Clinical characteristics of nodular fasciitis of the ear in children

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Abstract
The rate of early misdiagnosis in patients with nodular fasciitis of the ear is high. To provide a basis for clinical diagnosis and treatment, we aimed to summarise the clinical manifestations, imaging results, pathological findings, treatment strategies, and postoperative follow-up results for three cases of paediatric nodular fasciitis (two girls, one boy) treated in the Department of Otorhinolaryngology, Head and Neck Surgery, at Beijing Children’s Hospital of Capital Medical University from 2018 to 2020. The average age at diagnosis was 24 months. Lesions occurred in the left ear in two cases and right ear in one case. All patients had a history of biopsy before surgery. Rapid growth was observed following biopsy in two patients, and anti-inflammatory treatment was ineffective in all three cases. Fluorescence in situ hybridisation analysis of ubiquitin-specific peptidase 6 (USP6) was performed in two of the three cases, with positive results. The lesions exhibited hypo-intensity or iso-intensity on T1-weighted magnetic resonance imaging (MRI) and heterogeneous hyper-intensity on T2-weighted MRI. “Fascial tail” signs were observed on imaging in all cases. Surgical resection was performed in all cases. Intact ear appearance was observed at follow-up, and there were no cases of recurrence.

Conclusion: Combining clinical features with imaging findings may improve the accuracy of preoperative diagnosis in patients with nodular fasciitis. In addition to pathological findings, genetic testing for USP6 may aid in diagnosis. The final diagnosis should be based on comprehensive assessment. Complete surgical resection can prevent recurrence.

What is Known:
• Paediatric NF around the ear is rare and is easily misdiagnosed as other inflammatory masses that have a higher incidence in children.
• Most previous reports of NF were case reports.

What is New:
• Combining clinical and imaging findings with genetic testing for USP6 rearrangement may improve the accuracy of preoperative diagnosis in patients with NF. Nonetheless, the final diagnosis should be based on comprehensive assessment.
• The present paper is significant in that it represents the only report of three cases of ear NF in children with a complete medical history and prognosis.

Keywords Nodular fasciitis · Children · Ear · Manifestations · USP6 gene

Abbreviations
CT Computed tomography
FISH Fluorescence in situ hybridisation
H&E Haematoxylin and eosin
IHC Immunohistochemistry
MRI Magnetic resonance imaging
MYH9 Myosin heavy chain 9
NF Nodular fasciitis
SMA Smooth muscle actin
STIR Short tau inversion recovery
T1WI T1-weighted imaging
T2WI T2-weighted imaging
**Introduction**

Nodular fasciitis (NF) refers to a rare, benign, and self-limiting reactive soft tissue tumour composed of fibroblasts and myofibroblasts. This disease was first described by Konwaler et al. in 1955; at which time, it was termed pseudosarcomatous fasciitis [1]. NF can be misdiagnosed as a malignant lesion due to its rapid growth and high cellularity upon histopathological examination. The lesion can occur anywhere, although it is mostly observed in the upper limbs (48%), followed by the trunk (20%) and the head and neck (13–20%) [2, 3]. The age at NF onset is usually between 20 and 40 years, and the prevalence rate in children is approximately 10% [4, 5]. NF that occurs around the ear is rare, particularly in children. To the best of our knowledge, only a few cases of NF of the ear have been reported [6, 7]; thus, descriptions of the associated findings have been limited. To strengthen diagnostic awareness of paediatric NF around the ear, we retrospectively analysed the clinical features, diagnosis, and treatment of three cases encountered in our department.

**Materials and methods**

We conducted a retrospective analysis of data collected from three children with NF who were treated in the Department of Otorhinolaryngology of Beijing Children’s Hospital at Capital Medical University. Combined manifestations and cytomorphological and immunohistochemical features were used to confirm the diagnosis of NF.

All three patients underwent computed tomography (CT), magnetic resonance imaging (MRI), and ultrasonography (US). Patient 1 only underwent regular MRI and CT, while Patients 2 and 3 underwent both regular and enhanced MRI and CT. Imaging features of the lesions were characterised according to size, margin, signal intensity, and degree of enhancement. The density and signal intensity were documented as hypo-, iso-, or hyper-, respectively, relative to those of the muscle. The review of the imaging data (CT/MRI) was completed by four senior otolaryngologists who each have over 10 years of experience. Pathological sections were stained with haematoxylin and eosin (H&E). Immunohistochemical staining (IHC) results for smooth muscle actin (SMA), catenin, desmin, S-100, CD34, and CD68 were documented. Fluorescence in situ hybridisation (FISH) for ubiquitin-specific peptidase 6 (USP6) rearrangement was performed, and a USP6 gene two-colour separation probe was used.

