

## Case Report

# Recurrent nodular fasciitis of the external auditory canal: an intriguing diagnosis

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### ABSTRACT

The external auditory canal can sometimes present with unique pathology since it is a vibrant environment possessing a variety of anatomical structures including bone, cartilage, skin, ceruminous glands and fibro-collagen tissues. Nodular fasciitis is an intriguing entity which rarely presents in the external ear canal as a growth. The diagnosis is made only through histopathological confirmation after ruling out similar benign or malignant lesions including ceruminoma, granuloma or carcinoma. Fasciitis being of fibrous origin has propensity to recur wherein management becomes more challenging. This is a clinical profile of a case of recurrent nodular fasciitis which was surgically managed and followed up with no further recurrence.

**Keywords:** Nodular fasciitis, External auditory canal

### INTRODUCTION

Nodular fasciitis (NF) is a benign myofibroblastic proliferation, which presents as a rapidly growing mass resembling an aggressive lesion clinically.<sup>1</sup> Among all pathologic diagnosis, NF contributes to 0.025%. NF was first described by Konwaler as subcutaneous pseudosarcomatous fibromatosis.<sup>2</sup> Although many hypothesis have been postulated that subtle trauma is an etiological factor, however the exact etiology still remains unknown.<sup>3</sup> Extremities are the most common site of NF, followed by the chest and trunk. Although external auditory canal is a common site in paediatric population, head and neck region contributes to only in 7% to 20% in the adult population which includes the parotid region, forehead, zygoma, cheek, periorbital area, eyelid, and intraoral sites.<sup>4</sup>

NF often mimics a malignancy histopathologically, most common a sarcoma due to its features like increased cellularity and mitotic activity, rapid growth and ill-defined margins. Due to these features resembling a

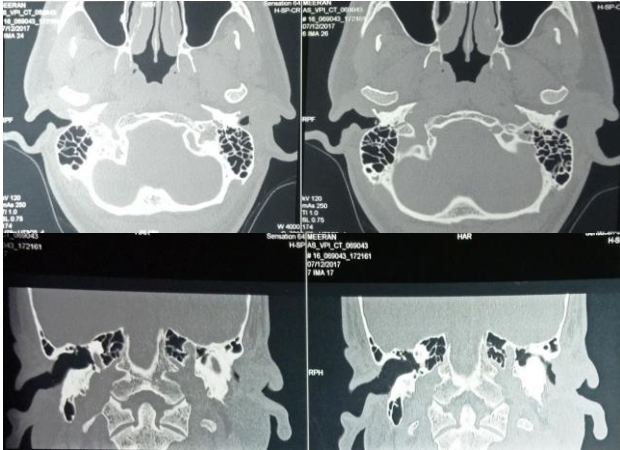
malignancy, aggressive treatment is often employed which causes increased physical and psychological morbidity to a patient. Thus, definitive diagnosis of NF must be made based on histopathological and clinical findings prior to treatment.<sup>5</sup> We report here an intriguing case of a middle-aged man who presented with a recurrent external auditory canal mass with a previous histopathological diagnosis of fibrous myxoid tumour, however, the histological diagnosis was that of a classical nodular fasciitis on revision surgery.

### CASE REPORT

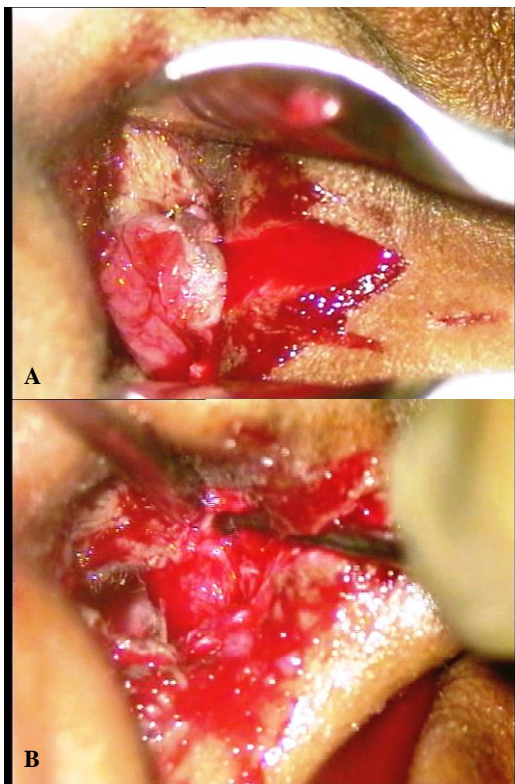
A 37 year old male presented to our institution with a rapidly enlarging swelling in the left ear canal for past 3 months. He had a previous history of similar complaint in the past 7 months for which excision biopsy of the mass was done 4 months ago and now recurrence happened at the same site soon after surgery. Histopathological report had previously showed fragments of tumour tissue with fibro-myxoid degeneration along with areas of congestion

and chronic inflammatory cell infiltration. There was no evidence of malignancy in this report.

