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Case Report

Metachronous Primary Muscular Neuroblastoma in an 8 Months Old Infant with Pepper Syndrome: Case Report

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Abstract

Neuroblastoma is the most frequent neoplasm in the first year of life and almost always arises from the adrenal glands or the sympathetic nervous system chain. We present an original case of simultaneous triple primary locations: adrenal gland, liver, and quadriceps.

Keywords: *Neuroblastoma; Pepper Syndrome; Muscular Location*

Introduction

Neuroblastoma (NB) is the second-most common solid tumor of infancy and childhood, exceeded in number only by brain tumors. It is the most common malignancy noted in newborns [1]. The peak incidence of neuroblastoma occurs at 2 to 3 years of age. Most primary tumors occur within the abdomen, mediastinum, retroperitoneum, and the paravertebral sympathetic ganglia in the neck or pelvis [2]. Neuroblastoma with primary or metastatic location in the skeletal muscle has rarely been described in the literature.

Case Report

An 8-year-old male with Pepper syndrome, which was diagnosed via CT scan (Figure 1). Initially, there was resection of the left adrenal gland where a well-defined mass was located, later confirmed through histopathology to be a neuroblastoma (Figure 2). The staging was determined to be INSS stage 4S. Subsequently, the child commenced rounds of chemotherapy.

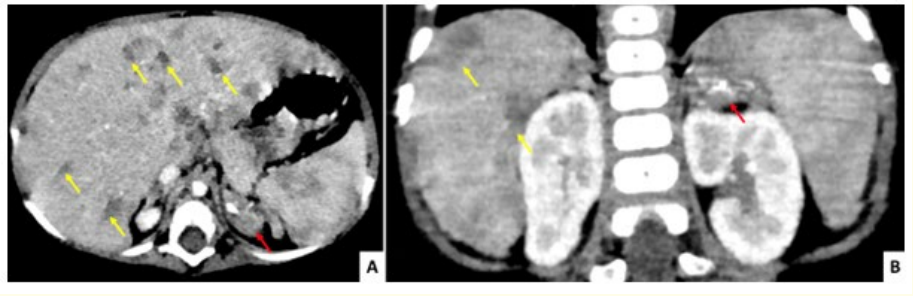

Figure 1: *CT of the abdomen injected in axial and coronal section showing: A: A left adrenal tissue mass, enhanced after injection, and containing calcifications (red arrow). B: Liver full of hypodense nodules related to secondary localizations (yellow arrows).*

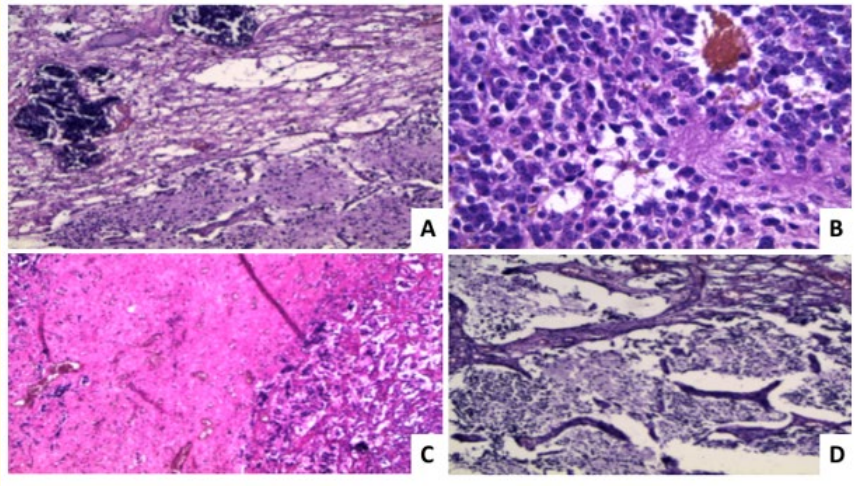

Figure 2: *Histological results concerning the adrenal mass: A: Poorly differentiated neuroblastoma characterized by an abundant neuropil background punctuated by basophilic nuclei of neuroblastic cells. Note at the top of the basophilic micro calcifications. Hematoxylin eosin; X 20. B: Poorly differentiated neuroblastoma. Neuroblast cells have rounded nuclei with finely nucleolated, granular chromatin. Note the fibrillar eosinophilic substance on the right below and on the right corresponding to the neuropil background. Hematoxylin eosin; x 40. C: Poorly differentiated neuroblastoma after chemotherapy. Fibro-inflammatory changes on the left and viable tumor on the right. Hematoxylin eosin; x 20. D: Neuroblastic tumor made.*

It appears that one year later, the child presented to your hospital for evaluation of a long-standing, painless left thigh mass. During the physical examination, no signs of insect bites or evidence of previous falls or trauma to the lower limbs were observed. The parents had noticed the mass a few months prior, and while it was initially painless and decreased in size temporarily, it has since progressively grown. No other clinical findings were noted. Ultrasound of the left thigh revealed a 3.5 x 3.5 cm, homogeneous, hypoechoic mass within the quadriceps muscle, with no calcifications and no vascular involvement observed on Doppler imaging (Figure 3).

