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RESEARCH ARTICLE

ADULT HEPATIC MESENCHYMAL HAMARTOMA: A CASE REPORT.

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Abstract

The mesenchymal hamartoma of the liver is a benign hamartomatous lesion whose pathogenesis is unknown. It usually occurs in children and has been rarely reported in adults. We report an unusual case of a cystic mesenchymal hamartoma of liver in an 65-year-old elderly female. A provisional diagnosis of gastrointestinal stromal tumor (GIST) was made and definitive histopathology confirmed the diagnosis. This case has been reported because of its rarity.

Introduction:-

Mesenchymal hamartoma of the liver (MHL) is an infrequent benign tumor that affects children mostly under 5 years old and is rarely reported in adult population [1,2]. It was Maresh who first described this tumor in 1903, the term MHL was used by Edmondson in 1956 [3]. This tumor usually manifests as an abdominal mass with signs of compression of adjacent organs. In some cases, it may be incidentally discovered during a radiological investigation [1]. This tumor is frequently cystic but can be solid or mixed, the cystic nature is attributed to the cystic degeneration of myxomatous mesenchyme or cystic dilatation of malformed bile ducts [4]. Although the precise pathogenesis of MHL is largely unknown, the most common theory is that it represents the development of aberrant primitive mesenchyme in the portal tract likely relating to the bile ducts [1]. We established from immunohistochemical studies that the mesenchymal hamartoma was composed of spindle cells (positive for smooth muscle actin and vimentin, negative for CD31, CD34, and S-100) and bile ducts (positive for cytokeratin-7 (CK-7) and negative for CK-20) [5].

We report and discuss the case of a 65-years-old woman with a MHL: an unusual case.

Case Report

Mrs. E.T, a 65-year-old Moroccan woman, with no particular medical history, presented to our hospital for abdominal discomfort which has been evolving for almost a month, associated with some episodes of postprandial vomiting. The patient's physical status on admission was as follows: body temperature 37.3, blood pressure 130/75, and pulse rate 74 beats /min. physical examination indicated a tenderness of the epigastric region without palpable mass. The hernia orifices were free, the rest of the clinical examination was normal. Her laboratory investigations, including complete blood count, prothrombin time, serum bilirubin, liver enzymes and tumor markers (carcinoembryonic antigen, alpha-fetoprotein and carbohydrate antigen 19-9) were normal. The abdominal ultrasound revealed an interhepatogastric cystic mass. Contrast-enhanced computed tomography (CT) showed heterogeneous tissue mass between the liver and the small gastric curvature. , measuring 48 x 26 mm, in favor of a

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gastrointestinal stromal tumor (GIST) associated with 3 hepatic hemangiomas, the largest of which (15 mm) is at segment VIII. (figure.1). An oesogastroduodenal fibroscopy was performed, and it was normal.

On the basis of symptoms and radiological investigations, a provisional clinical diagnosis of GIST was made and we decided to perform a surgical resection of this tumor.

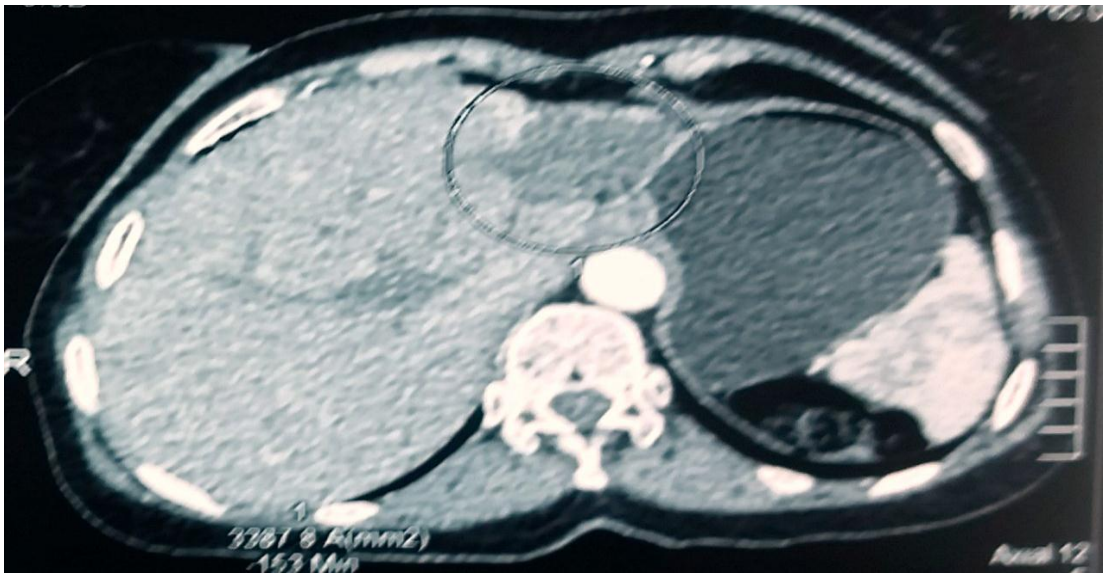

Fig 1:-Enhanced CT of the abdomen and pelvis evoking a GIST? (Mass surrounded by a circle).

