Auris Nasus Larynx 40 (2013) 569–572

Contents lists available at SciVerse ScienceDirect

Auris Nasus Larynx

journal homepage: www.elsevier.com/locate/anl

Infantile inflammatory pseudotumor of the facial nerve as a complication of epidermal nevus syndrome with cholesteatoma

Naohito Hato [a,*], Mika Tsujimura [a], Taro Takagi [a], Masahiro Okada [a], Kiyofumi Gyo [a], Mikiko Tohyama [b], Hisamichi Tauchi [c]

[a] Department of Otolaryngology, Ehime University School of Medicine, Ehime, Japan
[b] Department of Dermatology, Ehime University School of Medicine, Ehime, Japan
[c] Department of Pediatrics, Ehime University School of Medicine, Ehime, Japan

ARTICLE INFO

Article history:
Received 9 May 2012
Accepted 11 January 2013
Available online 19 February 2013

Keywords:
Facial nerve
Inflammatory pseudotumor
Epidermal nevus syndrome
Cholesteatoma

ABSTRACT

The first reported case of facial paralysis due to an inflammatory pseudotumor (IPT) of the facial nerve as a complication of epidermal nevus syndrome (ENS) is herein presented. A 10-month-old female patient was diagnosed with ENS at 3 months of age. She was referred to us because of moderate left facial paralysis. Epidermal nevi of her left auricle extended deep into the external ear canal. Otoscopy revealed polypous nevi and cholesteatoma debris filling the left ear. Computed tomography showed a soft mass filling the ear canal, including the middle ear, and an enormously enlarged facial nerve. Surgical exploration revealed numerous polypous nevi, external ear cholesteatoma, and tumorous swelling of the facial nerve. The middle ear ossicles were completely lost. The facial paralysis was improved after decompression surgery, but recurred 5 months later. A second operation was conducted 10 months after the first. During this operation, facial nerve decompression was completed from the geniculate ganglion to near the stylomastoid foramen. Histological diagnosis of the facial nerve tumor was IPT probably caused by chronic external ear inflammation induced by epidermal nevi. The facial paralysis gradually improved to House–Blackmann grade III 5 years after the second operation.

1. Introduction

Epidermal nevus syndrome (ENS) is a rare, congenital, sporadic neurocutaneous disease characterized by the presence of an epidermal nevus and various developmental abnormalities of the skin, eyes, and nervous, cardiovascular, and genitourinary systems [1,2]. Lesions are present at birth or develop during childhood, mostly in the first year of life. Signs and symptoms arise corresponding to a defect in the ectoderm, which is the outer layer of the embryo that gives rise to the epidermis and neural tissue. As far as we know, involvement of the ear canal is very rare. We herein report the first case of recurrent facial palsy by an inflammatory pseudotumor (IPT) of the facial nerve as a complication of ENS.

2. Case report

A 10-month-old female patient was referred to our department because of a 1-month history of left facial paralysis. According to her doctor (a dermatologist), she was diagnosed with ENS at 3 months of age because of admixed symptoms of left opticopupillary dysplasia, retinal atrophy, and congenital esotropia. Numerous brown rough plaques were noted on her left scalp, cheek, and auricle (Fig. 1a). The right ear appeared to be normal, whereas the left ear had an epidermal nevus that extended from the auricle to deep in the external ear canal (Fig. 1b). Because the ear canal was filled with epidermal nevi and debris, the tympanic membrane could not be observed. Computed tomography (CT) revealed that the ear canal was filled with a soft mass suggesting cholesteatoma and the horizontal portion of the facial nerve was enlarged, with destruction of the surrounding bone (Fig. 2). A facial neuroma was initially suspected; however, an IPT was presumed because magnetic resonance images (MRI) of the facial nerve were hypointense to the brain on T1 and T2. Bacteriological study of the left ear revealed coagulase-negative staphylococcus.

Eight days after the first visit, she underwent surgery via a transcanal approach because oral prednisolone administration

* Corresponding author at: Department of Otolaryngology, Ehime University School of Medicine, Shitsukawa, Toon City, Ehime 791-0295, Japan.
Tel.: +81 89 960 5366; fax: +81 89 960 5368.
E-mail address: nhato@m.ehime-u.ac.jp (N. Hato).

0385-8146/$ – see front matter © 2013 Elsevier Ireland Ltd. All rights reserved.
http://dx.doi.org/10.1016/j.anl.2012.11.016

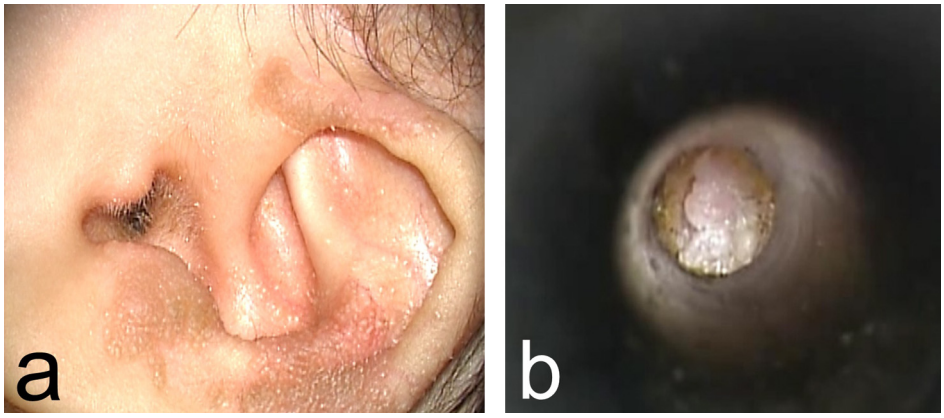

Fig. 1. Epidermal nevus of (a) the left auricle and (b) the left external ear canal.

