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Kidney clear cell sarcoma: About one case

Böbrek berrak hücreli sarkom: Bir olgu hakkında

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Abstract

Kidney Clear Cell Sarcoma (SRCC) is a rare malignant tumor of the child, recognized for its aggressiveness, its high bone metastatic potential and its tendency to recur. We report a case diagnosed SRCC in a 22-month-old infant with a large right lumbar mass. An observation of Kidney tumor type clear-cell sarcoma, whose diagnosis was placed on a piece of anatomo-pathology, insofar as the imaging remained uncertain as to the Kidney origin of this mass. Apart from the histological aspect, there is currently no criterion for its diagnosis. Its prognosis has been markedly improved by the introduction of new treatment regimens.

Keywords: Tumor, Kidney, Child, Sarcoma, Clear cells

Öz

Böbrek Berrak Hücreli Sarkom (SRCC), saldırganlığı, yüksek kemik metastatik potansiyeli ve tekrarlama eğilimi nedeniyle tanınan, çocuğun nadir görülen malign tümörüdür. Büyük bir sağ lomber kitle olan 22 aylık bir bebekte SRCC tanısı alan bir olgu sunuyoruz. Bu kitlenin tanısı bir anatomi-patoloji üzerine yerleştirilmiş şeffaf hücreli sarkomu olup, böbrek kitlesinin tanısı böbrek kanseri ile ilgili belirsiz kalmıştır. Histolojik özellik dışında, şu anda tanısı için bir kriter bulunmamaktadır. Yeni tedavi rejimlerinin uygulamaya konması ile prognozu belirgin bir şekilde artmıştır.

Anahtar kelimeler: Tümör, Böbrek, Çocuk, Sarkoma, Berrak hücreler

Introduction

Kidney clear cell sarcoma is a rare tumor (4% of Kidney tumors) in children [1]. It is one of the most frequent aggressive Kidney tumors, said to have adverse histology, belonging to the group of non-Wilms kidney tumors [1,2].

Case presentation

We report a case of SRCC diagnosed in a 22-month-old infant with a large right lumbar spine. Computed tomography (CT) showed one of a large right mediastinal mass, well limited, hypodense, heterogeneously enhanced after contrast laminating the renal parenchyma, measuring 11x8.5x12 cm. This mass represses and compresses the ipsilateral renal pedicle, the inferior vena cava (IVC) and comes into contact with the abdominal aorta on a <90 ° surface. It also displaces the digestive structure, the diaphragmatic pillar and the visceral face of the right liver with loss of greasy edema of separation, with no sign of invasion. It comes into contact with the lateral abdominal wall without evidence of invasion (Figure 1).

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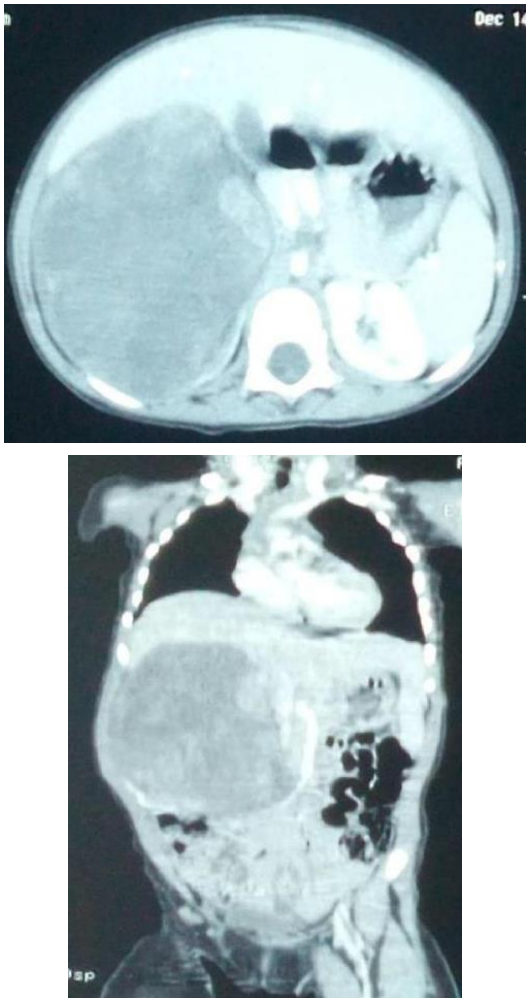

