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UNDIFERENTIATED EMBRYONIC SARCOMA OF THE LIVER: A CASE REPORT

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Abstract: Liver embryonic sarcoma (EHS) is a mesenchymal neoplasm of undetermined origin. This neoplasm is characterized by presenting solid and cystic components that may present points of necrosis or hemorrhage when in advanced stages. It is a malignant neoplasm, which affects in most cases children and adolescents between the first and second decade of life, with a peak incidence in children from five years of age. A good imaging exam associated with the clinical history (age at onset) and alpha-fetoprotein levels can help in the preoperative diagnosis. However, in clinical practice, especially when the disease affects another age group, we arrive at the diagnosis only through a post-surgical anatomopathological study. Among the differential diagnoses we can mention hemangioma, hepatoblastoma, hydatid cyst, hepatic cystadenoma and hepatic lymphangioma. Ultrasonography is the examination performed initially and helps in differentiating the components of the lesion; however, magnetic resonance imaging with the aid of liver-specific contrast (Primovist) is the gold standard for differentiating benign and malignant liver lesions, in addition to being essential for vascular assessment and involvement of adjacent structures, being essential to define the surgical strategy and resectability of the injury. This report describes the case of a 16-year-old female patient, who was admitted to the emergency room of our service due to vague and recurrent abdominal pain, associated with severe pain in the right shoulder and hyporexia. Patient was evaluated by our team, who chose to investigate the hospitalized patient. During hospitalization, magnetic resonance imaging of the abdomen was performed, which showed an oval hepatic subcapsular formation in segments VII and VIII of the right lobe and part of segment IVa. The lesion had thin walls, irregular internal membranes and possible signs of necrosis

or recent hemorrhage. The lesion measured 14x12 cm and was in close contact with the diaphragm. In view of this finding, it was decided to perform a segmental hepatectomy of segments IV, V and VIII, associated with excision of the diaphragmatic portion adhered to the lesion. In the anatomopathological and immunohistochemical study of the surgical specimen, a malignant mesenchymal neoplasm of indeterminate histogenesis and characteristics compatible with hepatic embryonal sarcoma (Ki-67 of 40%) with free surgical margins were observed. The patient had a satisfactory postoperative evolution, being discharged on the seventh postoperative day and is currently being followed up by the clinical oncology team for complementary treatment and follow-up.

Keywords: Sarcoma, embryonic, liver, surgery.

INTRODUCTION

Liver embryonic sarcoma (HES) is a mesenchymal neoplasm of undetermined origin that is characterized by presenting solid and cystic components that may present points of necrosis or hemorrhage when in advanced stages. It is a malignant neoplasm, which affects in most cases children and adolescents between the first and second decade of life, with a peak incidence in children from five years of age. A good imaging exam associated with the clinical history (age at onset) and alpha-fetoprotein levels can help in the preoperative diagnosis. However, in clinical practice, especially when the disease affects another age group, we arrive at the diagnosis only through a post-surgical anatomopathological study. Among the differential diagnoses we can mention hemangioma, hepatoblastoma, hydatid cyst, hepatic cystadenoma and hepatic lymphangioma. Ultrasonography is the examination performed initially and helps in differentiating the components of the lesion; however, magnetic resonance imaging with the aid of hepato-specific contrast (Primovist) is the gold standard for differentiating benign and malignant liver lesions, in addition to being essential for vascular evaluation and involvement of adjacent structures, helping to define the resectability of the lesion.

CASE REPORT

This report describes the case of a 16-year-old female patient, who was admitted to the emergency room of our service due to vague and recurrent abdominal pain, associated with severe pain in the right shoulder and hyporexia. Patient was evaluated by our team, who chose to hospitalize to carry out investigation. Among the imaging tests, magnetic resonance imaging of the abdomen was performed (image 1), which showed an oval hepatic subcapsular formation in segments V and VIII of the right lobe and part of segment IV a. The lesion had thin walls, irregular internal membranes and possible signs of necrosis or recent hemorrhage. The lesion measured 14x12 cm and was in close contact with the diaphragm.

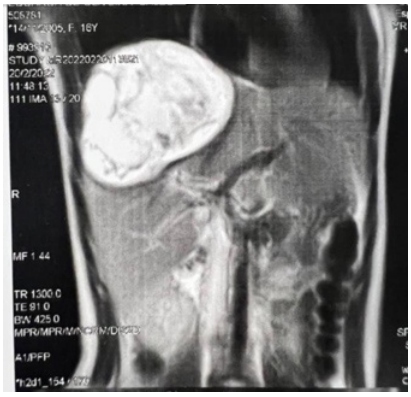

Image 1: Magnetic resonance imaging of the abdomen with contrast, coronal section

In view of this finding, it was decided to perform segmental hepatectomy surgery of segments V and VIII and part of IV, associated with exeresis of the diaphragmatic portion adhered to the lesion. The anatomopathological

and immunohistochemical study of the surgical specimen revealed a malignant mesenchymal neoplasm of indeterminate histogenesis and characteristics compatible with hepatic embryonic sarcoma, with free surgical margins. The patient had a satisfactory postoperative evolution, being discharged on the seventh postoperative day and is currently being followed up by the clinical oncology team for complementary treatment and follow-up.

DISCUSSION AND CONCLUSION

SEH is a rare malignant neoplasm with little known etiopathogenesis. It corresponds to only 7% of primary liver tumors. It must be remembered as a differential diagnosis in large liver lesions with solid-cystic characteristics, especially in the pediatric population.

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