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Images of Interest / Imagens de Interesse

High-Risk Adrenal Neuroblastoma with Bone Metastases in an 18-Month-Old

Neuroblastoma Suprarrenal de Alto Risco com Metástases Ósseas em Criança de 18 Meses

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Abstract

Neuroblastoma is the most common extracranial malignant tumor in the pediatric population and a significant cause of morbidity and mortality. It arises from neural crest cells and exhibits a highly variable clinical course, ranging from spontaneous regression to widespread metastatic disease. We report the case of an 18-month-old child presenting with a limping gait and swelling of the left elbow. Radiographs demonstrated aggressive bone lesions in the left humerus and right femur, while ultrasound identified lymphadenopathy in the left axilla and right iliac region, as well as a right adrenal mass. Contrast-enhanced Computed Tomography confirmed the adrenal mass, which demonstrated image-defined risk factors, namely local organ infiltration. These findings are consistent with metastatic neuroblastoma. This case highlights the importance of comprehensive imaging in the diagnosis, staging and management of high-risk neuroblastoma.

Keywords

Neuroblastoma; Metastasis; Neoplasia.

Resumo

O neuroblastoma é o tumor maligno extracraniano mais comum na população pediátrica e uma causa significativa de morbilidade e mortalidade. Tem origem nas células da crista neural e apresenta um curso clínico altamente variável, desde a regressão espontânea até à doença metastática generalizada. Relatamos o caso de uma criança de 18 meses com claudicação da marcha e tumefação do cotovelo esquerdo. As radiografias demonstraram lesões ósseas agressivas no úmero esquerdo e fémur direito, enquanto que a ecografia identificou adenopatias na axila esquerda e região ilíaca direita, bem como uma massa suprarrenal direita. A Tomografia Computorizada com contraste confirmou a massa suprarrenal, que evidenciava fatores de risco definidos por imagem, nomeadamente a infiltração de órgãos adjacentes. Estes achados são compatíveis com neuroblastoma metastático. Este caso sublinha a importância de uma avaliação imagiológica abrangente no diagnóstico, estadiamento e abordagem do neuroblastoma de alto risco.

Palavras-chave

Neuroblastoma; Metástase; Neoplasia.

Case Presentation

An 18-month-old female presented to the emergency department with a limping gait, swelling of the left elbow, a palpable left axillary mass and fever. Laboratory tests showed anemia and elevated lactate dehydrogenase (LDH) levels.
Radiographs of the hip and left elbow demonstrated aggressive bone lesions (Figure 1). Ultrasound revealed a right adrenal solid mass and multiple enlarged lymph nodes in the left axilla and right iliac region (Figure 2).
Contrast-enhanced Computed Tomography (CT) of the thorax, abdomen and pelvis identified a 3.4 cm heterogeneous, hypodense right adrenal mass with coarse calcifications and mild enhancement (Figure 3). The lesion infiltrated the adjacent liver and contacted the superior pole of the right kidney. It contacted the posterior wall of the inferior vena cava (IVC) with no evidence of vascular encasement. These imaging findings, together with the bone and nodal involvement, suggested high-risk metastatic neuroblastoma.
Histopathological confirmation was obtained through multiple procedures, including left axillary lymph node biopsy, bone biopsy of the humeral lesion and bilateral bone marrow aspiration and biopsies of the posterior superior iliac crests. Histological examination demonstrated poorly differentiated neuroblastoma with a high mitotic–karyorrhectic index

