Child's Nervous System (2019) 35:1603–1607
https://doi.org/10.1007/s00381-019-04057-8

CASE REPORT

Pediatric nodular fasciitis at the roof of the optic canal causing decreased vision: case report and review of the literature

Ako Matsuhashi[1] · Kenichi Usami[1] · Eitaro Ishisaka[1] · Hideki Ogiwara[1]

Received: 29 October 2018 / Accepted: 7 January 2019 / Published online: 1 February 2019

Abstract

We present a 3-year-old girl with decreased visual acuity of the left eye. Radiological studies revealed a mass lesion at the roof of the left optic canal with bony erosion, which compressed the left optic nerve. Gross total resection of the mass and decompression of the optic canal were performed. Histopathological study was consistent with nodular fasciitis. This is the first report of nodular fasciitis at the roof of the optic canal in a young child. Nodular fasciitis compressing the optic nerve should be included in differential diagnoses of optic neuropathy in young children.

Keywords Nodular fasciitis · Optic canal · Decreased vision

Introduction

Nodular fasciitis (NF) is a rare non-malignant fibroblastic proliferation, typically found in subcutaneous tissue of the extremities [1–4]. It is often confused with malignant tumor due to its rapid growth, high cellularity, and infiltrative borders in pathology [1, 5]. NF is commonly diagnosed in adults aged 20–40 years, and its prevalence in children is as low as 10% of reported cases [2, 6]. Tumor location varies, but NF preferably develops in the upper extremities and trunk; head and neck lesions compose 7–15% of the cases [3–5].

Published reports of pediatric cases of NF are limited due to the rarity of this condition, and among them, most cases present with symptoms of lesion growth or local pain. Recommended treatment is the complete local excision, though an extent of excision should be tailored to spare normal anatomy and function [2].

Herein, we report the first case of a young child with NF located at the roof of the optic canal, causing compressive optic neuropathy.

✉ Hideki Ogiwara
hideki_o@d5.dion.ne.jp

[1] Division of Neurosurgery, National Center for Child Health and Development, Okura 2-10-1, Setagaya-ku, Tokyo 157-8535, Japan

Case report

A 3-year-old girl was referred to our institute with decreased left vision. She was found to have strabismus at the age of 2.5 years. The eidoptometry could not be performed then because of her age. She was found to have decreased left vision at the age of 3 years. Her visual acuity was 20/200 in the OS, 20/20 in the OD.

At presentation at out hospital, her left visual acuity was decreased to 20 cm/numerus digitorum. Magnetic resonance imaging (MRI) of the brain revealed an enhancing mass lesion at the roof of the left optic canal. The left optic nerve was compressed by the mass. Computed tomography (CT) scan revealed a 6 × 5 × 5-mm mass and local osseous erosion at the roof of the left optic canal (Fig. 1). Differential diagnoses included osteohemangioma, osteoid osteoma, intraosseous hemangioma, intraosseous meningioma, fibrous dysplasia, and Langerhans cell histiocytosis. Whole-body CT scan revealed no other lesions. We proceeded to the surgical resection to diagnose the lesion and decompress the optic nerve.

Regarding the surgery, left fronto-temporal craniotomy was placed, and the lesion was approached via extradural subfrontal route. A part of the roof of the left optic canal was eroded by the lesion. The lesion was reddish-brown in color, elastic hard, and vascularity was intermediate. The roof of the optic canal was fully decompressed with partial anterior clinoidectomy, and the lesion was totally resected