Figure 1: Computed tomography appearance of a bulky abdominal mass, un-encapsulated, heterogeneous density and whose relationships with the kidney are imprecise

Pulmonary, cerebral and bone extension assessment did not indicate secondary localization. Preoperative chemotherapy was administered according to GFA-Nephro 2005, with no toxicity 0. At the end of preoperative chemotherapy, abdominopelvic CT showed no reduction in tumor volume. Surgery consisted of an extended right anterior trans-peritoneal nephro-uretectomy with lymph node dissection was performed. The operative piece measured 15x10x 9cm. The ureter measures 3x0.5 cm. At the opening, the entire kidney is occupied by a beige tumor, homogeneous, massively infiltrating the kidney and having cystic areas. Histologically, there was tumor proliferation arranged in layers. It is made of cells sometimes rounded, sometimes oval or fusiform. The nuclei are small, with fine chromatin. These cells are located on an abundant myxoid background with formation of cystic foci bordered by tumor cells. The tumor stroma has vessels and fine-walled capillaries, absence of anaplasia, heterologous component or nephrogenic residue with negative cleansing (latero-cellar + inter-aortic-cellar). Tumor cells diffuse vimentin and cyclin D1 and do not express BCL2 or desmin (Figure 2). Histological and immune-histochemical appearance compatible with a clear cell sarcoma, classified according to the SIOP (International Society of Pediatric Oncology) 2001 high risk, stage II.

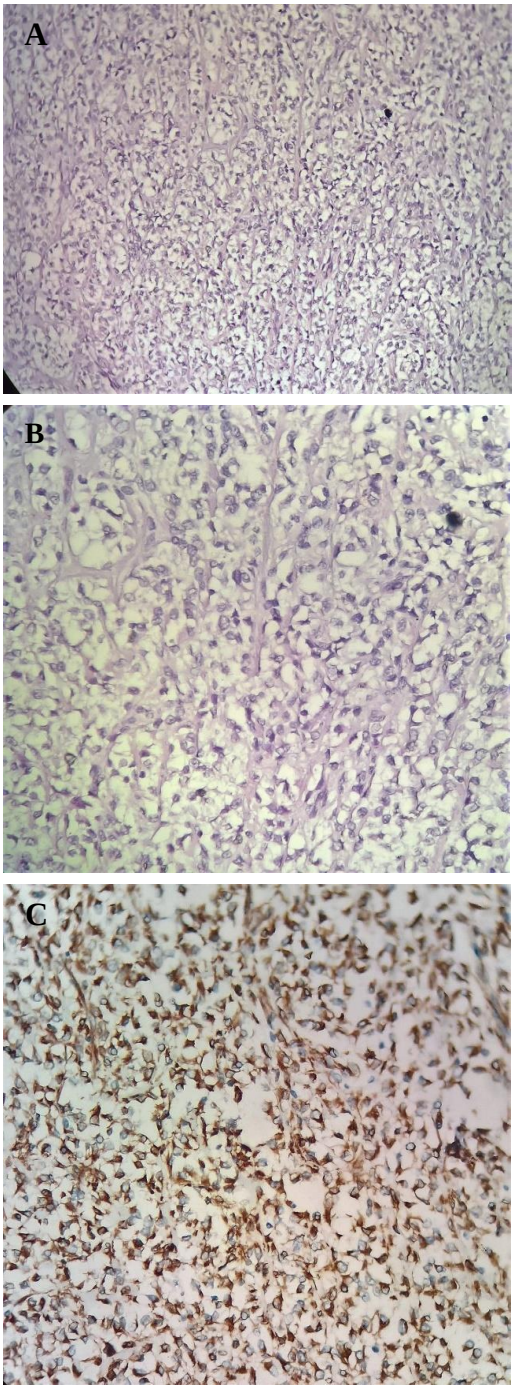

Figure 2: A: HES X 10: Tumor proliferation arranged in diffuse layer, made of clear cells, B: HESX400: tumor cells are round or oval, with clarified nuclei and clear cytoplasm; Note the presence of a fine vascularization connected, C: tumor cells strongly express vimentin

