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Primary peripheral neuroectodermal tumor or Ewing's sarcoma of the kidney: A rare case and review of the literature

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ABSTRACT

Peripheral neuroectodermal tumors (PNET), or Ewing sarcoma, are tumors that generally develop in bone; extraskeletal locations are rare. Renal PNETs are rare and are characterized by an aggressive clinical course and a poor prognosis. We report the case of a young patient who presented with abdominal and lumbar pain with a palpable abdominal mass. The imaging was in favor of a huge renal tumor, and the histological analysis allowed the diagnosis of a renal PNET. The therapeutic attitude was multimodal, including surgery and chemotherapy, allowing complete remission and a favorable outcome.

1. Introduction

Primary neuroectodermal tumors belong to the Ewing family of sarcomatous tumors, which all derive from the same stem cell.[1] Neural crest cells are suspected of being the cause of these tumors.[1] The predilection sites for these sarcomas are: the paravertebral region, the chest wall and the distal extremities.[1] The localization of PNET in the genitourinary system such as kidneys, bladder and prostate is rare.[2] Renal PNET is extremely rare and presents a very aggressive biological behavior with a poor prognosis.[2] Renal PNET is exceptional, with fewer than 100 cases reported.[1] The diagnosis of renal PNET is mainly based on histopathology and immunohistochemistry.[1] Given their rarity, the treatment of renal PNET is not codified in the literature.[1] The evolution of cases described in the literature is unfavorable with limited survival.[2] We present a new case of renal PNET in a young patient showing a favorable response to multimodal treatment including extensive surgery and chemotherapy.

2. Case presentation

We present the case of a 47-year-old patient who consulted for abdominal pain and left lower back pain for 1 month. On clinical examination, an abdominal mass with deformation of the abdomen is discovered. The CT scan showed a left renal mass measuring 20 × 12 cm with double components, both solid and cystic, and the tumor was heterogeneously enhanced with the presence of areas of necrosis (Fig. 1). The biological assessment was without notable abnormalities.

A bone scan was performed to eliminate a primary bone origin, which returned without abnormalities. The tumor extension assessment included a thoraco-abdomino pelvic CT scan, which did not reveal any secondary localization. Faced with these data, an extended total nephrectomy was performed without notable incident (Fig. 2).

Macroscopic examination showed the presence of a lesion with dual solid and cystic contents, fleshy in appearance, brownish heterogeneous, encapsulated, and without capsular rupture with non-invaded surgical edges. Histological analysis showed poorly differentiated tumor proliferation with a solid architecture. The tumor cells were poorly differentiated with abundant basophilic cytoplasm and equipped with elongated nuclei. Immunohistochemically, the tumor cells expressed high expression of CD99, CD56, and vimentin (clone V9), thus confirming the diagnosis of renal PNET (Fig. 3).

The patient benefited from an adjuvant chemotherapy protocol based on vincristine, doxorubicin, and cyclophosphamide, with survival without radiological recurrence for 2 years after diagnosis.

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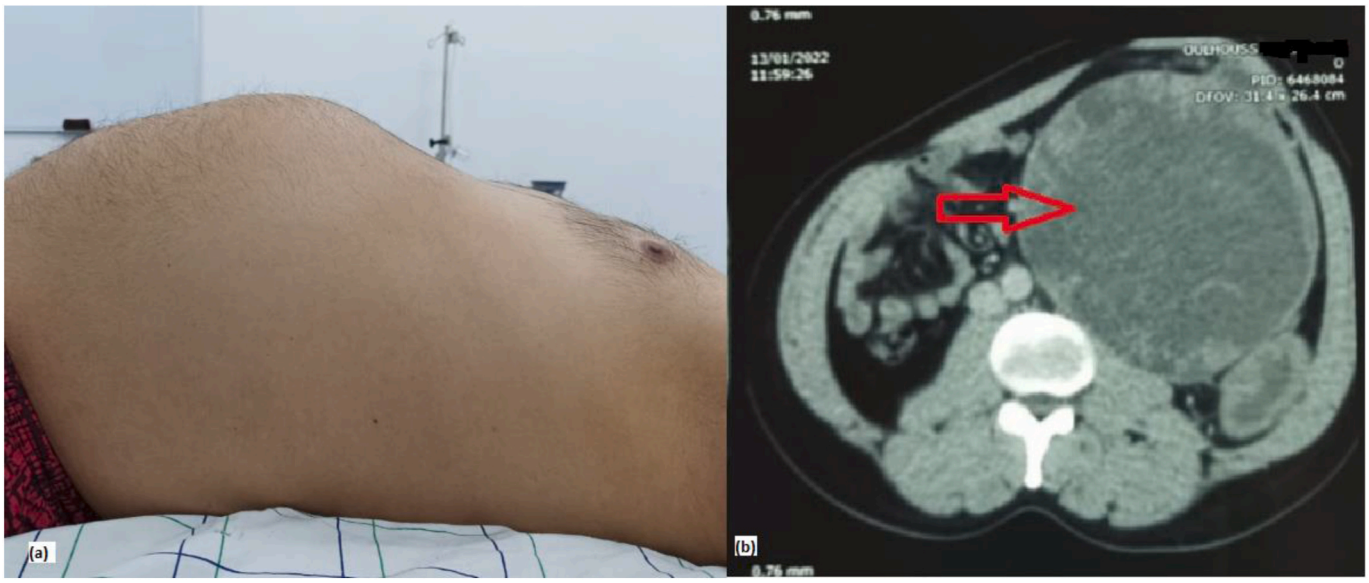