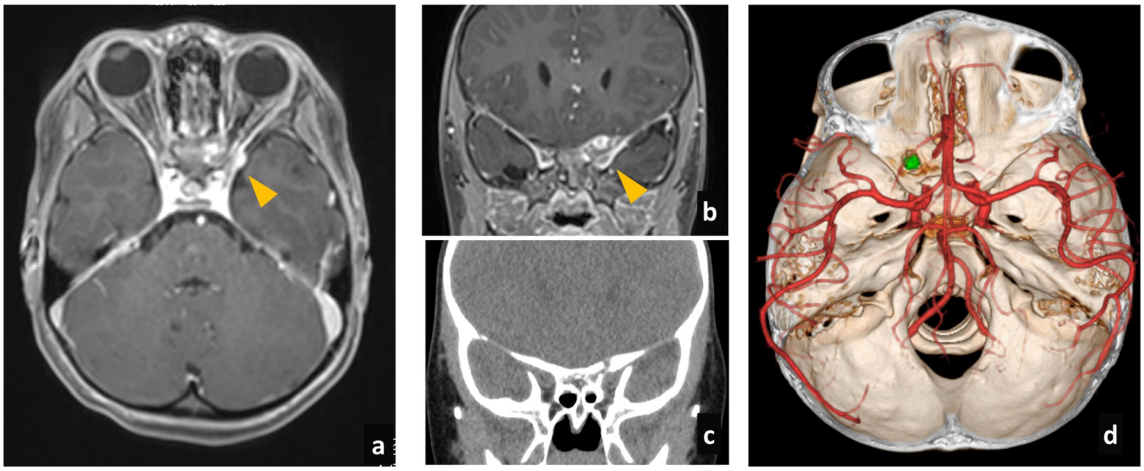

Fig. 1 Axial and coronal views show the enhancing lesion at the roof of the left optic canal (arrowhead) (**a**, **b**). Local osseous erosion in the left sphenoid bone is revealed on CT (**c**). 3D reconstruction imaging demonstrating bony erosion and the mass lesion (d)

(Fig. 2). There was no perioperative complications. Postoperative CT scan showed fully decompressed optic canal, and MRI showed no residual tumor (Fig. 3). Histopathological examination showed hyperplastic spindle cells that composed fibrotic interstitial tissue, which was consistent with nodular fasciitis (Fig. 4). The spindle cells showed no evidence of significant nuclear pleomorphism or malignant cytology. Ki-67 proliferative index was 2%, inflammatory cell markers CD 68 showed only rare positive cells. There was no recurrence at 3 months after the surgery. Her vision of the left eye has been unchanged at that time.

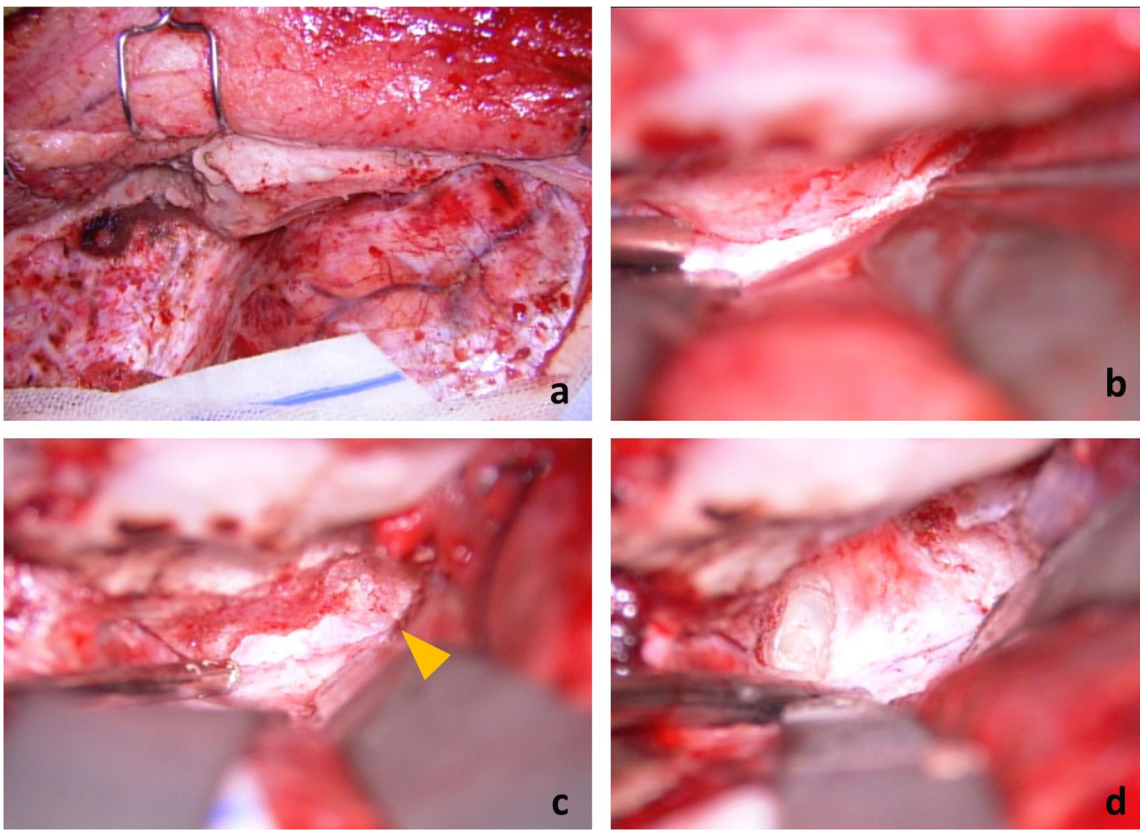

Fig. 2 The lesion was approached via extradural subfrontal route (**a**, **b**). The roof of the left optic canal was eroded by reddish-brown lesion (arrowhead) (**c**), which was fully decompressed (**d**)

