

# EPITHELIOID HEMANGIOENDOTHELIOMA: DIAGNOSTICS AND TREATMENT, A ONE-CENTER EXPERIENCE



**Stepanova Yu.A., Aivazyán Kh.A., Shirokov V.S., Kalinini D.V., Zhao A.V.  
A.V. Vishnevsky National Medical Research Center of Surgery, Moscow, Russia**

**EPITHELIOID HEMANGIOENDOTHELIOMA (EGE)** is a rare vascular tumor originating from vascular endothelial or preendothelial cells. The frequency of occurrence is less than 1% of all vascular tumors. Due to the rare occurrence, despite the diagnostic criteria described in the literature, the probability of an erroneous diagnosis is high.

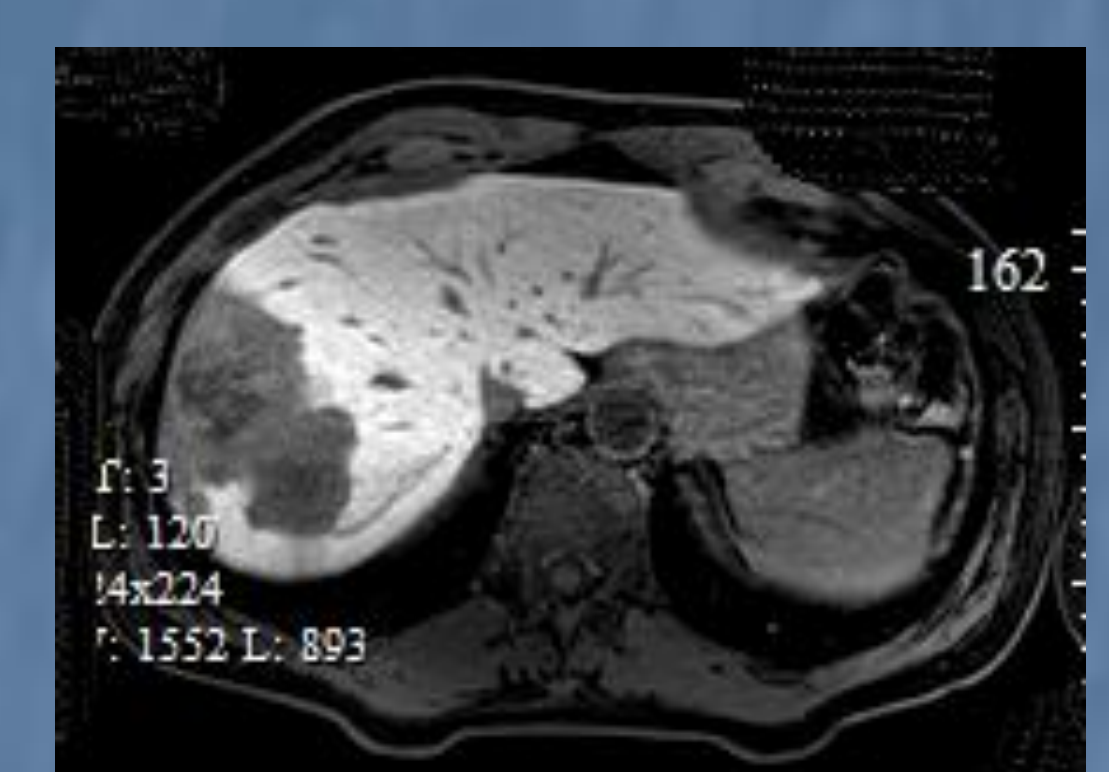
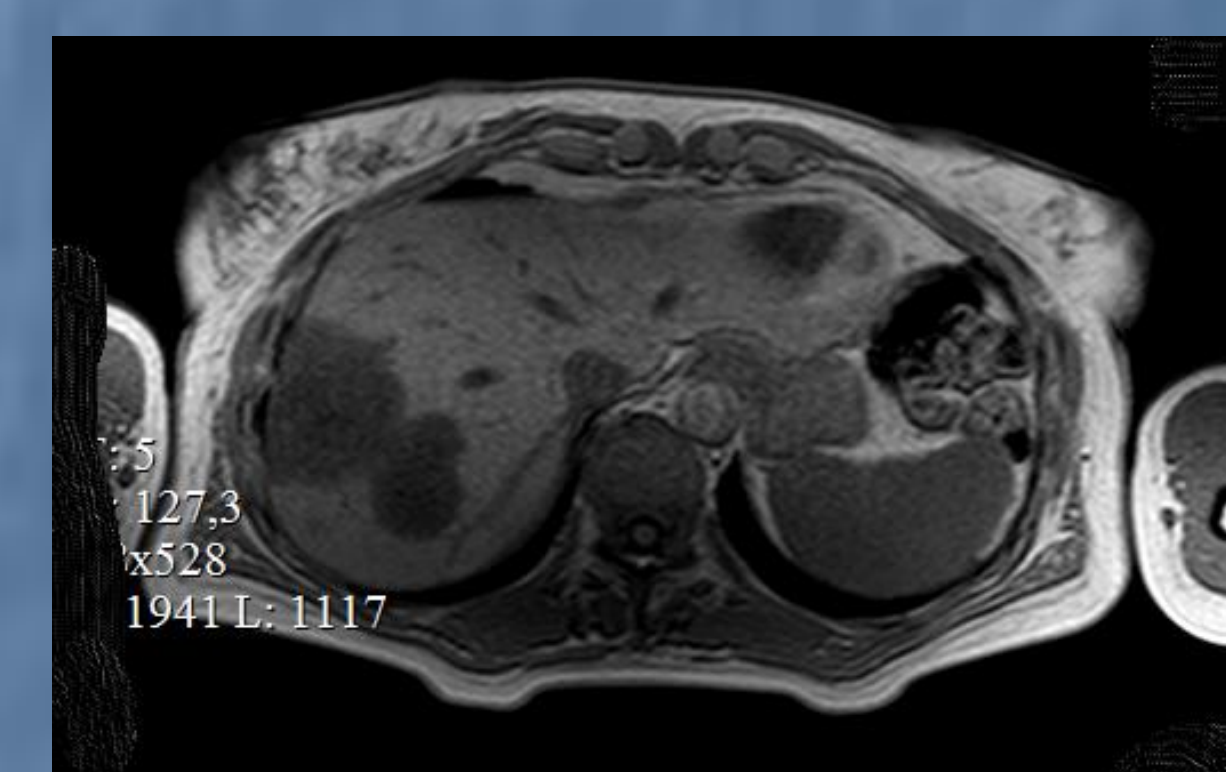
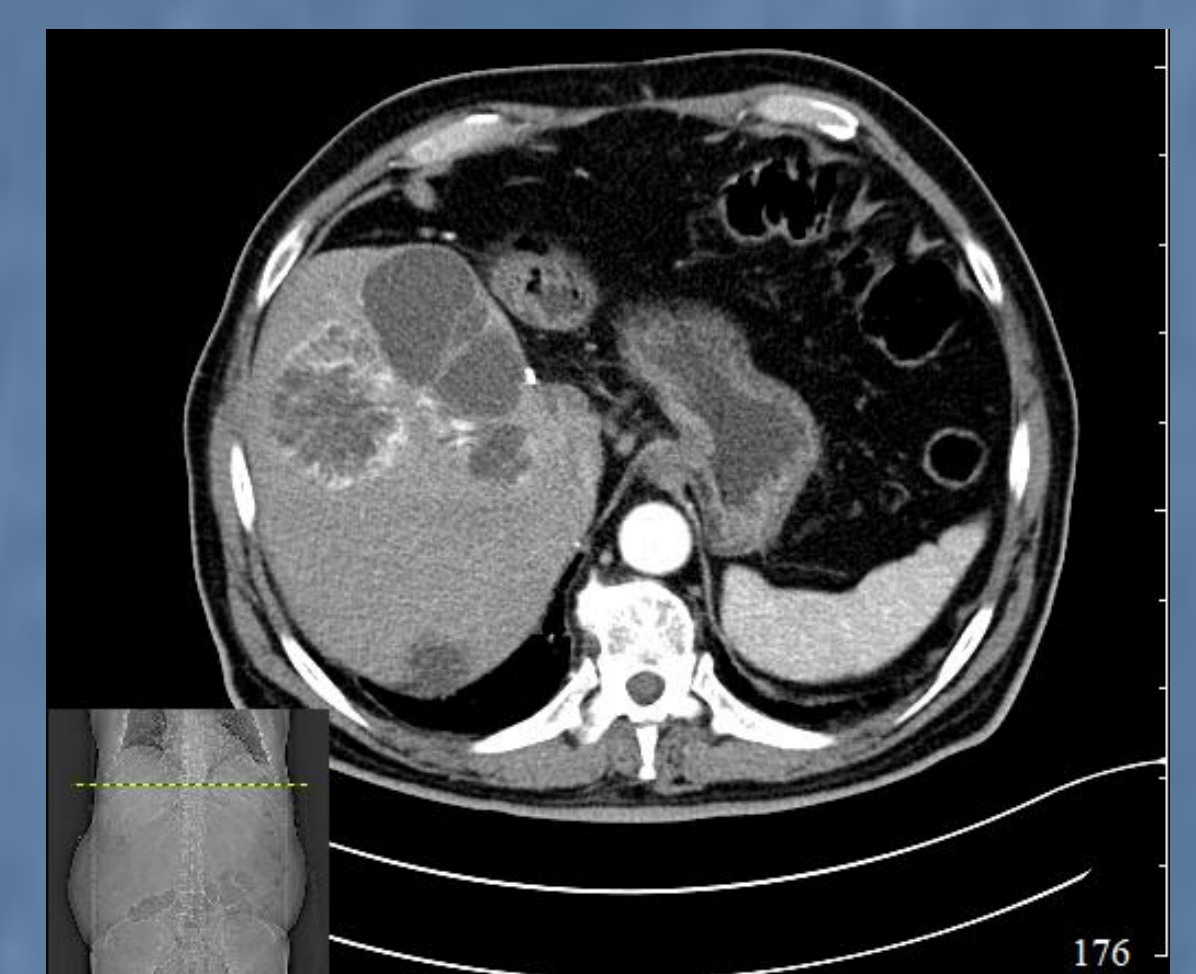
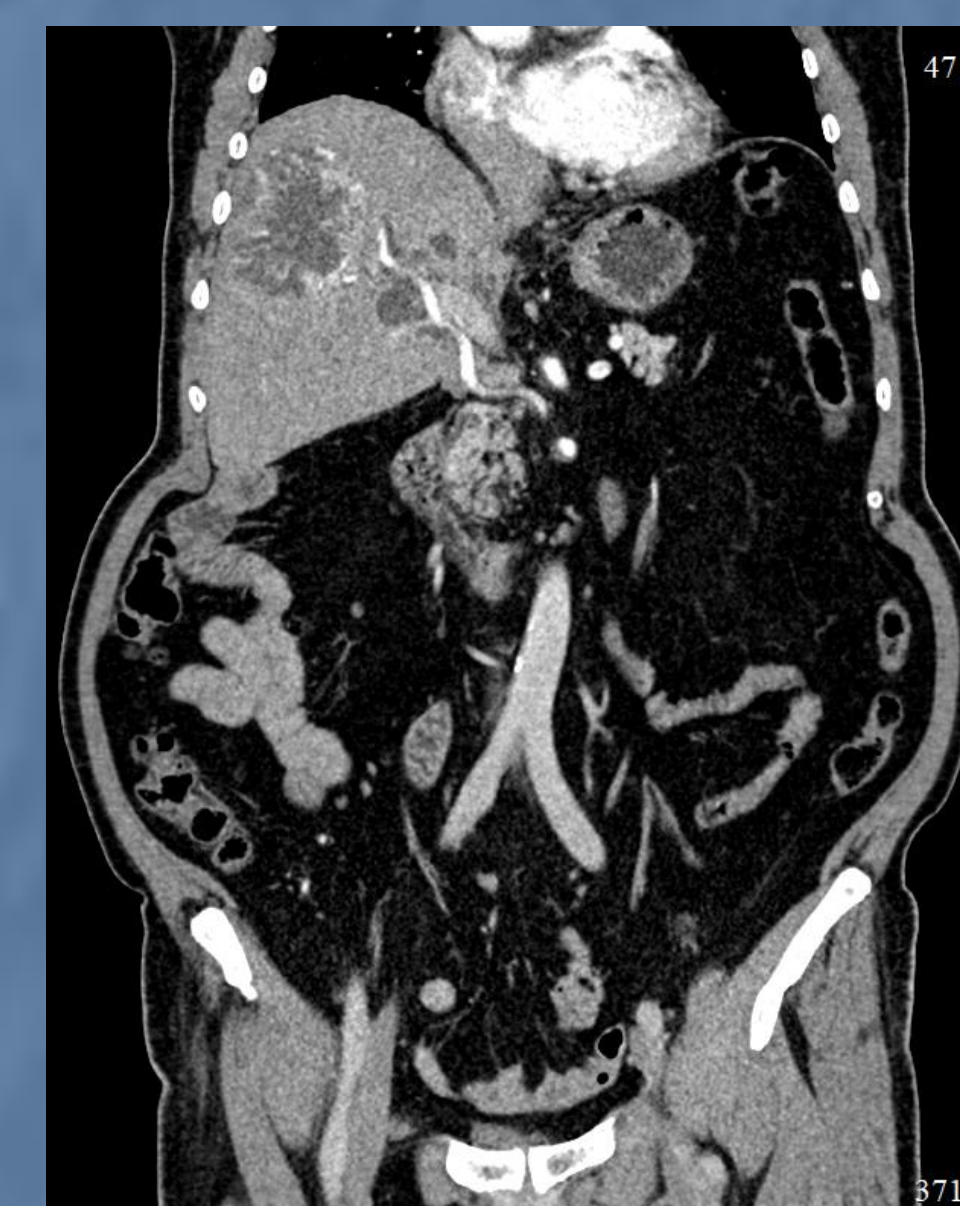
