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Case Report

Congenital neuroglial choristoma of the foot

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ABSTRACT

Neuroglial choristomas are rare malformations of heterotopic neural tissue that have been previously reported predominantly in the head and neck. Competing theories of embryogenesis propose their origin as encephaloceles that have undergone resorption of their cranial connection or displaced neuroectodermal cells which have undergone ectopic proliferation. Most cases occur in midline or para-midline structures. There have been no prior published cases of a neuroglial choristoma in the extremities. We present a case of a 13-month-old otherwise healthy child who presented to our institution with a slowly growing foot mass who was found to have a neuroglial choristoma. This case suggests an early embryological migration defect as the etiology and offers a unique differential consideration for a benign extremity mass.

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Introduction

Choristomas are heterotopic masses of differentiated cells. Neuroglial choristomas, sometimes called glial heterotopia, are exceedingly rare malformations of neural tissue which have been previously reported predominantly in the head and neck [1]. Cases are typically midline, though there have been cases reported involving the middle-ear structures [2]. Two principal theories have been advanced regarding the origins of neuroglial choristoma. In the head and neck, they may arise from herniated brain tissue (encephalocele), which has

subsequently undergone resorption of its intracranial connection. Alternatively, an error of early embryonic development may result in displaced neuroectodermal cells which undergo differentiation and ectopic proliferation [3]. Previously described cases typically have been in the midline head and neck, though cases have been reported in the thoracolumbar soft tissues and lungs [4–7]. To our knowledge, there have been no prior published cases of a neuroglial choristoma in an extremity. Almost all prior cases required surgical biopsy and subsequent pathology evaluation and immunohistochemical staining work up for diagnosis confirmation [8].

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Fig. 1 – Photos of the patient's right foot demonstrating soft tissue masses at the plantar aspect of the foot and phalangeal hypoplasia.

Case report

A 13-month-old otherwise healthy girl presented to our institution for evaluation of a slowly growing right foot mass (Fig. 1). The child was diagnosed prenatally with a right foot deformity on anatomy ultrasound. Since birth, her parents reported slowly increasing size of a plantar mass. On physical examination, she was noted to have 2 nontender masses on the plantar aspect of her foot, hypoplasia of the distal phalanges, and fusion of the hypoplastic distal second and third phalanges.

As the mass continued to grow, plain films of the right foot were obtained, which demonstrated hypoplasia of the distal phalanges, absence of the fifth ray, and partial fusion of the third and fourth metatarsal (Fig. 2). An MRI of the right foot with and without intravenous contrast was obtained. Two T1-isointense, T2/PD-heterogeneously mildly intense, and mildly enhancing soft tissue masses measured 2.3×1.9 cm medial to the calcaneus and 3.8×3.2 cm at the plantar aspect of the metatarsals extending superiorly between the second and third metatarsals. There was displacement of bone without osseous erosion or perilesional edema. The masses appeared to have a fibrous component and differential diagnoses that were considered included fibromatosis and infantile fibrosarcoma (Fig. 3).

An open biopsy of the larger soft tissue mass was performed and demonstrated bundles of collagenous fibrous tissue and interspersed nodules of neuroglial tissue. The

nodules mainly contained paucicellular glial tissue with scattered neurons (Fig. 4a). No atypia or increased mitotic activity was seen. A panel of immunohistochemical staining was performed to support the histologic findings (Fig. 4b-d). In summary, the nodules were diffusely positive for S100, neuron-specific enolase, neurofilament and glial fibrillary acidic protein, and NeuN highlighted scattered neurons, consistent with neuroglial choristoma. Due to the location of lesion, the possibility of nerve sheath tumors and ectomesenchymoma was considered. However, epithelial membrane antigen, desmin, and myogenin were negative and did not support these differentials and the diagnosis of neuroglial choristoma was made. Curative treatment with complete surgical excision and foot reconstruction is planned.