**Results**

**Patient 1**

A 3-year-old girl presented with a painless mass in front of the right ear that had been present for 1.5 years (Fig. 1). Regular anti-infection treatment was ineffective. The mass was biopsied before surgery, and histological examination revealed NF. Temporal bone CT showed an irregular soft tissue mass with uniform density in the anterior and inferior parts of the right external auditory canal. The mass exhibited hypo-intensity on T1-weighted imaging (T1WI) relative to skeletal muscle. Hyper-intense signals were observed on T2-weighted imaging (T2WI) and short tau inversion recovery (STIR) sequences. US examination revealed a 41 × 21 × 33 mm ovoid hypoechoic nodule with distinct margins.

The child underwent surgery under general anaesthesia. A longitudinal incision was made along the surface of the preauricular bulge. The subcutaneous tissue was then cut, and the mass was exposed. The border of the mass was distinct to the naked eye. After complete resection, the incision was sutured intradermally, and the specimen was sent for pathological examination. Histopathologically, the lesion was compatible with the diagnosis of NF. IHC staining indicated positivity for SMA and catenin, focal positivity for CD68, 10% positivity for Ki-67, and negativity for CD34, S-100, and desmin. There was no obvious scar in the incision after the operation and no recurrence 31 months after the operation, as confirmed by MR imaging.

**Patient 2**

A 17-month-old boy was admitted to our department with a painless mass in his right pinna, which had been present for 4 months (Fig. 2). He underwent two surgeries to remove the mass before admission. The pathological analysis revealed that the mass was a myofibroblastic tumour. After the second surgery, the wound did not heal, and the mass reappeared and grew rapidly. Anti-infection treatment was administered during this period, although it was ineffective. MRI performed prior to admission demonstrated an iso-intense nodule on T1WI, which was hyper-intense on T2WI and STIR sequences. Enhanced MRI showed heterogeneous enhancement at the edge of the lesion, and no enhancement within the lesion. US examination revealed a solid mass with irregular hypoechogeticity (uneven internal echo). The mass was approximately 30 × 18 × 28 mm in size.

A fusiform incision was made along the tumour on the posterior aspect of the pinna. We observed that the lesion had an incomplete capsule and exhibited a rotten fish-like appearance and crisp texture. The mass did not penetrate...
the fascia layer behind the ear, although it invaded the cartilage of the cavum concha and the skin of the postero-inferior wall of the external acoustic meatus. During the operation, we removed part of the cartilage and the shaved part of the perichondrium behind the ear. The involved skin was then removed. The incisional margins were sent for frozen pathology during the operation, and no NF tissue was found until triamcinolone acetonide was then injected into the surgical cavity. Tension-reducing sutures were fashioned at the retroauricular sulcus. The auricular cavity flap was rotated to repair the skin defect of the posterior inferior wall of the external acoustic meatus and enlarge the external acoustic meatus (Fig. 3). The external acoustic meatus was then tightly obturated with a piece of gauze coated with antibiotic ointment. Postoperative pathology confirmed the diagnosis of NF. IHC staining indicated positivity for SMA, focal positivity for CD68, 15% positivity for Ki-67, and negativity for CD34, desmin, and S-100. FISH test results showed that 34% of the 100 cells exhibited \textit{USP6} gene rearrangement. Nine months after the operation, the patient exhibited good recovery with a good reconstruction of the appearance of the ear. To date, no recurrence has been reported.

**Patient 3**

A 19-month-old girl presented with a painless and progressive mass at the external orifice of the left ear, which had been present for 6 months (Fig. 4). The mass had ruptured and bled during the last month. A biopsy was performed at another hospital, and the pathological
results suggested an inflammatory granuloma. The mass showed signs of enlargement after biopsy. Temporal bone CT showed an oval low-density tissue mass in the left auricle with surrounding soft tissue thickening. MRI features included hypo-intensity relative to skeletal muscle on T1WI and hyper-intensity on T2WI or STIR sequences, with heterogeneous enhancement after contrast injection. US indicated that the entire lesion was approximately $17 \times 16 \times 9$ mm in size. Although the lesion did not exhibit any intrinsic vascularity, the surrounding soft tissue exhibited increased vascularity on Doppler US examination.