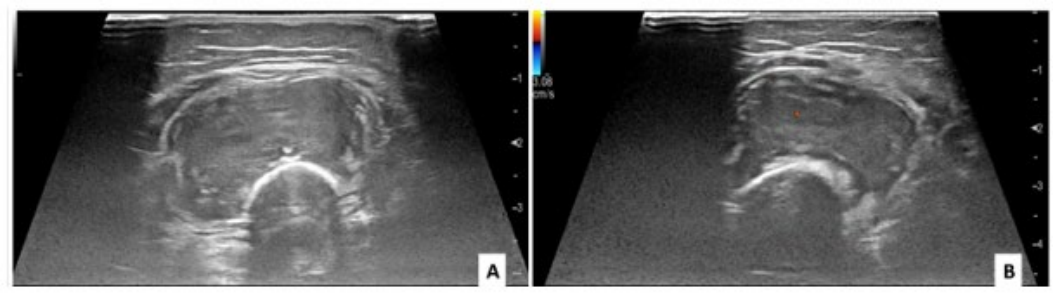

Figure 3: *B-mode ultrasound (A) and color doppler (B): oval mass, well limited depending on the quadriceps muscle coming into contact with the bone behind.*

Following the CT scan, which showed a well-defined mass originating from the quadriceps (Figure 4), the decision was made to perform an excisional biopsy. In the operating room, the mass appeared tan, grossly encapsulated, and measured. Subsequent histopathological analysis diagnosed the mass as a primitive neuroblastoma (stroma-poor), specifically of the poorly differentiated subtype (Figure 5).

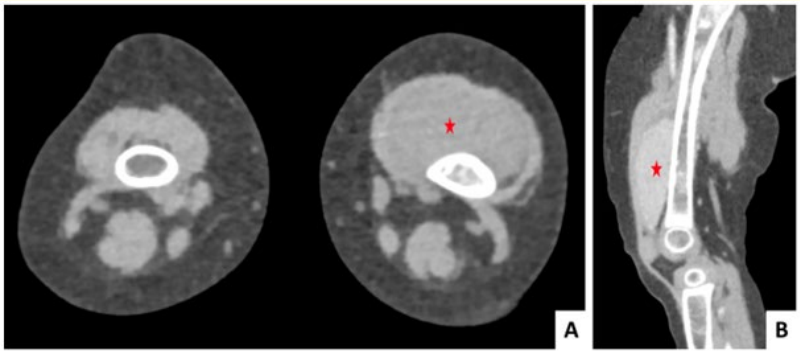

Figure 4: *CT of the knee without (A) and with injection of contrast agent (B), showing a well limited tissue like mass of the quadriceps muscle, enhanced after injection, and respecting cortical bone (red star).*

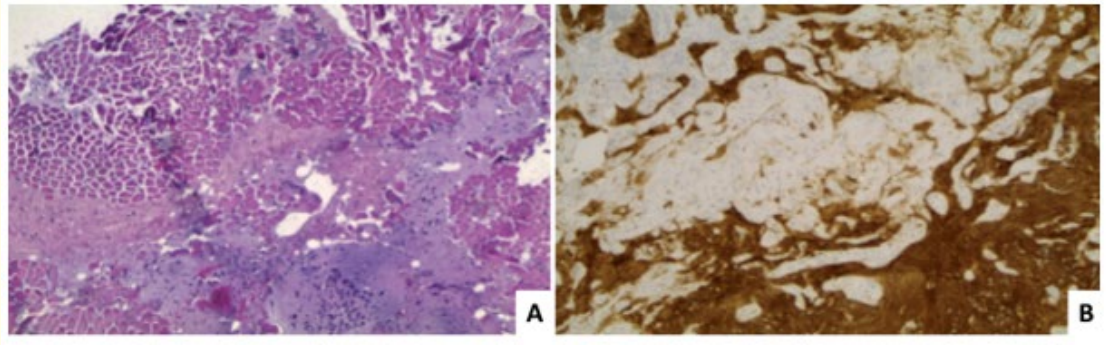

Figure 5: *Histological results of the muscular lesion: A: Muscle tissue from the quadriceps muscle infiltrated by patches of round neuroblast cells arranged on an abundant eosinophilic neurofibrillary background. Hematoxylin eosin; x 20. B: Tumor cells infiltrating muscle tissue as well as neuropil background are Synaptophysin positive. Immunohistochemistry; x 40.*

Discussion

The forearm is indeed an unusual site for the presentation of neuroblastoma, making your case potentially the first well-documented instance in pediatric literature of neuroblastoma originating from skeletal muscle. The primary metachronous neuroblastoma, particularly in this location, is rare and noteworthy [1].

Considering the appearance of another tumor after staging, several possibilities were considered for the left thigh mass: Unresolved metastasis of a stage 4S neuroblastoma: In this scenario, the primary lesion would be the adrenal mass. While there are few reports of infants with stage 4S neuroblastoma and multiple diffuse skeletal muscle nodules, they exhibit similar disease sites and risk factors, thus requiring chemotherapy [2].

Metachronous rare primary peripheral neuroblastoma: This hypothesis was confirmed based on histopathological results. Previous studies have shown favorable outcomes with fully resectable tumors despite adverse biological features [1,2].

Neuroblastoma metastasis with no identifiable primary site: While there are approximately 10 cases reported in the literature of neuroblastoma with unknown primary sites, they predominantly involve bone metastasis without muscle involvement, making this scenario less likely [1]. If the mass were smaller, it might have been missed on CT and better depicted by MRI due to its superior soft-tissue contrast resolution. Skeletal muscle lesions typically appear similar to surrounding muscle fibers with slight hyperdensity [2].

The patient underwent surgery after neoadjuvant chemotherapy, with the possibility considered that a primary tumor, possibly in the adrenal or another site, existed at birth but underwent regression, leaving the solitary deep thigh lesion undetected by later CT scans, especially in the absence of symptoms. Regardless of the scenario, the management remained consistent.

Conclusion

In conclusion, primary, peripheral, INSS-stage 1, 2A, or 2B, with favorable histology, neuroblastoma is extremely scarce if not unique and usually can be managed with an excisional biopsy as the only necessary treatment.

Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

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