Paraneoplastic Dermatomyositis Secondary to Hepatocellular Carcinoma in an Ectopic Liver: A Rare Case Report

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Abstract

Objective: To report a rare case of ectopic liver, complicated with hepatocellular carcinoma in a 75-year female who presented with dermatomyositis.

Case Report: A 75-year old Saudi female presented with erythema, pruritus and scaling eruptions on the face, neck, trunk and the extremities assuming violaceous color. The condition started five years earlier and was diagnosed as "photodermatosis", and was unsuccessfully treated with antihistamines and topical glucocorticoids. Three months prior to admission, she began to experience progressive proximal muscle weakness, dysphagia and severe weight loss. Neurological examination showed proximal, symmetric muscle weakness in upper and lower limbs. MRI showed inflammatory changes of the vastus lateralis muscle which was biopsied later to reveal histopathological changes consistent with dermatomyositis. Enhanced abdominal CT showed a well-defined rounded solid soft tissue mass in the subhepatic right lumbar region. A percutaneous biopsy and histopathology revealed that the patient had malignant tumor with hepatoid pattern of growth mostly in favor of hepatocellular carcinoma. The final diagnosis of our case was hepatocellular carcinoma originating from an ectopic liver tissue and associated with paraneoplastic dermatomyositis. Conservative palliative management was the only option since she was critically ill. The patient’s condition deteriorated rapidly. She developed fever with urinary tract infection and she died of septic shock three days later.

Conclusions: Physicians' awareness regarding the diagnosis of dermatomyositis should be raised. Screening for hidden malignancies is important in all cases of dermatomyositis.

Key Words: Ectopic liver – hepatocellular carcinoma – Dermatomyositis – Paraneoplastic.

Introduction

ECTOPIC liver is a rare developmental anomaly. In 1940 the incidence was about 0.23% in a series of 5500 autopsies [1]. Heterotopic liver tissue may be classified either as accessory liver, when it is connected with a thin stalk to the main liver or as a true ectopic liver when no such relationship can be established as in our case and usually show normal histological architecture.

Most ectopic livers are clinically silent. But they can suffer from recurrent abdominal pain caused by torsion or intraperitoneal bleeding [3]. Fatty infiltration and alpha-1-antitrypsin deficiency have been described in ectopic liver [2,4]. However, the development of a hepatocellular carcinoma in the ectopic liver is of utmost significance. In fact, hepatocellular carcinoma can be observed in about 30% of ectopic livers [5].

Case Report

In December 2012, a 75-year old Saudi female was admitted to the Internal Medicine Department of Aseer Central Hospital, Abha City, Saudi Arabia, with erythema, papules, pruritus and scaling eruptions on the face, neck, chest, and the extremities assuming violaceous color, which started to show up five years earlier. This heliotrope rash led to the previous diagnosis of her condition as "photodermatosis". Conservative palliative management was the only option since she was critically ill. The patient’s condition deteriorated rapidly. She developed fever with urinary tract infection and she died of septic shock three days later.

Conclusions: Physicians' awareness regarding the diagnosis of dermatomyositis should be raised. Screening for hidden malignancies is important in all cases of dermatomyositis.

Key Words: Ectopic liver – hepatocellular carcinoma – Dermatomyositis – Paraneoplastic.
Neurological examination showed proximal, symmetric muscle weakness in upper and lower limbs (grade 3 muscle strength), with preserved deep tendon reflexes, coordination, sensation and cranial nerves. The rest of physical examination was within normal range.

Laboratory findings revealed high ESR, high liver and muscle serum enzymes and anemia of chronic inflammation.

The clinical suspicion of "dermatomyositis" was strongly raised. MRI of the thigh was done and showed inflammatory changes of the vastus lateralis muscle which was biopsied later to reveal histopathological changes consistent with dermatomyositis. There was perifascicular distribution of atrophic, degenerating, and regenerating myofibers with abnormal capillary morphology and capillary loss.

Once our clinical suspicion was confirmed, an underlying neoplasm was investigated. CT chest, high digestive endoscopy and colonoscopy showed no abnormality.

Enhanced abdominal CT showed a well-defined firmly rounded solid soft tissue mass in the subhepatic right lumbar region. The lesion had homogenous texture and well-defined border. It measured 4 x 4cm in diameter. It was totally separated from the liver, bowel loops, pancreas and right kidney (Fig. 1).

A percutaneous (under radiological guidance) biopsy was immediately taken from the mass. The histopathology report revealed a tumor composed of thick trabeculae of irregular nuclei, prominent nucleoli, occasional binucleation, clumped chromatin and abundant eosinophilic cytoplasm. Vague acinar formation and brisk mitotic activity including atypical ones were also seen. The tumor cells were positive for CD10 (canalicular pattern), while negative for AFP, CK7, CK20 and vimentin and chromogranin.

So, the patient had malignant tumor with hepatoid pattern of growth mostly in favor of hepatocellular carcinoma. Another percutaneous biopsy was taken from the patient's liver which showed completely normal liver. Finally, after one week of investigations, the diagnosis of our case was settled as "hepatocellular carcinoma" originating from an ectopic liver tissue, associated with paraneoplastic dermatomyositis.