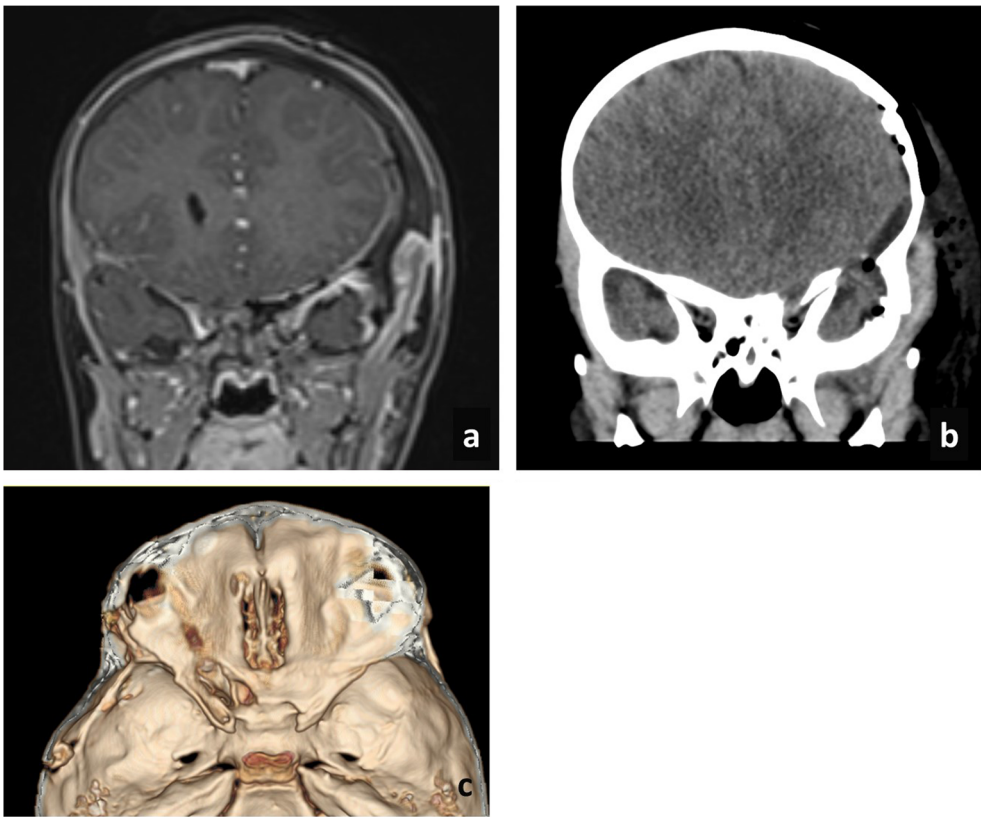

Fig. 3 Postoperative MRI showed no residual tumor (**a**). Postoperative CT showed fully decompressed optic canal with partial anterior clinoidectomy (**b**, **c**)