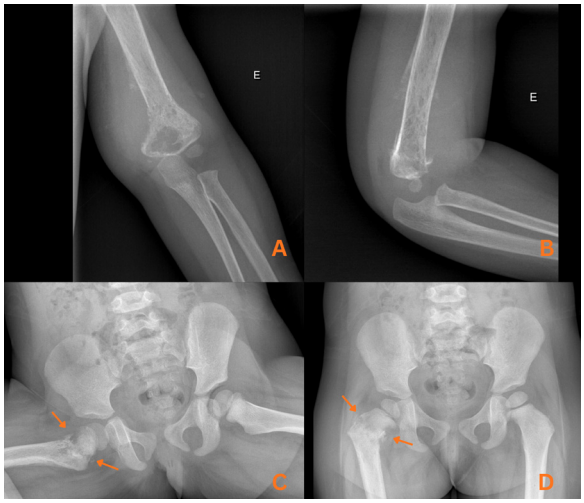

Figure 1 – Radiographs of the left elbow (A, B) show patchy, ill-defined lucencies in the distal humerus, cortical involvement with an exuberant periosteal reaction including a Codman triangle and a larger lytic lesion in the distal humeral epiphysis. The hip radiographs (C, D) demonstrate a lytic lesion in the proximal femur with cortical irregularity and disruption, compatible with a pathologic fracture (arrows). These findings are characteristic of aggressive bone lesions and metastatic disease should be considered in the differential diagnosis.

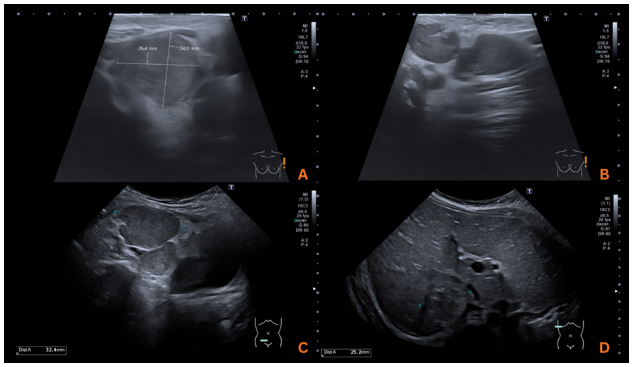

Figure 2 – Ultrasound images demonstrate multiple enlarged, heterogeneous lymph nodes in the left axilla (A, B) and right iliac region (C). A heterogeneous, predominantly hypoechoic mass is seen in the right adrenal region (D). Together with the previously identified aggressive bone lesions, these findings are compatible with a malignant process, most likely neuroblastoma, with nodal and skeletal involvement and warrant further imaging (with a CT or MRI) and clinical evaluation for staging and management.

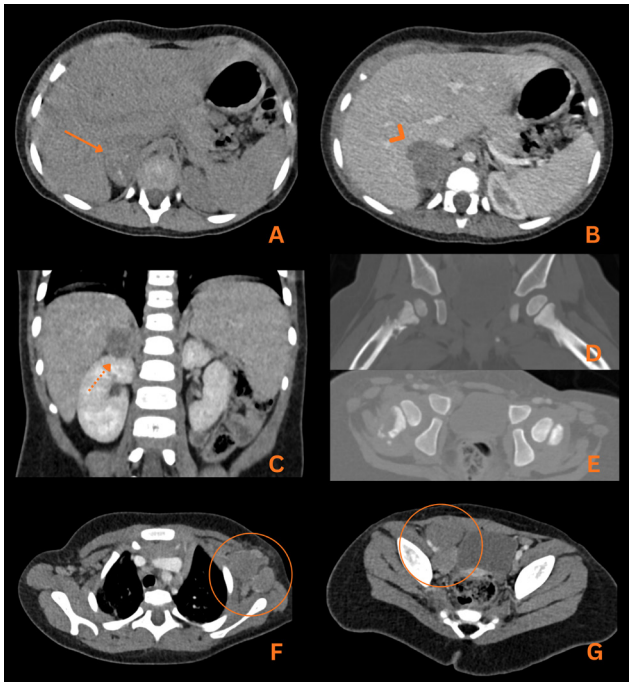

Figure 3 – Axial non-contrast CT image (A) shows a heterogeneous right adrenal mass (arrow) with coarse calcifications. Post-contrast images (B, C) show mild enhancement, with extension into the liver (arrowhead), contact with the inferior vena cava with no evidence of encasement and contact with the superior pole of the right kidney (dashed arrow). Coronal reconstruction (D) and axial image (E) show the previously identified lytic femoral lesion with cortical disruption consistent with a pathologic fracture. Axial images (F, G) confirm the previously noted lymphadenopathy (circles). These findings support metastatic adrenal neuroblastoma.

and unfavorable histology. Immunohistochemical staining was positive for synaptophysin. Molecular analysis of the tumor tissue showed amplification of the N-MYC gene, with additional chromosomal alterations including 1p-, 3p-, 2p+ and 17q+, which are associated with a worse prognosis and more aggressive disease. Bone marrow aspirates showed metastatic infiltration at approximately 15% of cells.