However, our patient's condition was deteriorating rapidly. She was referred to the Critical Care Department. The oncology and hepatology teams were consulted. They decided for conservative and palliative management since she was critically ill. Other options for treatment of hepatocellular carcinoma could not be suggested. However, the patient’s condition deteriorated rapidly. She developed high fever with urinary tract infection. Broad spectrum antibiotics were started, but she died of septic shock three days later.

Discussion

Dermatomyositis is an uncommon inflammatory myopathy characterized by pain and weakness in the proximal muscles and cutaneous manifestations. The association of this disease with malignancy is well known [6].

Our patient presented with heliotrope skin eruptions which started to appear five years before admission to our hospital. Her condition was misdiagnosed by dermatologists as "photodermatosis", for which she received antihistamines and topical glucocorticoids, but, her condition did not improve. It is obvious that dermatomyositis was not considered by dermatologists as a possible differential
diagnosis of the patient’s condition. Moreover, it seems that the patient chose to live with her skin condition that did not respond to treatment. So, our patient’s diagnosis remained missing for five years.

However, three months prior to admission to our hospital more serious symptoms appeared. She began to experience proximal muscle weakness and wasting, for which she sought medical consultations at several health institutions. Again, it is obvious that dermatomyositis was not considered by physicians as a differential diagnosis for the condition.

Callen and Wortmann [7] stated that Gottron’s papules and heliotrope rash have been conventionally described as highly characteristic features in cases of dermatomyositis.

Miller [8] noted that the diagnosis of dermatomyositis is based on the five Bohan and Peter criteria which include the presence of typical dermatologic features, symmetrical proximal myopathy, raised muscle enzymes, abnormal electromyogram and inflammatory myositis in muscle biopsy. The diagnosis of dermatomyositis is definite if there are presence of typical dermatologic features together with 3 or 4 other criteria. Otherwise, the diagnosis would just be probable dermatomyositis if typical dermatologic features with 2 other criteria fulfilled; and possible dermatomyositis if typical dermatologic features with another criteria fulfilled.

Once our strong clinical suspicion was directed toward dermatomyositis, MRI of the thigh was performed which revealed inflammatory changes of the vastus lateralis muscle. Muscle biopsy sowed histopathological changes consistent with dermatomyositis. So, we had to identify or exclude the presence of any underlying neoplasm.

Enhanced abdominal CT was performed to our case, which revealed the presence of a subhepatic ectopic liver at the right lumbar region. Histopathological examination of a biopsy from the ectopic liver showed that the patient had hepatocellular carcinoma.

Kubota et al. [11] stated that ectopic liver is a rare condition. Its diagnosis is quite uncommon. Lundy et al. [12] noted that the cause of ectopic liver is an aberrant migration during the embryologic development of the liver. Abnormalities of this migration or a displacement of a portion of the cranial part of the hepatic diverticulum of the liver bud to other sites may be the main cause of ectopic liver.

Catani et al. [3] added that detection of ectopic liver tissue by means of imaging studies is rare. This may be due to the small size of most ectopic livers, the lack of awareness of this unusual condition among radiologists, difficulty interpreting the imaging and the frequent lack of symptoms.

In our case, the histopathology report revealed thick trabeculae of irregular nuclei, prominent nucleoli, occasional binucleation, clumped chromatin and abundant eosinophilic cytoplasm. Vague acinar formation and brisk mitotic activity including atypical ones were also seen. The diagnosis of hepatocellular carcinoma was confirmed.

Beltran et al. [13] stated that the histological findings of ectopic liver tissue are similar to those of the liver proper, including regular lobules, a central vein, and normal portal spaces. As in the liver itself, ectopic liver can show fatty infiltration, cholestasis, hepatitis, hemosiderosis, cirrhosis, or malignant degeneration to hepatocellular carcinoma.

Griniatsos et al. [14] pointed out that there is a strong evidence that ectopic liver is at increased risk of developing hepatocellular carcinoma. Arakawa et al. [15] noted that ectopic liver tissue is more susceptible to the development of malignancy because it does not have a complete vasculature or ductal system like a normal liver, and is perhaps functionally impaired. This altered hepatic function may lead to chronic inflammation or cirrhosis, which increases the possibility of developing hepatocellular carcinoma.

Our patient died 3 days after reaching the definitive diagnosis for her condition. She was extremely unlucky to have a rare condition of dermatomyositis that has been totally missed by clinicians for 5 years. The diagnostic clinical criteria of dermatomyositis were completely overlooked by dermatologists. She also had ectopic liver which could not be identified by any physician or radiologist for 75 years, allowing hepatocellular carcinoma to develop and progress. Moreover, her definitive diagnosis was settled too late, offering her a very bad prognosis.
Tang and Thevarajah [9] emphasized that if we could early identify the presence of a specific type of cancers in cases of dermatomyositis, it would help greatly in the early diagnosis and better management of the malignancy.

In conclusion, physicians' awareness regarding the clinical picture and diagnostic criteria of dermatomyositis should be raised. Screening for hidden malignancies is important in all cases of dermatomyositis. Early diagnosis and management of dermatomyositis can make the difference in patients' prognosis.

References