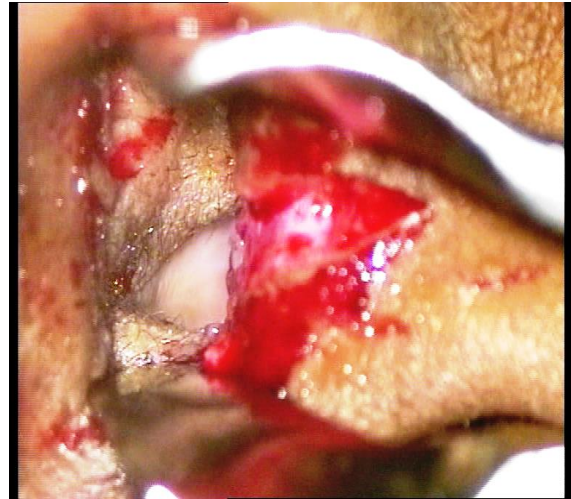
Computed tomography of temporal bone was done to assess the extent of this recurrent lesion. This showed a lobulated soft density mass lesion in the confines of the left external auditory canal causing distension of the same. No imaging features of bony or cartilagenous involvement was found (Figure 1).



**Figure 1:** CT images with soft tissue mass seen completely occluding the left external auditory meatus.



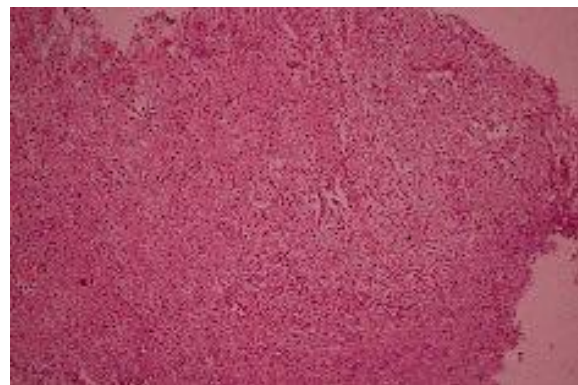
**Figure 2 (A and B):** Warty irregular mass lesion seen occluding the left EAC which was approached through a Lempert type 2 end-aural incision.



**Figure 3:** Scraping of tumour pedicle attached to the underlying perichondrium of the conchal cartilage.



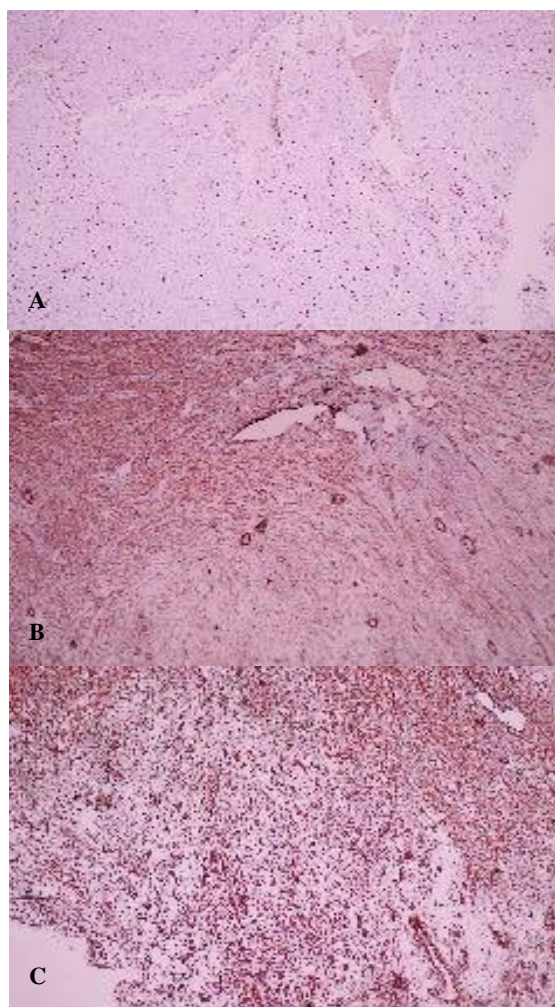
**Figure 4:** Left EAC after complete wide excision of mass. Note that tympanic membrane and bony ear canal are not involved by the lesion.



**Figure 5:** Histopathological examination under high power - fibromyxoid cellularity with collagen matrix and inflammatory infiltrates.

On examination the mass was warty, firm and non-tender, completely occluding the left external auditory canal.

Tympanic membrane was not visualized. The rest of ears, head and neck were unremarkable. There was no palpable cervical lymph nodes. The patient had moderate to severe conductive hearing loss in this ear. The contralateral ear was unremarkable.