(1 mg/kg/day for 3 weeks) proved ineffective. The ear canal was filled with many epidermal nevi, and open-type cholesteatoma was found behind them. The mastoid antrum and attic were filled with the cholesteatoma. They were removed together with the tympanic membrane, which was completely adhered to the promontory. The middle ear ossicles were unrecognizable. When the epidermal nevi and cholesteatoma were removed, a large tumorous facial nerve was exposed at the horizontal portion, occupying most of the middle ear cavity. The surrounding bony wall of the facial canal, which was thin in nature, was removed. Because the facial nerve was firm, enlarged, and rich in vasculature while keeping its original configuration, we believed that it might be involved in an inflammatory process and compatible with IPT. Radical mastoidectomy was conducted, but further manipulation of the nerve was not performed. The postoperative course was uneventful. Histopathology of the polypous mass in the ear canal was compatible with epidermal nevus: hypertrophic epidermis with hyperkeratosis, hypergranulosis, and prominent papillomatosis and many hair follicles with hypertrophic outer root sheaths. The patient was discharged from our hospital 15 days after the operation. The left facial palsy improved until it was almost normal 2 months postoperatively.

The patient presented again to our hospital 5 months after the surgery due to recurrence of the left facial paralysis. Bacteriological study of the left ear revealed no infection. CT showed that the radical cavity of the mastoid was filled with a soft mass suggesting recurrent cholesteatoma and that the facial nerve was enlarged from the geniculate ganglion to the mastoid portion, with destruction of the surrounding bone. MRIs of the facial nerve were hypointense to the brain on T1 and T2 and slightly enhanced on T1-Gd (Fig. 3). Despite three oral steroid treatments (prednisolone 1 mg/kg/day for 5 days, tapered thereafter), her facial paralysis gradually worsened to almost complete palsy (House–Blackmann grade VI). A second operation was conducted 10 months after the first surgery. The cholesteatoma occupied the radical mastoid cavity and tympanic space, including the Eustachian tube. The inner ear and petrous part were kept intact. Total removal of the recurrent cholesteatoma revealed that the exposed facial nerve was further enlarged. As the facial nerve at the horizontal portion was herniated, the bony wall at the geniculate ganglion and the vertical portion were removed to restore the smooth shape. After the decompression of the enlarged facial nerve, resection of the thin nerve sheath was performed from the geniculate ganglion to near the stylomastoid foramen. The firm and swollen nerve form was stable before and after the sheath resection. The chorda tympani was also enlarged, continuing to the facial nerve. A small piece of the vertical portion of the enlarged facial nerve with the nerve sheath and the chorda tympani near the exit from the facial nerve trunk were obtained separately for histological study using a small scalpel (Fig. 4). The wound was closed without reconstruction of the ossicular chain because the stapes footplate was unrecognizable.

Histological study revealed denervation in most of the nerve fibers of the specimen resected from the mastoid portion of the facial nerve (Fig. 5). Few nerve fibers remained in the specimen of the distal portion of the chorda tympani. Extensive proliferation of collagen fibers in the presence of capillaries and scattered inflammatory cells were noted. No anaplastic cells could be seen. Findings of reactive fibrous tissue with abundant lymphocytes and plasma cells with small regular nuclei were consistent with IPT. We speculated that the facial paralysis was caused by abnormal proliferation of collagen fibers inside the facial nerve, possibly from the interstitial space between the nerve fibers.

At the 5-year follow-up, the patient's perioral muscular tone had visibly improved, and her mouth and cheek movements had been restored to some degree. The patient could blow the cheek out, whistle, grin, and depress the lower lip. In contrast, slight improvement in muscular tone or movement was notable in the

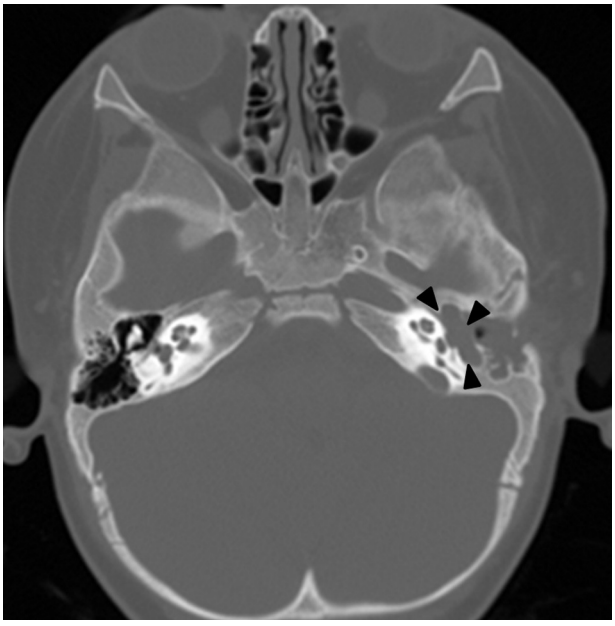

Fig. 2. CT image on the horizontal plane. The left mastoid was filled with a soft mass. The left facial nerve was enlarged at the horizontal portion, with destruction of the surrounding bone (black arrowheads).