In accordance with the therapeutic recommendations developed by the SIOP 2001 Committee, prolonged postoperative chemotherapy according to GFA-nephro 2005 post op. The tumor was classified as high-risk stage II R0, so the action to be taken was the irradiation of the right flank of 10.8 Gy in 6 fractions of 1.8Gy / Fraction, without boost, at the end of the radiotherapy, we do not have toxicity. Currently, the child is in complete remission more than 8 months from the diagnosis.

Discussion

Kidney clear cell sarcoma accounts for about 4% of childhood kidney tumors and usually occurs in children aged 18 months to five years (average 30 months). Because of the age of their occurrence, the absence of specific imaging and the histological similarity of certain variants, these tumors can pose

diagnostic difficulties. Indeed, the Kidney clear cell sarcoma is a rare tumor of the child and exceptional in the adult [3]. The abdominal mass sums up the whole clinical history and is the essential reason for consultation. Associated abdominal ultrasonography and / or abdominal CT scan alone usually provide the diagnosis of renal tumor [4].

Histologically, it is a tumor with an in fi ltering tendency, unlike nephroblastoma, which is encapsulated. In the classical form, it is a homogeneous tumor and in 90% of cases, it is a richly vascularized proliferation, arranged in nests or cords. The cells are fusiform or oval, with a clear cytoplasm and nucleus. A small fibro-vascular stroma typically lobulates this tumor. The presence of tubes and glomeruli enclosed within the tumor is frequently observed [5,6]. Our case is consistent with the literature data.

Immunohistochemistry is not specific. Only vimentin is positive. The expression of Bcl-2 is not constant, that of epithelial markers is only objectified at the renal tubes engulfed in proliferation. In our observation, cyclin D1 was also expressed [7]. The differential diagnosis of Kidney clear cell sarcoma can in children discuss a nephroblastoma in its sarcomatous or pure blastematous variety, a mesoblastic nephrome in its cellular form, a rhabdoid tumor and an embryonic rhabdomyosarcoma, some histological, immunohistochemical and molecular elements allow their distinction [5,7].

Rare cases of reported SRCC have t (10; 17) translocation and / or 14q deletion. No specific genetic criterion for Kidney clear cell sarcoma is currently known. However, the study of Huang et al [8]. Demonstrates that these four tumors have different genetic profiles.

To date, few studies have focused on therapeutic management. The current treatment, according to the SIOP 2001 protocol, recommends extensive nephrectomy, radiotherapy and intensive and prolonged multidrug therapy based on alkylants, anthracyclines, epipodophyllotoxins and platinum derivatives. National Wilms' Tumour Study 1, 2, 3 and 4 studies [9,10] show that the addition of doxorubicin (D) improves prognosis with a six-year relapse-free survival of 63.4% versus 25% without (D). In the same way, a multidrug therapy prolonged for 15 months offers a better survival at eight years, namely 87% versus 60.6% for a period of six months.

The utility of radiotherapy in intermediate-risk tumors is more debated, with Europeans generally favoring a chemotherapeutic approach, while North Americans are more inclined to associate a radio-therapeutic approach. High-risk tumors, including clear-cell sarcomas, are treated by the combination of radiotherapy and additional cytostatics such as carboplatin, etoposide and cyclophosphamide. The total duration of treatment, administered mostly outpatient, varies according to stages and classification of the tumor between one and eight months. Close monitoring is necessary given the tendency to recur and metastases, mainly bone [9,10].

In conclusion, kidney clear cell sarcoma is a rare, highly aggressive and often histologically discovered pediatric tumor that is considered to be a high-risk tumor. Such progress has been made over the years through improved combined management, medical-surgical, and better stratification of less-risk tumors, leading to better selection of required treatment intensity.

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