed only by elements of red pulp. Follicles and dendritic cells of the white pulp are lacking but contain few fibrous trabecula. In the hamartoma there may be foci of extramedullary hematopoiesis, lipid-loaded macrophages, and areas of lipophagia and plasmocytosis, which are seen with immunohistochemistry. The classic characteristic is that the cells lining the vascular channels express CD8 (Figure 4) and CD68+ macrophages are appreciated in the cords of the pulp along with cells positive for vimentin.

Essential histopathological elements for diagnostic confirmation are conglomerates of sinusoids covered by endothelial cells, basal membrane of connective tissue, absence of white pulp and a thin layer of connective tissue, which outlines the lesion.

A variety of surgical procedures are used for treating splenic hamartoma such as partial or total splenectomy, whether by laparotomy or laparoscopy. Laparoscopy is the preferred technique because it offers better cosmetic results, less pain, fewer days of hospitalization and does not require blood transfusion. The most common treatment for splenic hamartoma is complete intraabdominal splenectomy.

In conclusion, hamartomas of the spleen and other locations are benign lesions that are incidentally found. Final diagnosis is supported by histopathological study. Its definitive treatment is surgical resection. Complete laparoscopic intraabdominal splenectomy is the preferred technique. Even though its incidence is very low in Mexico, it is of vital importance for the physician to take this condition into consideration as a differential diagnosis when evaluating a tumor.

REFERENCES