Per operatively, a large lesion was seen replacing the whole left lobe and so the patient underwent resection of left lobe with drainage. Left lobectomy specimen was sent to the pathology laboratory for histopathological examination.

Gross examination revealed a large solid rounded tumor. It was well demarcated without capsule. It occupied the entire left lobe. The cut surface was yellow-white and slightly myxoid appearance with hemorrhagic and cystic areas (figure 2).

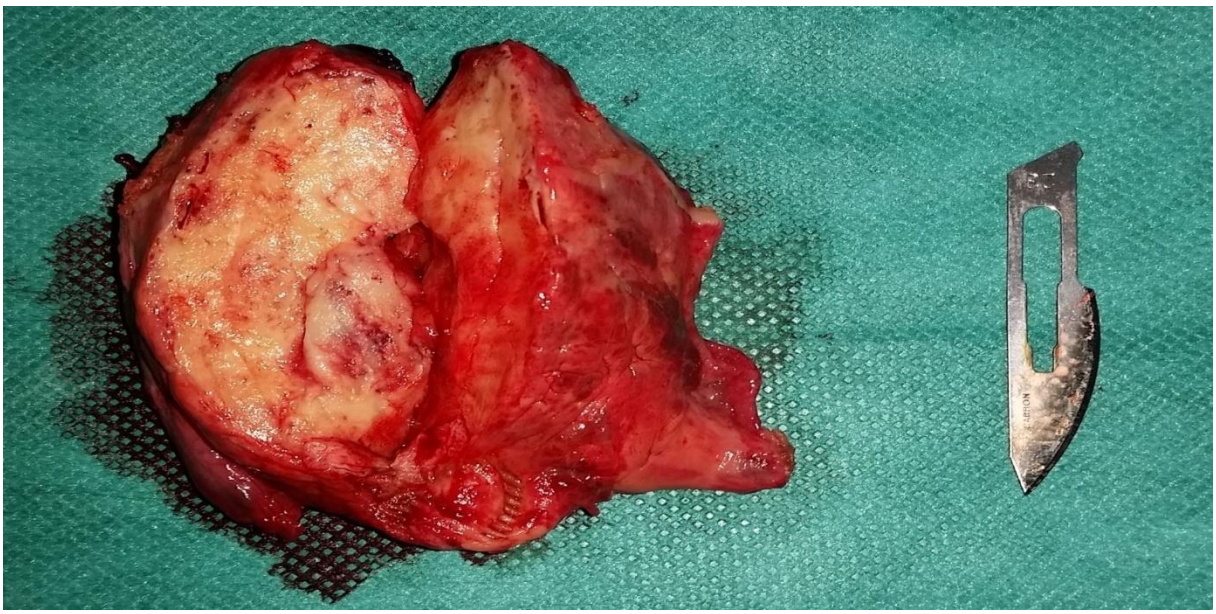

Fig 2:-the gross appearance of the cut surface of tumor.

Microscopically examination showed a mixture of epithelial and mesenchymal components. The epithelial component was represented by irregular and branching bile ducts without atypia. The ducts showing cystic change focally. The mesenchymal components is characterized by fibrous and myxoid stroma with myofibroblast-like cells, dilated vessels and mixed inflammatory infiltrate consisting of lymphocytes, plasma cells. Focally, this stroma entrapped a small area of hepatic parenchyma. (Fig.3 a, b).

The final histological diagnosis of mesenchymal hamartoma was made.