**Figure 6 (A-C): Immunohistochemistry panel: Ki 67, smooth muscle actin and vimentin positivity - pathognomonic for nodular fasciitis.**

Thus with a diagnosis of recurrent inflammatory myofibroblastic tumour, surgery was planned for endaural complete excision of the mass along with a tumour free margin. Intra-operatively the mass had a pedicle which was attached to the posterior superior wall extending up to the bony cartilaginous junction (Figure 2). A Lempert's type 2 incision was made and complete excision of the mass was done en-block along with the perichondrium and skin in its base (Figure 3). The bony EAC and tympanic membrane was found to be intact (Figure 4). Subcutaneous approximation was done as possible with vicryl sutures in order to cover the exposed cartilage. A betadine pack was applied in the ear canal over the remaining raw areas to stimulate healing of the ear canal skin by secondary intention. The patient recovered uneventfully over the next two weeks.

On histopathology, grossly the mass was irregular, fragile and grey brown in colour measuring 1.3×0.7×0.5 cms. The grossed cut-surface was grey white and firm. Microscopic examination revealed hyperkeratotic epidermis with underlying dermis showing spindle shaped cells in a myxoid edematous stroma with no cellular atypia (Figure 5). Immunohistochemistry was recommended and this showed that the spindle cells were positive for Vimentin, smooth muscle actin and Ki-67 (Figure 6) and it was negative for Desmin, Myogenin and CD-34. Such a presentation is pathognomonic of nodular fasciitis and hence clinched the diagnosis. The patient was counselled regarding the same and meticulously followed up over the past 18 months with no recurrence till date.

## DISCUSSION

Nodular fasciitis of the external auditory canal is a very rare pseudo-tumour with only less than 200 cases have been reported till date in world. NF is a diagnosis of exclusion after exclusion of every other condition based on immunohistochemical studies.<sup>6</sup>

Nodular fasciitis is often diagnosed as an malignant lesion due to its rapid growth. Histopathologically the lesion vary from mild cellular atypia with moderate cellularity and mitosis to a malignant tumor (carcinoma in-situ, especially ceruminocarcinoma). Thus it is very prudent to diagnose nodular fasciitis correctly to avoid additional surgery and radiotherapy.<sup>7</sup>

Histologically, the differential diagnosis for NF include spindle cell spindle cell tumor-like lesions which include inflammatory pseudotumor, reactive spindle cell nodule after biopsy, fascicular variant of pseudoangiomatous stromal hyperplasia and tumors which include myofibroblastoma, benign fibroblastic spindle cell tumor, leiomyoma, schwannoma, spindle cell lipoma, solitary fibrous tumor and myxoma posing a challenging quest for the pathologist to arrive to a diagnosis of nodular fasciitis.<sup>8</sup> NF is composed of myofibroblasts arranged in a fascicular or storiform pattern with varying cellularity which are positive for smooth muscle actin, but negative for desmin and caldesmon. Immunostains for CD34, S-100, ALK, keratin and p53 are also negative. Cytoplasmic staining for  $\beta$ -catenin is seen in one-third of the cases.<sup>9</sup>

Local recurrences are also more common due the nature of the lesion and incomplete surgical excision due to its location and surgical inaccessibility surrounding the ear canal. More rarely, frequent recurrences may transform the lesion to fibrosarcoma. A few case reports have described NF of the auricle. The largest series of nodular fasciitis has been reported by Thompson LD et al, who in their retrospective study mentioned fifty cases of NF of the auricular region which included 22 females and 28 males, aged 1 to 76 years among whom only 5 patients recalled antecedent trauma. All patients had surgical

excision of which four patients (9.3%) developed local recurrence and were alive and disease free at last follow-up. Local recurrence was due to the incomplete surgical excision due to inaccessibility around the ear.<sup>10</sup>

Subtle trauma which triggers fibro-myxoid proliferation is proposed as an causative factor for NF but the exact aetiology is still.<sup>11</sup> Head and neck contributed to only about 13-20% of all cases of NF and are more common in third to fifth decades of life with no definite gender predilection.<sup>12</sup> NF has known to manifest in a variety of head and areas as literature has cited this in peri-orbita, parotid, neck, face, auricle, temple, post-aural and buccal regions.<sup>13</sup>

Recent research has identified the *MYH9-USP6* fusion gene as a tumor marker for nodular fasciitis. This gene provides evidence for clonal neoplastic origin of nodular fasciitis and suggests the basis of a new theory of locally aggressive neoplasm-like transformation of soft tissues in response to triggers like trauma, also called as “*transient neoplasia*”. Thereby apart from immunohistochemistry, gene detection may become clinically useful as a diagnostic tool for nodular fasciitis in the future.<sup>14-16</sup>

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