Discussion

The evaluation of pediatric soft tissue masses typically begins with ultrasound and plain film. In this case, X-ray findings indicated the presence of an atypical underlying mass, prompting MRI evaluation. MRI characteristics of neuroglial choristoma are similar to brain, with T1 isointensity, mild T2/PD heterogeneous hyperintensity, and mild postcontrast enhancement. Neuroglial choristomas are exceptionally rare, usually occur in the head and neck, and have never before been reported in the extremities. Two theories posit that



Fig. 2 – X-ray images of the patient’s feet. Bilateral AP (a), right lateral (b), and oblique (c) projections demonstrate hypoplasia of the right distal phalanges, absence of the fifth ray, and partial fusion of the third and fourth metatarsal.

they may arise as encephaloceles that have lost their cranial connection or as displaced neuroectodermal cells which have undergone proliferation. In this case, given its distal location, the choristoma likely arose from an early embryological

migration defect. Neuroglial choristoma should be considered in the differential diagnosis of a pediatric soft tissue mass, especially in the midline and head and neck area, or when pathology features suggest neural differentiation.



Fig. 3 – MRI of the patient’s foot. Axial T1 fat-saturated (a) and PD fat-saturated (b) images demonstrate a soft tissue mass at the plantar aspect of the anterior foot which extends between the metatarsals (arrows), with isointense T1 signal and heterogeneous mild hyperintense PD signal without adjacent edema. Coronal T2 FS precontrast (c) and postcontrast (d) images demonstrate 2 masses (arrowheads) with heterogeneous mild hyperintense T2 signal and mild postcontrast enhancement. Sagittal T1 fat-saturated precontrast (e) and postcontrast (f) demonstrate well margination of the 2 masses (arrows) without osseous invasion and mild postcontrast enhancement.

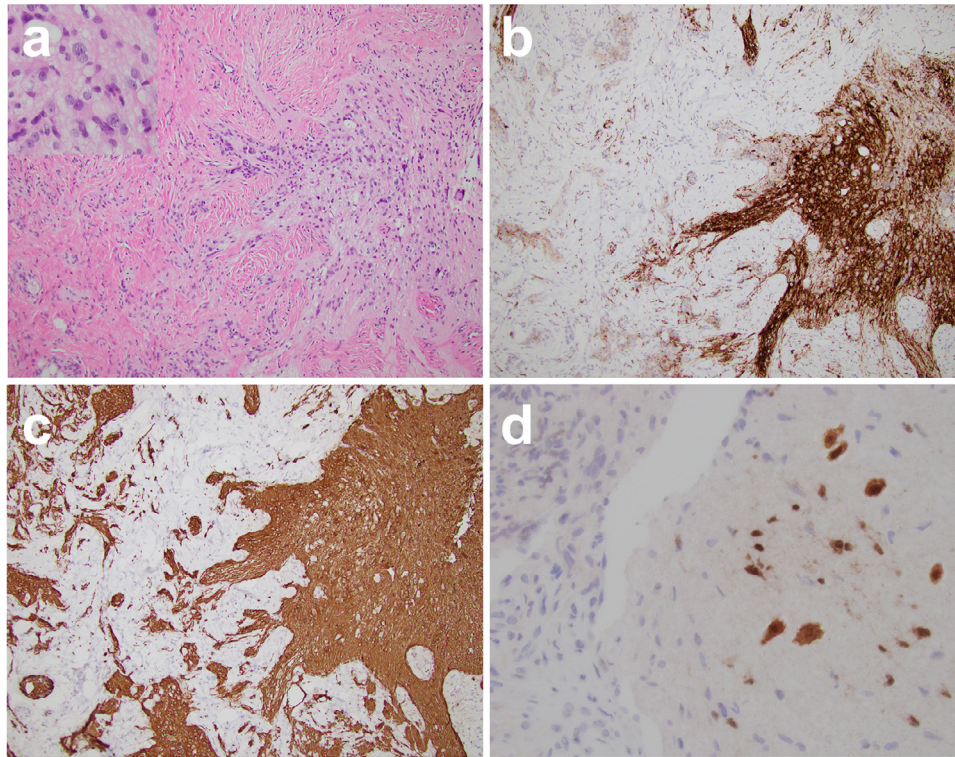


Fig. 4 – Biopsy specimen. Microscopic sections show fibrous tissue containing interspersed nodules of neuroglial tissue (a, H&E, $\times 100$), inset shows mainly paucicellular glial tissue with scattered neurons (H&E, $\times 400$). Neuroglial nodules are diffusely positive for NF (b) and GFAP (c) immunohistochemical staining, ($\times 100$). NeuN immunohistochemistry highlights scattered neurons (d, $\times 400$).

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2019.03.025](https://doi.org/10.1016/j.radcr.2019.03.025).

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