Renal primary angiosarcoma


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Abstract Angiosarcomas account for 2% of all soft tissue sarcomas and of them, primary renal angiosarcomas represent 1%. Twenty-four cases have been published in the English specialised literature. We report the second case to be described in a middle-aged female, with pulmonary metastases at diagnosis, and fatal outcome despite surgery and chemotherapy.

Key words Angiosarcoma • Kidney


Introduction

Angiosarcomas are rare malignant neoplasms that originate in the endothelial cells of blood and lymphatic vessels that account for less than 2% of all soft tissue sarcomas [1].

Visceral sarcomas are less common than sarcomas of the skin and soft tissues (57%) and kidney involvement is generally metastatic. Twenty-four cases of primary renal angiosarcoma have been reported in the English specialised medical literature since it was first described in 1942 [2], accounting for 1% of all angiosarcomas.

Primary renal angiosarcomas are more frequent in white males, between 60 and 70 years of age. It most commonly presents as macroscopic haematuria (81%), pain in the flank (38%) or a palpable kidney mass (31%) [3]. Diagnostic suspicion is based on radiological findings and confirmation is histological. There is no standard of care and prognosis is poor, with a median survival time of 8 months following diagnosis; the major prognostic factors are tumour size and the presence or absence of metastasis.

We present the case of a 42-year-old female who debuted with haemoptoic expectoration due to pulmonary metastases. She was treated with palliative surgery and chemotherapy and died 5 months after diagnosis.

The peculiarities of the case are analysed and a comparative review is made of other cases described in the literature.

Case report

The patient was a 42-year-old female, professional hairdresser. She was a non-smoker and non-drinker with no personal or family history of interest or exposure to environmental pollutants.

The patient presented at the Emergency Department due to constitutional syndrome with 2 months of evolution and haemoptoic expectoration. Physical examination of the abdomen revealed slight tenderness in the left paraumbilical region; all remaining findings were strictly normal. Routine blood work revealed hypochromic microcytic anaemia (Hb 9.8 g/dl, Htc 29%). All the other analyses performed were normal, including platelet count, coagulation, tumour markers (CA 19.9, CEA, CA 15.3, CA 12.5) and urinalysis.

A simple X-ray of the chest and abdomen was obtained (Fig. 1) that revealed multiple nodular lesions in the lung and a mass in the left half of the abdomen, respectively. The computerised axial tomography (CT) of the chest, abdomen and pelvis confirmed the presence of multiple, bilateral pulmonary lesions measuring approximately 1 cm, predominantly bilateral basal, with areas of ground-glass (Fig. 2), as well as an 11-cm mass in the kidney with a hypodense centre that displaced the hilar vessels, splenic artery and the pancreas.

Respiratory function tests were performed with results within normal and a bronchoscopic examination was carried out with bronchoalveolar lavage, aspiration and brushing that failed to reveal any evidence of malignancy.
The decision was made to proceed with surgery of the primary tumour based on the suspicion of metastatic renal carcinoma.

During surgery and upon gross examination, a mass was observed that involved the left kidney and adrenal gland with areas of necrosis. A radical left nephrectomy was performed and the pathology study described a kidney weighing 577 g, measuring 16x10x8 cm, with a 9-cm mass located on the superior pole. This mass was protuberant and a destructured 3x2 cm reddish-violet area was seen in the interior. The cut surface revealed heterogeneous tissue with a large necrotic area and cavitation in the centre, which spared the renal pelvis and hilar vessels, made up of epithelioid cells, with spindle-shaped areas lining the variably defined vascular channels; large hemorrhagic areas, occasional necrosis and 10 mytoses per 10 high-powered fields were also revealed (Fig. 3). These cells stained highly positive for CD-31 and CD-34 antibodies, which led to a definitive diagnosis of primary renal angiosarcoma, involving the superior pole of the left kidney and left adrenal gland.

Prior to initiating treatment with chemotherapy, the patient presented several episodes of symptomatic anaemia that required several transfusions of packed red blood cells. It was related to probable pleural bleeding due to the pulmonary metastases following the chest CT where extravasation of contrast was seen at that level and embolisation was ruled out in light of the scant probability of arterial bleeding. For this reason and given the poor Performance Status (PS), this patient was not eligible to participate in a clinical trial and first-line palliative chemotherapy was started with adriamycin (75 mg/m2: 115 mg) every 21 days, which was well tolerated except for grade III neutropenia subsequent to the second cycle.

The re-evaluation studies carried out after 2 cycles of treatment showed local-regional and pleuropulmonary progression of the disease; that line of treatment was therefore terminated and, pending the initiation of second-line treatment with weekly paclitaxel as the procedure to be followed for compassionate use, the patient was admitted for acute respiratory failure and died 5 months following diagnosis.

Discussion

In this text we review the 24 cases of primary renal angiosarcoma published in the English specialised medical literature from 1942 until 2006, which bears out just how uncommon this kind of malignant neoplasm is and even more so, its appearance in the kidney as the primary tumour site (Table 1).

Our case presents certain unique features, such as the fact that the patient was female [4] and younger than the usual median age (60–70 years). At present, the greater prevalence of this tumour in males has not been explained, albeit it may be due to the role of androgens, smoking or occupational factors [5].