Fig. 1. (a): a visible mass with deformity of the abdomen (b): the scanner reveals a large heterogeneous mass with areas of necrosis.

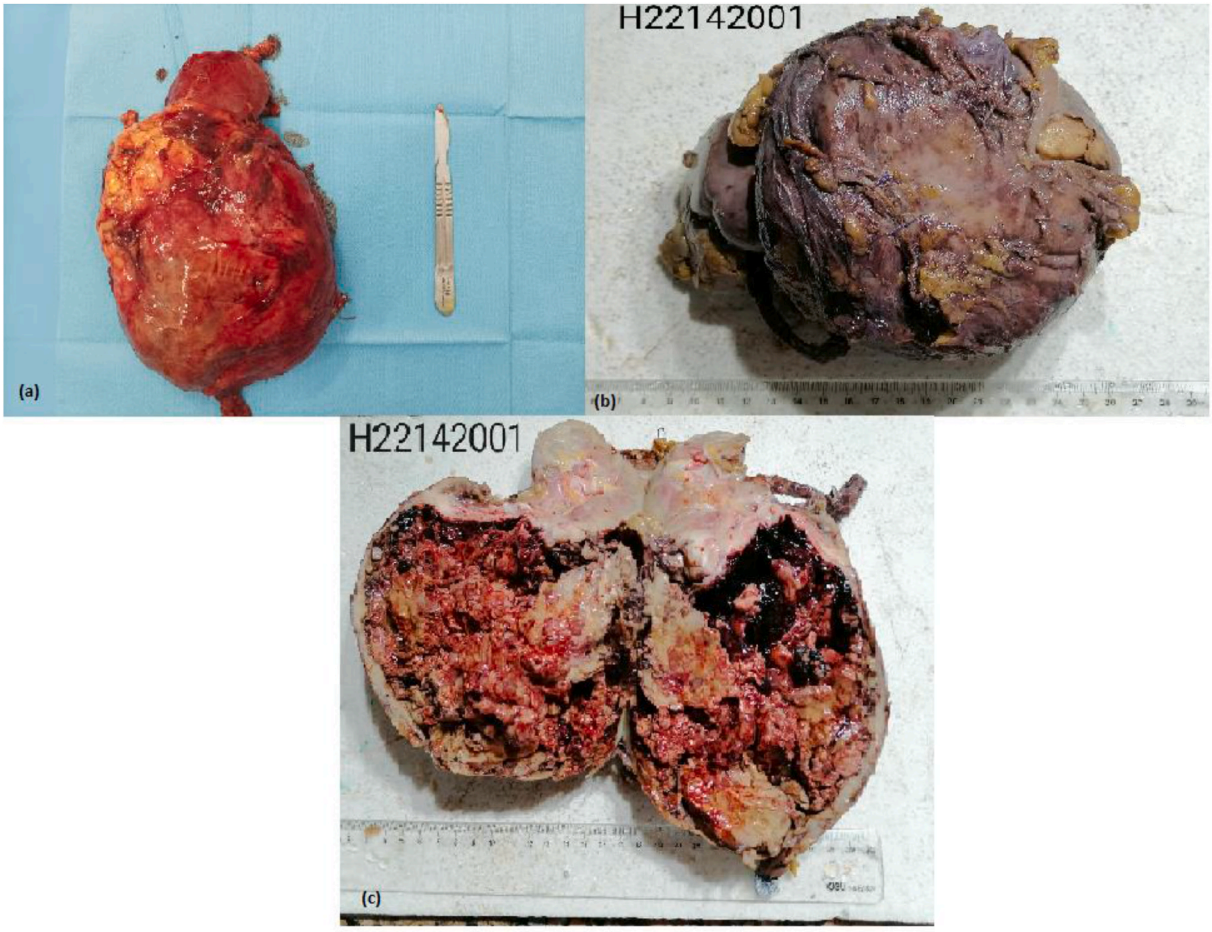

Fig. 2. (a) (b): the macroscopic appearance of the mass after extraction (c): the macroscopic appearance of the mass after opening.

3. Discussion

Primary renal neuroectodermal tumors were described for the first time by STOUT in 1918 and are known in the literature under different names: primitive neuroectodermal tumor, malignant neuroepithelioma, peripheral neuroblastoma, primitive neuroectodermal tumor of soft tissues, or peripheral neuroectodermal sarcoma of the soft tissues.[1] Renal PNETs constitute a rare primary visceral localization; few cases are reported in the literature. The authors agree on the aggressive potential of these tumors, which usually metastasize to lymph nodes, liver,