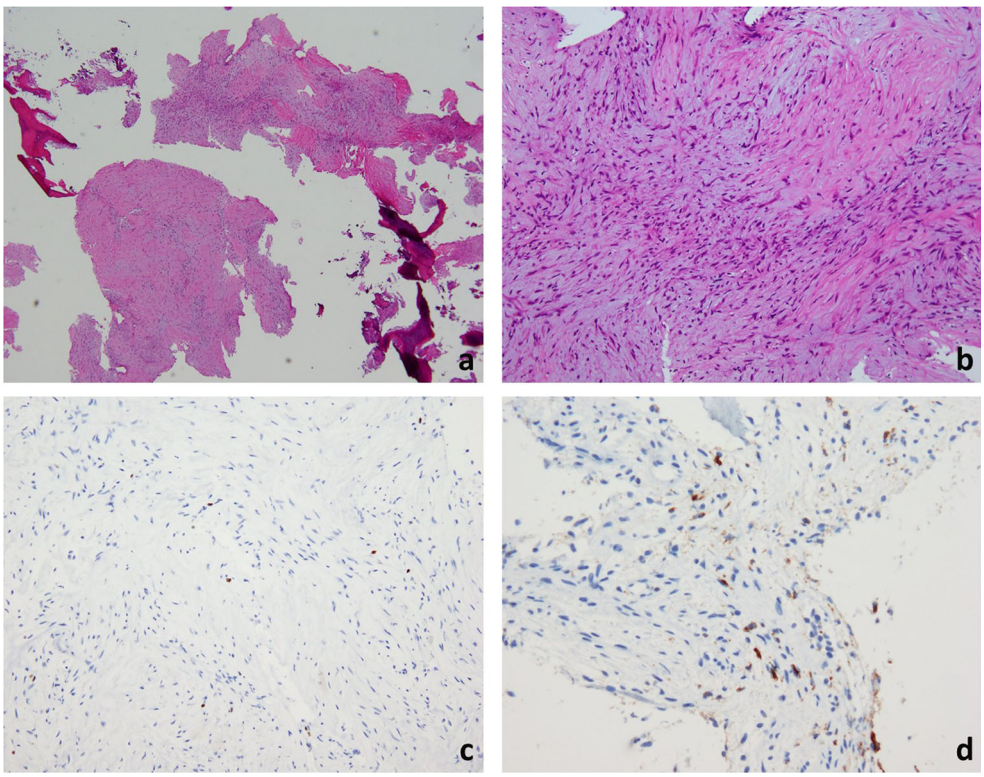

Fig. 4 Histopathological examination showed hyperplastic spindle cells that composed fibrotic interstitial tissue (**a**, **b**). Ki-67 proliferative index was 2% (**c**), inflammatory cell marker CD 68 showed only rare positive cells (**d**). H & E (**a**, **b**), Ki-67 (**c**), and CD 68 (**d**), original magnification × 40 (**a**), × 200 (**b**, **c**), and × 400 (**d**)

Discussion

Clinical manifestations in children with nodular fasciitis of the orbit

NF in the periorbital area is uncommon, and composes less than 1% of NF [3]. The most common clinical manifestations of NF is a solitary, rapidly growing mass with frequently associated shooting pain of the local area, numbness, and paresthesia, caused by compression of peripheral nerves [7, 8]. As for NF in the periorbital area, patients most frequently present with palpable and painful lump in the periorbital area without proptosis or displacement of the globe [9]. Though there are a few reports of cases that presented with unilateral proptosis or medial deviation of the eye globe, NF tends to occur in anterior periocular structures and it is extremely rare for NF to rise in the deep orbit [10, 11]. To the authors' knowledge, there has been only one report of NF in the deep orbit that caused compressive optic neuropathy [16]. They reported of NF in the orbital apex, for which subtotal resection and decompression of the optic canal were conducted.

Radiological features of nodular fasciitis of the orbit

NF has moderate to marked enhancement on CT and MR, and is a relatively well-defined, soft-tissue mass of superficial location [7]. However, radiological features of NF being nonspecific, radiological differential diagnoses include a wide variety of tumors such as rhabdomyosarcoma, fibrosarcoma, neurogenic tumor, dermoid or epidermoid, hemangioma, and malignant fibrous histiocytoma [7]. Dermoid cyst is more likely to produce a bony fossa and is not as readily enhanced as NF [11]. Some NF that are deep-seated tend to be large and have ill-defined margins, making it impossible to differentiate NF from malignant tumors radiographically. Therefore, surgical excision is generally considered appropriate for definitive diagnosis [7, 11].

Histopathological features of nodular fasciitis of the orbit

Histopathological study of NF shows proliferation of spindle-shaped fibroblasts which are frequently arranged in parallel bundles [12]. Although differentiation from other fibrous tumors, such as fibromatosis, fibrous histiocytoma, and fibrosarcoma, is difficult, NF tends to be more circumscribed with a higher mitotic activity, a greater resemblance to granulation tissue, and an increased myxoid matrix [7, 13]. Sarcoma lesions are marked with abundant spindle cells and atypical mitosis [14]. While mitoses may be numerous in NF, the spindle cell nuclei are never hyperchromatic and atypical mitoses are virtually never seen.

Treatment strategy and follow-up for nodular fasciitis of the orbit

Complete surgical excision is recommended when feasible [15]. Since all reported cases of NF in the ocular area have been managed by surgical excision, the natural course of this condition is not well established, though it is speculated that NF may respond to corticosteroids or undergo spontaneous regression [11]. Local recurrence is reported as extremely rare following complete excision [12].

Nodular fasciitis of the orbital apex

Kloek et al. have reported five cases of compressive optic neuropathy in adults (mean age 43), secondary to benign-appearing tumor at the orbital apex [17]. All cases underwent surgical bony decompression of the optic canal without tumor resection nor biopsy and demonstrated improvement in visual function. This shows that orbital decompression is an effective treatment for compressive optic neuropathy.

It is important to consider NF involving the orbital apex as one of the diagnoses when detecting a rapid decline of visual acuity. Difficulty of detecting deterioration of visual acuity in young children at an early phase makes it crucial to reach a definite diagnosis and find the best possible cure as quickly as possible.

Conclusions

We reported the first case of nodular fasciitis at the roof of the optic canal in a young child causing unilateral decreased vision. Nodular fasciitis involving the orbital apex should be included in differential diagnoses of optic neuropathy in young children.

Compliance with ethical standards

Conflict of interest On behalf of all authors, the corresponding author states that there is no conflict of interest.

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