![Fig. 2 Findings in Patient 2. a, b Pre-operative clinical photographs showing a mass in the cavum conchae and back of the auricle. c Axial T2-weighted image (T2WI) showing high signal intensity and a “fascial tail” sign (arrow). d Photograph of the ear 5 months after the operation showing an intact ear appearance. e Histological changes associated with nodular fasciitis (haematoxylin and eosin [H&E] stain, ×200). f Immunohistochemistry revealed positivity for smooth muscle actin (SMA). g Fluorescence in situ hybridisation (FISH) showing rearrangements of the ubiquitin-specific protease 6 (USP6) locus (separation of green and red signals).]

![Fig. 3 In the leftmost diagram, the sawtooth line represents the boundary between normal skin and diseased tissue. The middle picture panel shows the process of removing the diseased tissue and the invaded cartilage, and the shadow represents the cartilage of the auricular cavity. The last panel shows the reconstructed auricular cavity and the enlarged external auditory canal created using the rotating flap.]

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A surgical incision was made along the edge of the mass. During the operation, we observed that the tumour had an incomplete capsule, and that the texture was solid. The incisional margins were sent for frozen pathology, and the lesion was completely resected. The skin of the mastoid area was taken as a full-thickness free skin flap and transplanted into the skin defect of the incisura intertragica, following which a strip of gauze containing antibiotic ointment was applied. IHC staining indicated positivity for SMA and CD68, 10% positivity for Ki-67, and negativity for CD34, S-100, and desmin. NF was diagnosed, and FISH test results showed that 33% of the 100 cells exhibited \textit{USP6} gene rearrangement. Two weeks after the operation, the gauze was removed, and we observed that the flap had survived well. Eight months after the operation, the patient had recovered well without recurrence.

Summary of cases

Table 1 summarises the clinical characteristics and examination data of the three children with ear NF (two girls, one boy). The average age at onset was 14 months, and the maximum diameters of the three lesions were 2.9 cm, 2.8 cm, and 1.7 cm, respectively. None of the three children had a history of trauma. Anti-inflammatory treatment was ineffective in all three patients. Patients 1 and 3 had a history of biopsy, and Patient 2 had a surgical history of mass resection. All three masses were surgically removed in our hospital. Regular follow-up was performed after the operation, and no cases of recurrence were observed. All patients exhibited an intact ear appearance at the last follow-up.

Discussion

NF is a relatively rare mass-forming and self-limiting subcutaneous form of pseudosarcomatous myofibroblastic proliferation. Paediatric NF around the ear is rare and difficult to distinguish from other soft tissue lesions. It is also easily mistaken for other inflammatory masses that have a higher incidence in children. Therefore, in this study, we analysed clinical manifestations, imaging results, pathological findings, treatment strategies, and postoperative follow-up results in paediatric patients with NF. As indicated above,
we observed that NF was initially misdiagnosed and treated without effect using antibiotics in all three of our patients. Our results ultimately indicate that combining clinical features with imaging findings may improve the accuracy of preoperative diagnosis in patients with NF, although genetic testing for USP6 can also be used to support diagnosis. Nonetheless, the final diagnosis should be based on comprehensive assessment.

In the 2013 World Health Organization (WHO) soft tissue tumour classification, NF was classified as fibroblast/myofibroblastoma [8]. Thompson et al. [9] reported the largest number of cases of external ear NF in 2001, in which patients had a mean age of 27.4 years (range: 1–74 years). The authors did not provide further details with respect to age. Our literature search indicated that most previous reports were case reports [6, 7, 10]. The present paper is significant in that it represents the only case series of NF of the ear in three children, with a complete medical history and prognosis.

The cause of this disease remains unclear. Previously, NF was thought to be related to local trauma. In 2011, Erickson-Johnson et al. reported that the mechanism of NF seems to involve transcriptional upregulation of USP6 due to its high level of fusion with myosin heavy chain 9 (MYH9) [11]. While this provides evidence for the clonal neoplastic origin of NF, further investigations in this field are required. In our study, none of the three children had a history of trauma. However, rapid enlargement after biopsy and reappearance and rapid growth after surgical resection were observed in Patients 3 and 2, respectively. We speculated that external stimuli may cause enlargement of the existing mass.

Radiologically, MR imaging of the ear can better evaluate the size, extent, and nature of the mass than CT. In most cases of classical NF, T1W1 shows hypo-intensity or isointensity relative to the adjacent skeletal muscles, while heterogenous hyper-intensity can be observed on T2W1 or STIR sequences [12, 13]. In this study, the signal characteristics of the lesions on MRI were similar to those reported in previous studies. In addition, the “fascial tail”—which was observed in all three of our patients—is valuable for the diagnosis of NF [14]. The fascial tail sign is characterised as thickening of the adjacent fascial layer with obvious linear or tail-shaped enhancement on MR images. However, the fascial tail sign is not specific to NF, and some malignant soft tissue tumours are also associated with tail signs [15]. However, the relatively specific clinical manifestations of NF combined with the fascial tail sign on MRI can help to promote diagnosis of NF.