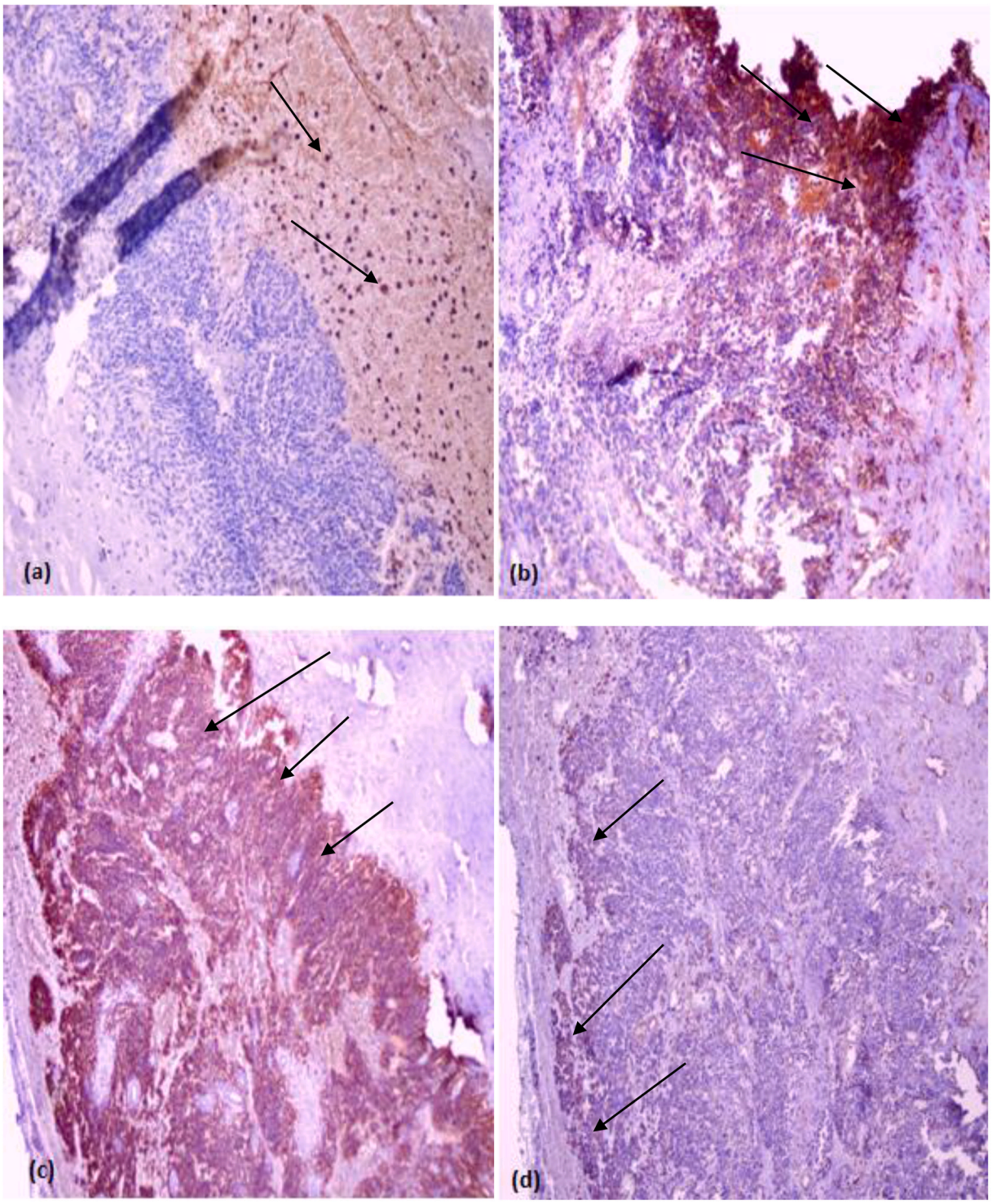

Fig. 3. (a): microscopic appearance: shows hyperchromatic, poorly differentiated tumor proliferation with irregular nucleolated edges with multiple mitotic images; (b): IHC: Anti-CD99 antibody (clone 12 E 7, DAKO): positive with fairly diffuse membrane and cytoplasmic marking; (C): IHC: Anti-CD56 antibody: with fairly diffuse membrane and cytoplasmic marking; (D): IHC: anti-Vimentin antibody (clone v9): positive with heterogeneous marking.

kidney, and bone.[1]

The average age of onset of renal PNET is 32.7 years (7–62 years). There is no sex predominance.[1] The clinical expression of renal PNETs is not pathognomonic.[2] The diagnosis time is relatively short, reflecting its aggressiveness. Urinary signs such as low back pain, renal colic, and palpable mass are their main manifestations; hematuria and deterioration of general condition reflect an advanced stage of the disease and indicate a poor prognosis.[2] The radiological assessment must include a CT scan, which often shows a mass syndrome with areas of necrosis or hemorrhage.[2] The diagnosis of renal PNET is based on a pathological examination. Macroscopically, the tumor is rounded, oval, or multi-nodular, well defined, and without encapsulation. When cut, it is gray-beige or yellow, and its consistency is soft or crumbly. Necrotic or hemorrhagic changes are common.[2] Calcifications are sometimes observed.[2] Immunohistochemically, renal PNET is most often positive for CD99, an essential element for the diagnosis. Also, almost two-thirds

of the expression of Fli-1 was observed.[3] Other markers such as vimentin, cytokeratin, NSE, and S-100 have also been detected.[3]

