

Antenatal adrenal neuroblastoma revealed by subcutaneous nodules

Jihane El Houssni, Jabour Soukayna, Siham El Haddad, Abourak Chaimae, Saber Abdellah Bassel, Tantaoui Mehdi, Latifa Chat, Nazik Allali

ABSTRACT

Introduction: Neuroblastoma is the third most common malignancy in pediatrics, subcutaneous metastases of antenatal neuroblastoma are described in the literature and they represent 1% of secondary localizations of neuroblastoma.

Case Report: We report the case of a 12-month-old male infant with an antenatal neuroblastoma of the left adrenal gland revealed by inguino-scrotal and forearm subcutaneous nodules whose anatomopathological study was in favor of cutaneous metastases of a neuroblastoma.

Conclusion: Neuroblastoma is a very frequent embryonic tumor in pediatrics, subcutaneous metastases are often revealing of antenatal neuroblastoma, the diagnosis of certainty is essentially based on anatomopathological study.

Keywords: Adrenal gland, Metastases, Neuroblastoma, Subcutaneous nodules

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INTRODUCTION

Neuroblastoma is a malignant solid embryonal tumor very common in pediatrics, originating in sympathetic tissue derived from neural crest cells. Imaging plays an important role in the management of patients with neuroblastoma and is essential to assess the metastatic extension. Subcutaneous metastases represent 1% of neuroblastoma metastases [1]. We report the case of an infant with left adrenal neuroblastoma in whom cutaneous metastases were the telltale sign.

CASE REPORT

An infant, male, 12 months old, from a consanguineous marriage, the onset of the symptomatology dates back to birth with the presence of subcutaneous nodules in the inguino-scrotal area (Figure 1) and in both forearms, mobile. An ultrasound (US) of the soft tissues was carried out, showing hypoechoic, rounded, well limited, regular contours subcutaneous lesions, poorly vascularized by color Doppler, containing calcifications (Figure 2).

The anatomopathological study of these subcutaneous nodules was in favor of a cutaneous localization of a round cell tumor whose morphological aspect and immunohistochemical profile are compatible with an undifferentiated neuroblastoma (Figure 1).

Abdominal US of this infant revealed a mass in the splenorenal space, oval, heterogeneous, well limited, containing calcifications, vascularized on color Doppler, pushing the kidney up and out with persistence of a separation line, encompassing the renal pedicle which remains however permeable (Figure 2).

Contrast-enhanced computed tomography (CT) performed as part of an extension workup showed a

hypodense, heterogeneous, well-limited, with scattered calcifications, retroperitoneal left adrenal mass, enhanced after contrast injection, measuring 31×42×52 mm (AP×H×T). Externally, it encompassed the left renal pedicle without endoluminal invasion, superiorly, it came into contact with the splenic parenchyma with loss of separation line without signs of infiltration, medially, it exceeded the midline and came into contact with the aorta and the celiac trunk, which however remained permeable, this CT also showed a heterogeneously enhanced, suspicious-looking thickening of the medial arm of the right adrenal gland, coelio-mesenteric adenopathies and multiple diffuse subcutaneous abdomino-pelvic, inguino-scrotal, and sacro-coccygeal nodules (Figure 3).

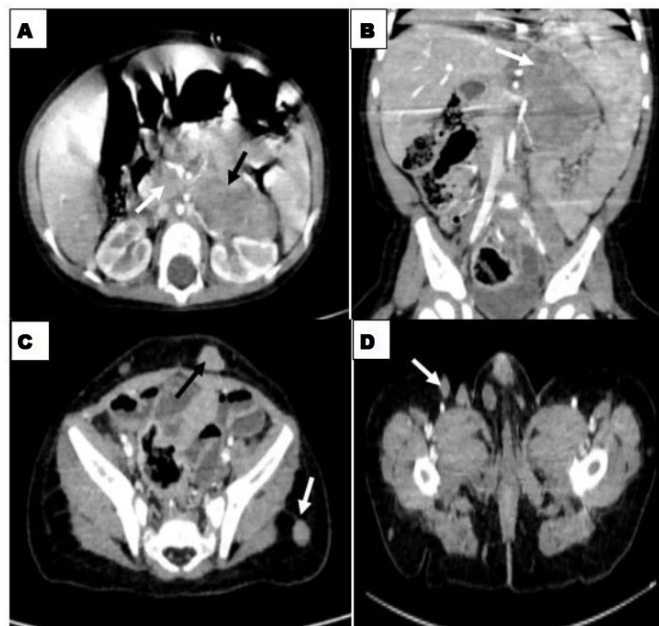


Figure 3: Contrast-enhanced abdominal and pelvic computed tomography: (A) Axial scan shows left retroperitoneal mass, hypodense, heterogeneous, well limited, containing calcifications, enhanced after contrast injection (black arrow), with celiac adenopathy (white arrow). (B) Coronal reconstruction showing left retroperitoneal adrenal process. And hypodense subcutaneous nodules in axial scans, (C) lumbar region (white arrow) and anterior abdominal wall (black arrow), and (D) in inguino-scrotal region (white arrow).