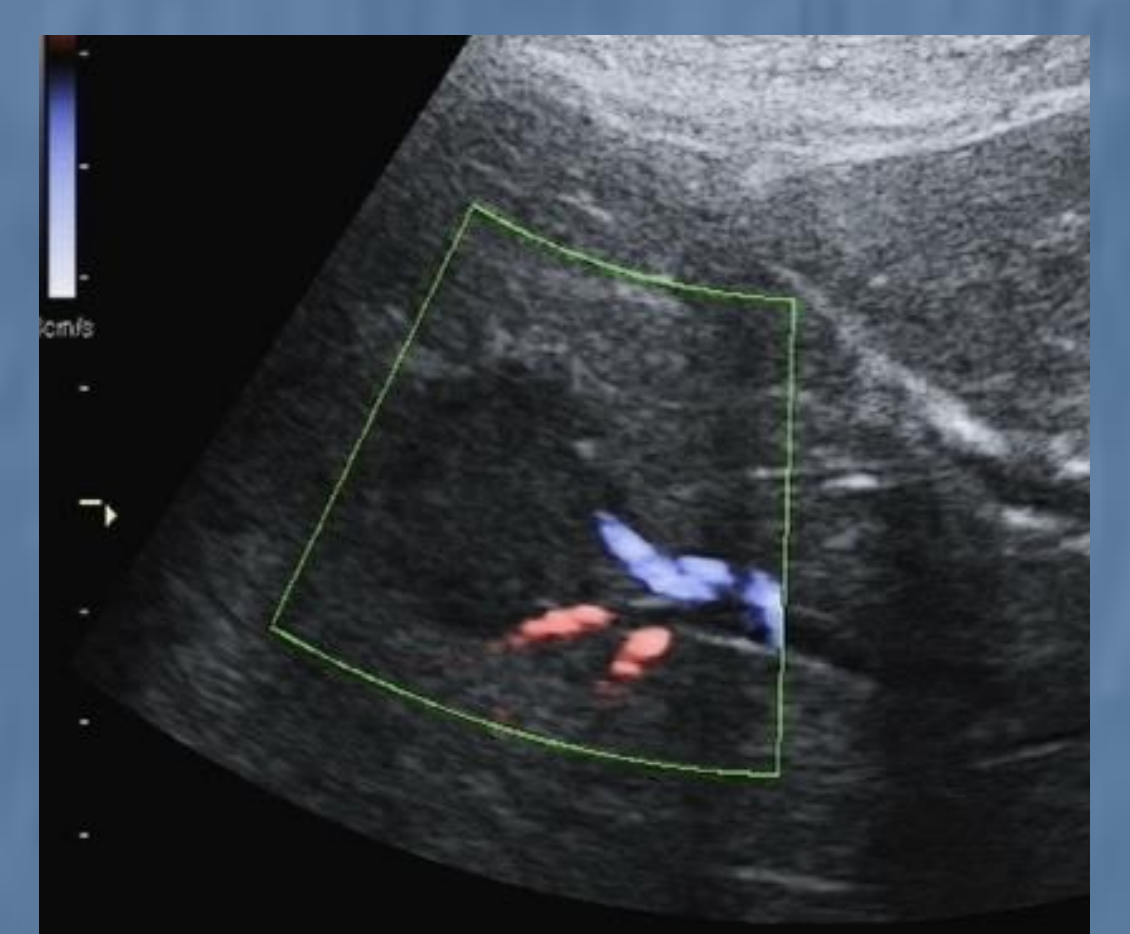
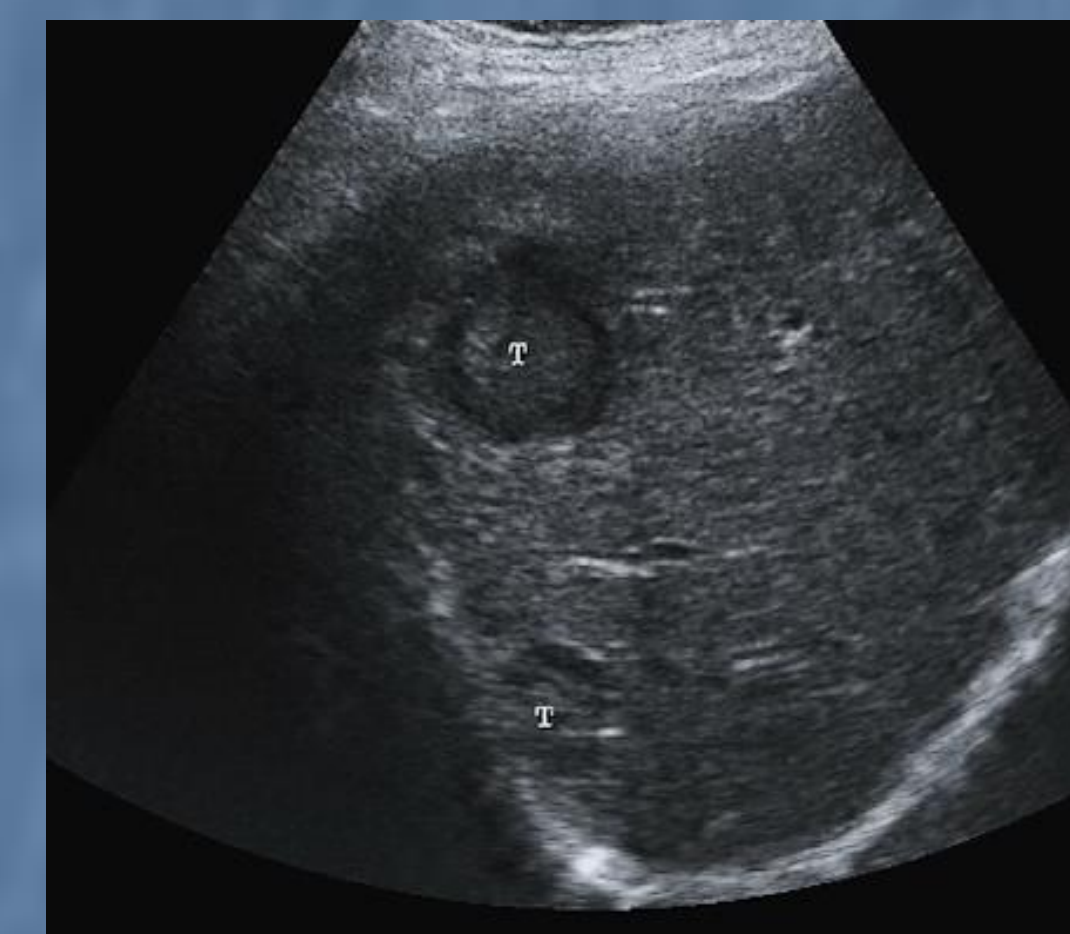
**Objective:** to evaluate the results of diagnosis and treatment of liver EGE.

