Primary undifferentiated embryonal sarcoma (UES) of the liver is a highly malignant mesenchymal origin tumor and has a peak incidence between the ages of 6 and 10 years. We hereby report a case of primary UES of the liver in a 7-year-old male patient who initially was misdiagnosed and treated as hydatid cyst of the liver. The tumor was occupying almost the entire right lobe of the liver and had a mostly cystic appearance with some solid components in it. Because hydatid disease is endemic in this region, it can often lead to misdiagnosis. The correct diagnosis was established after a biopsy and following neo-adjuvant chemotherapy the patient underwent a successful right hepatic lobectomy with complete resection of the tumor. The patient also received adjuvant chemotherapy and is currently disease-free in the present six month period. Primary UES of the liver has a predominantly solid appearance on US in contrast to its mostly cystic appearance on CT and MRI. These paradoxical imaging findings should be kept in mind in order to be able to distinguish this rare tumor from other entities, especially hydatid cyst. Thus, early diagnosis and prompt surgical resection of these tumors together with adjuvant and/or neo-adjuvant chemotherapy can provide complete remission.

Key-words: Neoplasms, in infants and children – Sarcoma.

Fig. 1. – US of the liver demonstrates a large complex cystic-solid appearance mass located in the right lobe. The color doppler reveals no flow-related enhancement in the mass.
which resulted as negative. Currently at follow-up, although the patient is still in his 6 months, there is neither recurrence nor any distant metastases on imaging studies.

**Discussion**

UES is a highly malignant primitive mesenchymal tumor that occurs predominantly in children and has a predilection for the ages of 6 to 10 years. Although it represents about 9-15% of all hepatic tumors in children, only about 150 cases have been reported in the literature (2, 3). UES is found more frequently in the right lobe of the liver (59%) than the left lobe (22%), but sometimes simultaneously in bilateral lobes (20%) (5).

Diagnosis of UES is always challenging due to the lack of specific
presenting symptoms, lack of serological markers, nonspecific findings on radiological imaging and the rarity of the disease. The histogenesis of this tumor still remains undetermined. Some authors have claimed that it is the malignant counterpart of mesenchymal hamartoma (2). Patients frequently present with abdominal pain or a palpable abdominal mass. Other symptoms are anorexia, nausea, vomiting, weight loss and jaundice. Due to these nonspecific symptoms UES sometimes can be misdiagnosed as acute appendicitis (6).

UES often show a misleading cystic appearance on CT and MRI in contrast to a predominantly solid appearance on US (7). The typical finding of UES in US is a single, large, solid echogenic mass with a cystic portion (8). CT scan typically demonstrates a large mass with cystic attenuation, while on MRI, large portions of the mass are hypointense on T1 weighted images and have a high signal intensity on T2 weighted images. With gadolinium administration, there is mild heterogenous enhancement in most of the tumor, consistent with extensive central necrosis or cystic change. This cystic appearance may lead to misdiagnosis such as hydatid cyst or amebic abscess as in our case. In the literature there have been several case reports of UES of the liver being mistaken for hydatid disease (9, 10, 11). According to Buetow et al. (7) the increased water content within the abundant myxoid stroma of UES accounts for the attenuation lower than that of soft tissue on CT scans and the high signal intensity on T2 weighted MRI, as was in our case (7). Moon et al. (8) have also described the discrepancy of internal architecture on US and CT, as an important characteristic of UES. Therefore, a diagnosis of UES is strongly suggested by the presence of a large hepatic lesion that has an apparent cystic appearance on CT and MRI but a largely solid appearance on US images. The use of delayed-phase contrast-enhanced CT and MRI is also demonstrated as useful because these techniques reveal tumoral enhancement and help to exclude purely cystic lesions in differential diagnosis (12).

It has been known that the clinical outcome of UES is poor (4). However, many authors suggest that multiagent, adjuvant or neo-adjuvant chemotherapy followed by resection offers the best long-term results and possibly a cure (13). Patients who underwent complete resection followed by adjuvant chemotherapy had significantly better survival rates compared with patients who underwent surgical resection alone. In addition, better survival was reported with intensive adjuvant therapy, even when there was residual tumor after radical resection (14). There have been three reports of liver transplantation for UES in children (15, 16, 17), however the use of liver transplantation for primary hepatic sarcoma in adults is controversial.

In conclusion, we hereby present a 7-year-old male child with UES of the liver which was initially misdiagnosed and treated as hydatid cyst. Although the differential diagnosis with imaging modalities seems to be challenging between these two entities, awareness about the paradoxical imaging findings (i.e. solid appearance on US vs. cystic appearance on CT and MRI) is crucial, because early correct diagnosis and curative surgery are mandatory for the favorable outcome of these tumors.

References