Treatment strategies for renal PNET are multimodal, including surgery, adjuvant chemotherapy, and radiotherapy.[4] The often late diagnosis of these tumors explains the frequency of locally advanced cases where cancer surgery cannot be conceived, and the use of other therapeutic means finds its place, in particular chemotherapy, which makes it possible to optimize local treatment and control metastatic disease via its systemic action.[4] In the absence of therapeutic standards, there are multiple cytotoxic drugs active on renal PNETs, often used in combination for a better therapeutic result.[5] The protocols described in the published series are different; vincristine, adriyamycin, and cyclophosphamide are the most commonly used cytotoxic drugs; etoposide and ifosfamide also seem to have good results.[5]

The prognosis essentially depends on the existence of metastases, the tumor volume, and the treatment received, with surgical treatment combined with chemotherapy providing the best prognosis.[4] The average survival time is 12 months for metastatic stages.[4,5] For our patient, VAC-type chemotherapy allowed locoregional and remote control of the disease, with no sign of progression 24 months after the end of treatment.

4. Conclusion

Renal PNTEs are extremely rare tumors. Given the immense difficulty of proposing a reference therapeutic protocol for renal neuroectodermal tumors and their rarity, the management of these tumors is multidisciplinary. We report a clinical case of therapeutic success thanks to surgery followed by polychemotherapy based on cyclophosphamide, doxorubicin, and vincristine. The evolution was favorable without recurrence 2 years later.

CRediT authorship contribution statement

Idriss Ziani: Visualization, Resources, Formal analysis, Conceptualization. **Hicham Ouaziz:** Writing – original draft, Supervision, Resources, Investigation, Funding acquisition. **Kaoutar Aammou:** Investigation. **Imane Azzam:** Formal analysis, Data curation, Conceptualization. **Ahmed Ibrahimi:** Visualization, Validation. **Yassine Nouini:** Visualization, Supervision.

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