Histologically, NF involves proliferative, tumour-like, immature fibroblastic lesions with rich cellularity and mitotic activity. Due to these features, NF is often misdiagnosed as sarcoma. Furthermore, Erickson-Johnson et al. [11] first reported USP6 gene rearrangement on chromosome 17p13 as a specific finding in NF in 2011. They also reported a sensitivity of 93% and specificity of 100% for USP6 rearrangements as diagnostic markers for NF, as confirmed by FISH. The result was considered positive if >10% of 200 cells showed split signals [11, 16]. At present, USP6 rearrangement and USP6 fusion genes can be used as diagnostic tools for NF. Two of our three patients were positive for rearrangement of the USP6 gene, although the test was not performed in the remaining patient.

Treatment options for NF include observation and surgical resection. Local resection is an effective treatment, with a recurrence rate of only 1–2% [17]. According to Thompson et al. [9], NF has a higher propensity for local recurrence in

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Table 1  Clinical characteristics of three patients

|                | Patient 1 | Patient 2 | Patient 3 |
|----------------|-----------|-----------|-----------|
| Age¹, month/sex| 36/G      | 17/B      | 19/G      |
| Side           | Right     | Right     | Left      |
| Location       | Preauricular | Pinna     | External auditory meatus |
| Diameter², cm  | 2.9       | 2.8       | 1.7       |
| Course, month  | 18        | 4         | 6         |
| Trauma history | Without   | Without   | Without   |
| Clinical features of the mass³ | Stiff, painless | Stiff, painless, and rapid growth | Stiff, painless, and rapid growth |
| Plain/enhanced CT scan | Yes/No | Yes/Yes (Other hospital) | Yes/Yes (Other hospital) |
| Regular/enhanced MR scan | Yes/No | Yes/Yes | Yes/Yes |
| Sonography     | Yes       | Yes       | Yes       |
| FISH for USP6  | No        | Yes       | Yes       |
| Follow-up, month/recurrence | 31/No | 9/No | 8/No |

¹ Age at diagnosis  
² Maximum diameter on MRI  
³ Clinical features of the mass when it was first observed
the auricular area (9.3%). They explained that this occurs due to increased auricular trauma and the difficulty in ensuring complete excision due to the anatomical position of the lesion. Our three children were followed up for 31, 9, and 8 months, and no recurrence was observed in any patient. Some authors have reported regression of NF after fine-needle aspiration biopsy [18]. Even without excision, spontaneous regression has also been observed [19]. We recommend conservative observation for a period when the patient exhibits good function and appearance. However, once the deeper tissue has been invaded, complete surgical resection should be performed. Surgical resection can cause ear skin defects, and it is particularly important to reconstruct ear appearance in children. As the first child in our series had no obvious skin defects, we adopted a counter-subcutaneous suture. In Patient 2, we utilised a local rotating skin flap in the auricular cavity, while full-thickness skin transplantation was employed in Patient 3. All three children experienced exceptional cosmetic results.

Conclusion

NF should be considered in the differential diagnosis of young children presenting with rapidly enlarging painless masses around the ear who have failed to respond to anti-infection treatment. The combination of clinical manifestations, imaging findings, and preoperative biopsy results can reduce the misdiagnosis rate. For ear NF, the significance of preserving or reconstructing local appearance is higher than that for lesions at other positions, making it necessary to consider the patient’s prognosis and appearance during treatment. We found that complete local resection can be performed without substantial defects in ear appearance.

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Authors’ contributions  All authors contributed to the conception and design of the study. Material preparation, data collection, and analysis were performed by Zhang, Wang, Liu, He, Chen, Shao, Zhang, Ma, and Li. The first draft of the manuscript was written by Xiaoxu Wang, and all authors commented on the subsequent versions of the manuscript. All authors read and approved the final manuscript.

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Availability of data and material  All data and materials used are reported in the current manuscript.

Code availability  Not applicable.

Declarations

Ethics approval  Ethical approval was waived by the local ethics committee of Beijing Children’s Hospital at Capital Medical University given the retrospective nature of the study and because all procedures were performed as part of routine care.

Consent to participate  Informed consent was obtained from the parents/guardians of all participants included in the study.

Consent for publication  The parents/guardians of participants consented to the submission of the original article to the journal.

Conflict of interest  The authors declare no competing interests.

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