

## Malignant rhabdoid tumour of the liver: an unusual case report

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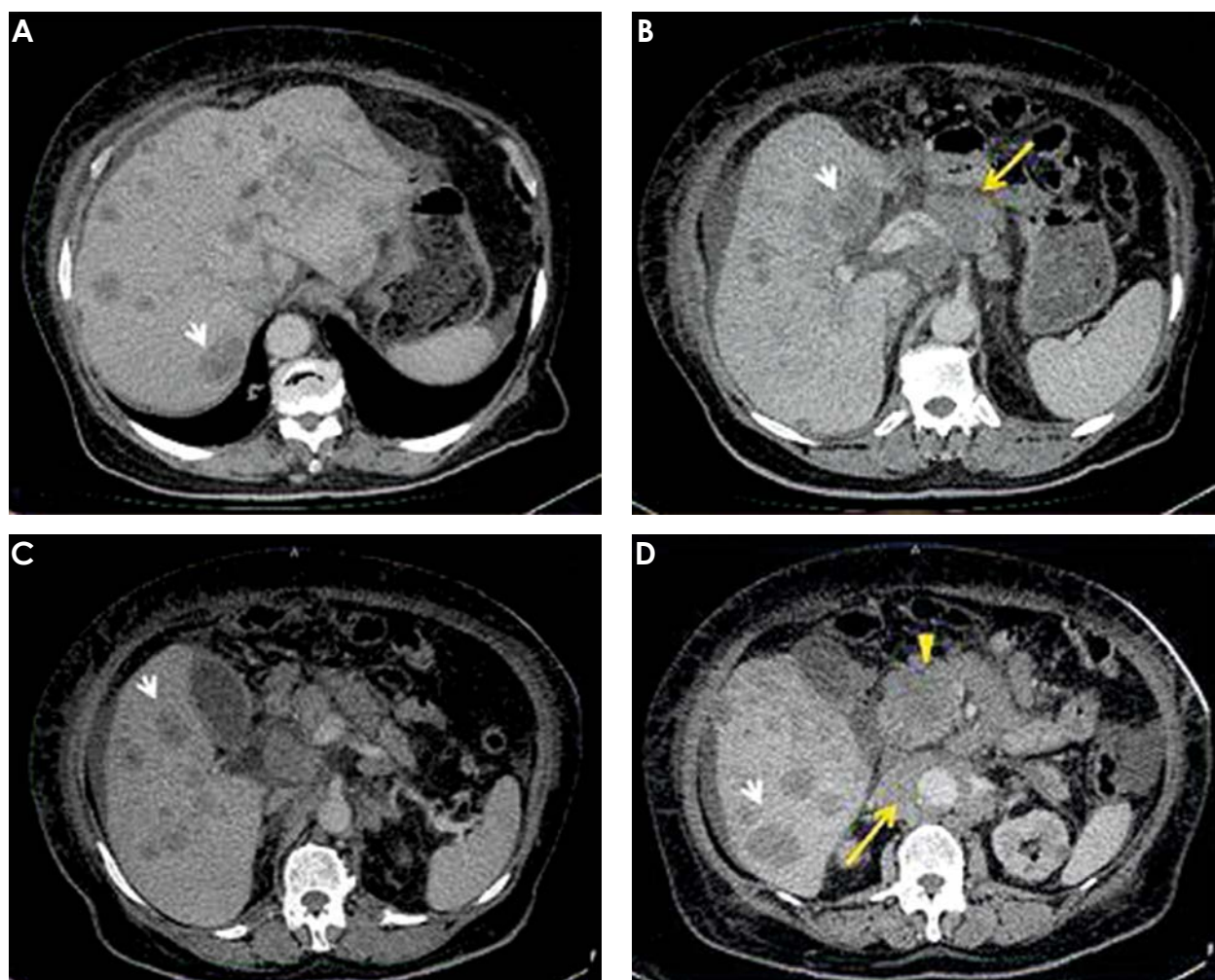
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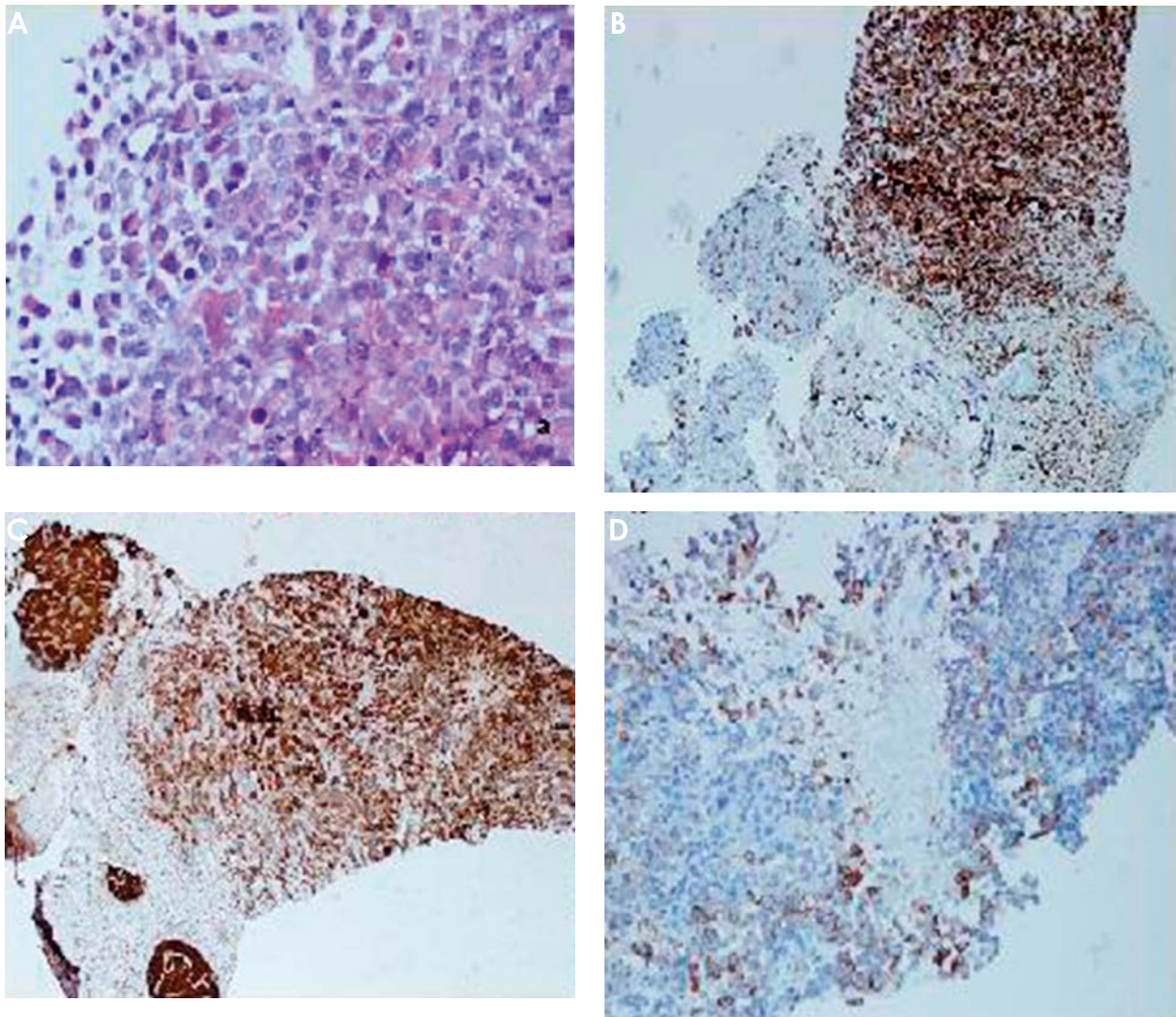
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Malignant rhabdoid tumour (MRT) is a rare and aggressive childhood neoplasm, first described as an aggressive variant of Wilms' tumour by Beckwith and Palmer in 1978 [1]. The first case, an in-