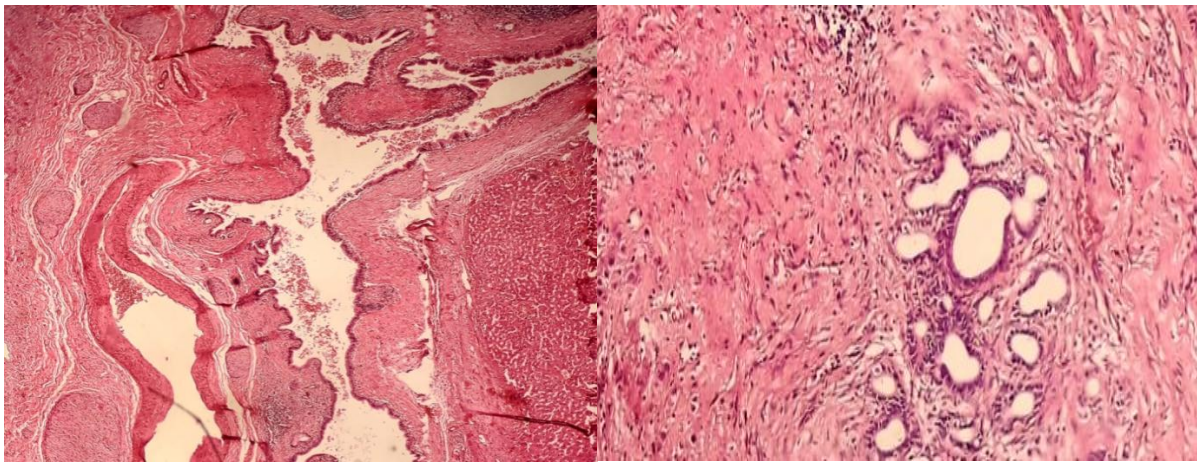

Figure 3:-histological feature of mesenchymal hamartoma of the liver showing a round, irregular branching and cystic bile ducts within a fibrous stroma. (HE : hematoxylin-eosin ; A: x100; B: x200).

Discussion:-

MHL is a benign lesion of the liver representing the second most common pediatric liver tumor (after hepatoblastoma) and constitutes 8% of all pediatric tumors. Eighty percent of the lesions are found by the age of two and the remainder are identified by five years of age with the male to female ratio being 2:1 [5].
Intrauterine MHL cases have been reported in the literature, Laberge et al. [6] reported 12 cases of fetuses with MHL, among them, 8 surviving to term and a surgical resection was performed after birth, while the remaining four cases caused intrauterine fetal death due to the mass effect of the tumor compressing the heart, kidneys or umbilical veins.

Only 32 cases of adult mesenchymal hamartoma have been previously reported as individual case reports in the English literature. [7] Of these, 23 (72%) cases have been reported in females and only 9 (28 %) in males with an average age of 40 and 59 years, respectively. in fact, males are more often affected in the pediatric population while females are more often affected in the adult population.

Location of the tumor in the liver appears to be different between pediatric and adult patients. MHL in children is primarily localized to the right lobe [8], while in adults, there seems no predilection for a particular lobe, MHL affecting both lobes of liver was seen in almost 22% of cases. In 17 cases (53,1%), gross appearance of mesenchymal hamartoma was cystic, in 9 (28,1 %) cases was solid and in 6 cases (18,.8 %) was both solid and cystic [7] .

The clinical signs of MHL seem to be influenced by the age of the patient; in children, it is often a painless abdominal mass, in some severe cases there may be ascites, jaundice or dyspnea by compression of the diaphragm and lungs [1]. In adults the symptoms of MHL include hepatomegaly, right hypochondriac or diffuse abdominal pain, in some few cases, the MHL was asymptomatic [5,9]. Ultrasonography is often the initial morphological examination for suspected hepatic masses since it is readily accessible, can quickly determine the cystic or solid nature of the lesion, and provides an estimate of mass size. CT imaging and/or MRI habitually follow ultrasonography, especially in preparation for surgical resection of the tumor. Because of the absence of specific radiological signs of MHL, morphological exploration could establish only a provisional diagnosis; it is the surgical excision of the lesion with histological examination which gives the definitive diagnosis [10].

Currently, the pathogenesis of mesenchymal hamartoma is still under study, the hypothesis of its origin from synchronous abnormal mesodermal development has been postulated. It gradually enlarges up to large sizes, some can spontaneously regress and few can undergo malignant transformation to undifferentiated (embryonal) sarcoma. Karyotype study of undifferentiated embryonal sarcoma has revealed chromosomal rearrangement in 19q13.4, which is similar to that in mesenchymal hamartoma. [11]. In addition to cytogenetic similarities a histological resemblance was established from the studies of Lauwers et al. [12] and Begueret et al [13]. This can be the probable explanation for the fact that mesenchymal hamartoma is potentially a premalignant lesion and might be a precursor lesion to undifferentiated embryonal sarcoma.

In adult, a solo case of hepatic angiosarcoma arising from MHL was recently reported by Li and al [14],we think more evidence is needed to include hepatic angiosarcoma in the malignant potential of MHL.

Surgical resection with negative margin is the standard treatment of MHL, the possibility of a huge tumor growth and the malignant potential of this lesion (embryonal sarcoma and/or angiosarcoma) constitute arguments against careful clinical monitoring particularly if the patient is a reasonable surgical candidate [15].

Conclusion:-

Mesenchymal hamartoma is a rare hepatic lesion in adults, usually presents as a cystic lesion and can be a premalignant lesion. His pathogenesis remains a controversial subject. Symptoms are typically nonspecific, laboratory results are noncontributory and radiographic imaging is variable and inconclusive. Surgical resection is the main therapeutic modality and necessary to establish a definitive diagnosis. Our case is distinguished by the fact that this tumor is extremely rare in the adult population.

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