Figure 1: (A) Subcutaneous inguino-scrotal nodules, determining elevation of the skin contour, (B) Histopathology image shows tumor proliferation of rounded, monomorphic cells, their cytoplasm is not very abundant, with a high mitotic index higher than 4%, associated with fibro-vascular septa.

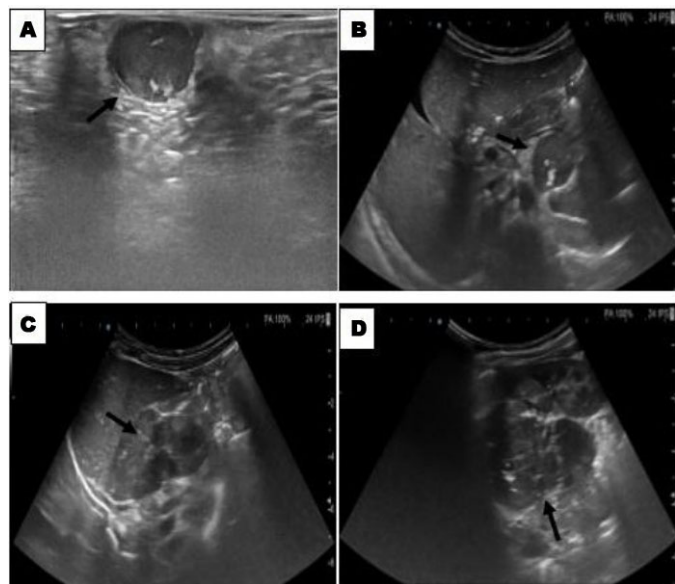


Figure 2: (A) US of superficial soft tissue and (B–D) abdominal: (A) Subcutaneous inguino-scrotal nodule, (arrow), hypoechoic, well limited, rounded, containing calcifications. (B) Coeliac adenopathy, (arrow), hypoechoic, well limited, containing calcifications. (C and D) Mass in the splenorenal space (arrow), oval, heterogeneous, well limited, containing calcifications, pushing the kidney up and out, with persistence of separation line.

DISCUSSION

Neuroblastoma is the second most common abdominal tumor in children after Wilms' tumor [2]. It is the third most frequent malignant tumor in pediatrics [3] with a clear male predominance [2]. Neuroblastoma is metastatic in 75% of cases [4]. It accounts for nearly 15% of cancer deaths in children [2]. Neonatal or antenatal neuroblastoma is described in the literature [2], it is almost always of adrenal origin (90%), it is metastatic in 50% of cases at the time of diagnosis [2], metastatic subcutaneous nodules are frequent in the newborn (32% of neonatal cases) [5]. Wesche et al. reported eight cases of neuroblastoma with cutaneous metastases [6]. Chen et al. described a case of antenatal neuroblastoma with subcutaneous nodules [7].

Our patient presented with a left adrenal neuroblastoma revealed by metastatic subcutaneous nodules present from birth.

Nodules usually occur on the trunk and extremities, other locations have been described such as the scrotum, soles of the feet, and palms of the hands [5].

In our case, the subcutaneous nodules are located in the inguino-scrotal, abdomino-pelvic, and sacro-coccygeal regions.

Ultrasound of superficial soft tissue, these nodules appear as heterogeneous, well-limited nodules with little vascularity on color Doppler [8]. This was like to our patient.

The second skin manifestation reported in patients with neuroblastoma is periorbital ecchymosis (Raccoon eyes) which are secondary to orbital metastases [5].

The main differential diagnoses of cutaneous metastases of neuroblastoma are subcutaneous nodules secondary to congenital TORCH infections (blueberry muffin Sd), histiocytosis, vascular tumors including hemangiomas, cutaneous metastases of hematological malignancies such as lymphoma and leukemia [5, 8].

CONCLUSION

Neuroblastoma is a very common embryonic tumor in pediatrics, subcutaneous metastases are often revealing of this disease, the diagnosis of certainty is essentially based on anatomopathological study.

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Author Contributions

Jihane El Houssni – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically

for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Jabour Soukayna – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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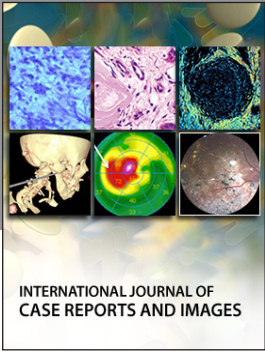
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