**Materials and methods.** At A.V. Vishnevsky National Medical Research Center of Surgery accumulated experience in the diagnosis and treatment of **12 patients with morphologically verified EGE of the liver** (aged 21 to 70, women predominated - 9 (75%). Preoperative examination: ultrasound, MSCT and/or MRI.

## Results.

Clinical manifestations of liver EGE were nonspecific. Pain in the right upper quadrant of the anterior abdominal wall, nausea, vomiting, hepatomegaly, splenomegaly (4 cases), weight loss (3 cases), jaundice (2 cases) were noted. Approximately half of the cases were asymptomatic. The duration of clinical manifestations before diagnosis varied from 3 months to 2 years.

- **Ultrasound.** The lesions were defined as multiple peripheral compactly located (almost merging) solid nodes of reduced or mixed echogenicity with a hypoechoic rim, accompanied by capsule retraction. A dimly expressed (due to the small caliber of the vessels) peripheral vascular rim was located in large lesions at duplex scanning.
- **MSCT.** Rounded lesions, hypodense in the central part and hyper- or isodense to the liver parenchyma along the periphery (sign "target") visualized in the native phase. The detected foci differed in the nature of the accumulation of the contrast agent. More often, the lesions accumulated a contrast agent in the peripheral regions in the form of a rim or a target in the arterial phase with progression of accumulation to the portal and delayed phases. Rarely, progressive accumulation of the contrast agent throughout the volume of the lesion was noted. In 9 cases, in patients with multiple lesions, the formation of a chain of spherical growths of tumor tissue (nodules) connected by narrow (3–7 mm in diameter) threads was noted (symptom "rosary", Rozengauz E.V. et al., 2020). If this pattern was not evident in the axial section, it could be identified and followed up with multiplanar reconstruction.
- **MRI.** The foci were characterized by low signal intensity on T1 WI. There were hyperintense relative to the unchanged liver parenchyma and form a "white target" pattern: the central parts with a high-intensity signal, and the peripheral parts were slightly hyperintense on T2 WI.



The size of the lesions varied from 13 mm in diameter to a large volumetric confluent lesion. There were multiple in 10 observations. Three patients had bilobar multiple lesions. In 11 cases, retraction of the liver contour was determined at the site of the lesion localization.

Surgery was in 9 patients: hemihepatectomy - 5 (55.6%); resection of two and three segments of the liver - 4 (44.4%).

With bilobar lesions, patients undergo dynamic ultrasound monitoring within 11 to 24 months. Despite the fact that, according to the literature, EGE chemotherapy is considered ineffective, in 1 case it was decided to perform TACE (Oxaliplatin 75 mg + Lipiodol 7.5 ml). According to the results of which, a significant positive trend was noted (regression of tumors by an average of 30% of their volume).

All the patients are observed in the postoperative period from 6 months to 5 years. All patients are currently alive. 5-year cumulative survival - 100%.

**Conclusion.** EGE is a rare mesenchymal liver tumor. With an integrated approach to its diagnosis and treatment, good immediate and long-term results are possible.