fant, with a primary liver neoplasm was described as MRT of the liver in 1982 [2]. Although MRTs are generally seen in the kidneys, primary MRT of liver, retroperitoneum, central nervous system and other



**Fig. 1.** On the axial abdominal contrast-enhanced CT sections, multiple hypodense lesions in the liver compatible with metastasis (A–D, short white arrows), celiac and paraaortic lymphadenopathies (A and D, long yellow arrows) and irregular contoured hypodense mass in the head of pancreas (D, arrowhead) are seen. There is a small amount of perihepatic and perisplenic fluid



**Fig. 2.** Prominent nucleoli and scarce vesicular nuclei containing discohesive neoplastic cells with rhabdoid morphology (A). The neoplastic cells stained Vimentin (B), pancytokeratin (C) and EMA (D) positively

sites were reported. Extra-renal malignant rhabdoid tumors are more rare tumors than the former one. Up to today less than 40 primary malignant rhabdoid tumors of liver were reported in the literature [3]. One third of the patients have high alpha-fetoprotein (AFP) levels, but two thirds of them have normal AFP levels. Although rare, it should be considered in the differential diagnosis of primary liver tumors with normal AFP.

A 67 year-old man was admitted to our clinic with a 2-month history of abdominal pain and jaundice. Physical examination showed hepatomegaly and ascites. The biochemical tests showed aspartate aminotransferase: 85 U/l, alanine aminotransferase: 57 U/l, alkaline phosphatase: 777 U/l, gamma glutamyl transferase: 1132 U/l and total bilirubin: 8.8 mg/dl. Inflammation markers were elevated (WBC: 18 110/ml and C-reactive protein: 8.75 mg/dl). An abdominal ultrasound showed the multiple heterogeneous and hypodense solid lesions of the liver parenchyma and

a percutaneous needle liver biopsy was performed. In addition to the masses in the liver and ascites, the abdominal and chest computed tomography (CT) showed a large number of lymphadenopathies on the retroperitoneal, mesenteric and celiac areas as well as a mass in the pancreas. Alpha fetoprotein was 1.5 ng/ml. An upper gastrointestinal endoscopy and a colonoscopy revealed normal findings.

On the immunohistochemical examination of the liver biopsy, the tumour cells stained positively with cytokeratin (AE1+ AE3+), vimentin, epithelial membrane antigen (EMA+); and negatively with CD34, CD99, CD117, CD37, CD45, Hep Par, cytokeratin 20, cytokeratin 7, desmin, TTF-1, glypican, S-100 and melan-a. With histopathological and immunohistochemical features, the patient diagnosed as malignant rhabdoid tumour.

During the clinical follow-up the patient was consulted to medical oncology and, we planned chemotherapy. He died on the 15<sup>th</sup> day of hospitalization

due to multiorgan dysfunction, particularly liver and respiratory failure.

Malignant rhabdoid tumour was first described as an aggressive variant of Wilms' tumour by Beckwith and Palmer in 1978 [1]. The first case of a primary liver neoplasm was described in 1982 [2]. To date, fewer than 40 cases were reported. Malignant rhabdoid tumour is a disease of pediatric population and generally originates in the kidney. Patients in the literature are usually less than 2 years of age [3]. Our patient is 67-years old, which is an advanced age for this diagnosis.

Extrarenal MRTs have poor prognosis like their renal counterparts [3]. For the treatment of MRT, systemic chemotherapy regimens are most often employed. Actinomycines, platinum-based antineoplastic agents, topoisomerase inhibitor, anthracycline antibiotics, vinca alkaloids, antimetabolites and nitrogen mustard alkylating agents can be used for chemotherapy [4]. Surgical treatment and liver transplantation are possible for a small number of patients because of frequent metastasis [5]. However, Matthew *et al.* reported that nonsurgical treatment modalities were not satisfactory and the average life expectancy was about 21 weeks. Five of 36 reported cases were still alive at the end of follow-up. Four of them were treated with surgical resection or liver transplantation and one patient had received only systematic chemotherapy [3]. We planned to administer chemotherapy, however he died on the 15<sup>th</sup> day of the diagnosis before the administration of chemotherapy.

Renal and extrarenal MRT is characterized by the presence of 'rhabdoid' cells; rhabdoid cells resemble rhabdomyoblasts, and this is the basis for the term rhabdoid tumor. Rhabdoid cells have eosinophilic perinuclear inclusion bodies, which are comprised of whorls of intermediate filaments. The most consistently positive reactions for these intermediate filaments are with antibodies to vimentin and cytokeratins (CK) [4]. Our patient's liver biopsy material from the mass lesion showed us prominent nucleoli containing discohesive neoplastic cells with malignant rhabdoid morphology. On the immunohistochemical examination of the liver biopsy, the tumour cells stained positively with cytokeratin (AE1+ AE3+), vimentin, epithelial membrane antigen (EMA+). However simultaneous pancreatic mass on the CT scan resulted in a confusion regarding the primary focus of the tumour. Since the central nervous system (CNS) scanning had not been performed, we don't know about the CNS involvement. Neither abdominal USG nor abdominal CT scan showed any mass in the kidneys, thus it's certain that this tumour is an extrarenal MRT.

In conclusion, this unusual tumour that typically affects the pediatric population should be considered in the differential diagnosis of adult liver mass-

es with no elevation of AFP. In the future, we will learn more about the treatment options of this poor prognosis disease and we will hopefully improve the survival.

## REFERENCES

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