Functional imaging with iodine-123 metaiodobenzylguanidine (I-123 MIBG) scintigraphy was also performed. Image quality was partially limited by motion and positioning artifacts, which restricted full skeletal evaluation. Within these limitations, the International Society of Paediatric Oncology Europe Neuroblastoma Group (SIOPEN) score was ≥53/72, indicating a high metastatic skeletal burden.

Given the histological and molecular confirmation obtained from metastatic sites and according to international diagnostic criteria for neuroblastoma, an additional biopsy of the primary adrenal lesion was not considered necessary. Chemotherapy was promptly initiated.

Discussion

Neuroblastoma is the most common extracranial solid tumor in children and originates from neural crest cells. It typically occurs in early childhood, with a median age at diagnosis of 18 months and most often arises in the adrenal medulla.[1] Tumors can also occur along the sympathetic chain, including the cervical, thoracic and abdomino-pelvic regions. Common symptoms include an abdominal mass or systemic signs such as fever, fatigue or weight loss. Diagnosis relies on imaging, histopathological examination and molecular testing, including assessment for N-MYC gene amplification, which is associated with a worse prognosis.

Imaging is essential for the diagnosis, staging and management of neuroblastoma. Ultrasound is often the first-line modality, typically revealing a heterogeneous mass with calcifications. More detailed anatomical assessment is achieved through CT or Magnetic Resonance Imaging (MRI).[2] CT is useful for evaluating tumor size, calcifications, local invasion and vascular involvement. MRI is usually superior to CT, typically showing a heterogeneously T2 hyperintense, T1 hypointense mass with heterogeneous enhancement and is ideal for assessing spinal canal involvement when the tumor is near the spine. Both modalities help identify image-defined risk factors (IDRFs), such as encasement of major vessels or organ infiltration, which are linked to poor surgical outcomes and higher risk categories.[3] Functional imaging, particularly I-123 MIBG scintigraphy, is the preferred modality for detecting neuroblastoma lesions and metastases.

The International Neuroblastoma Risk Group (INRG) Staging System incorporates imaging and clinical features to stratify patients into risk categories.[3] In this case, the presence of bone metastases, lymphadenopathy and a right adrenal primary tumor with IDRFs is consistent with stage M disease.[3] Organ infiltration is defined by tumor extension causing the margins between the tumor and the infiltrated structure to be lacking or ill defined.[4] In our patient, this was evident in the liver, while involvement at the superior pole of the kidney was less certain and could have been better evaluated with MRI. According to INRG criteria, encasement of a vessel is defined as tumor contact involving more than 50% of the vessel circumference or a flattened vein with no visible lumen.4 In this case, tumor contact with the IVC did not meet these criteria and was therefore not considered an IDRF. These criteria differ for renal vessels, where even isolated contact is considered an IDRF because of the potential implications for surgical morbidity and renal preservation.[4]

Although MRI provides superior soft-tissue contrast and might have allowed a clearer delineation of potential renal involvement, contrast-enhanced CT was performed in the initial phase of the investigation during the evaluation of the child's clinical presentation, when the diagnosis of neuroblastoma had not yet been confirmed and was considered adequate for initial staging and therapeutic decision-making. Given the presence of extensive metastatic disease and IDRFs (namely hepatic infiltration), surgical resection was not considered and rapid induction chemotherapy was promptly initiated. MRI could have been considered at a later stage

in the event of surgical management. This approach aligns with international staging and treatment recommendations, which recognize both CT and MRI as acceptable modalities for initial evaluation.[3,4]

This case highlights the importance of early recognition and comprehensive imaging in high-risk neuroblastoma with metastatic involvement. The integration of multimodal imaging, histopathology and molecular data underscores the crucial role of radiology in diagnosis and staging, offering valuable educational insights into disease assessment and management.

Ethical Disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

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Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

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