B-ENT, 2010, **6**, 131-133

Nodular fasciitis of the external auditory canal

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Key-words. Nodular fasciitis; external auditory canal

Abstract. *Nodular fasciitis of the external auditory canal.*
Problem: Nodular fasciitis (NF) is a reactive myofibroblastic proliferation that may be misdiagnosed as a sarcoma because of its rapid growth, rich cellularity, and mitotic activity. NF is uncommon in the auricular region. We describe a case of nodular fasciitis of the external auditory canal in an 8-year-old male.
Methodology: An excisional biopsy was performed, and the pathologic examination was consistent with nodular fasciitis; however, the lesion recurred within a month requiring that a total excision be done.
Results: The patient is without recurrence 7 months after the total excision.
Conclusions: Because its histologic features closely mimick a malignant lesion, NF must be considered in the differential diagnosis of a mass originating from the external auditory canal to avoid overly aggressive and functionally debilitating treatment.

Introduction

Nodular fasciitis (NF) is a benign, self-limiting pseudosarcomatous reactive process composed of fibroblasts and myofibroblasts. It typically occurs on the upper and lower extremities of adults, particularly on the forearm. Although 7-20% of these tumours are located in the head and neck region, involvement of the ear is extremely rare.[1] We present a case of NF arising in the left external auditory canal.

Case report

An 8-year-old male was admitted to our Otolaryngology Department with a 2-week history of a mass protruding from his left ear. He had a history of trauma at the site of the lesion 3 weeks earlier. The mass was elastic and hemorrhagic, and measured 1.7 cm × 1 cm × 1 cm (Figure 1). An examination of the head and neck revealed no other abnormality. Computed tomography of the temporal bone showed a well-defined, superficial soft-tissue mass in the left EAC (Figure 2). An excisional biopsy was performed, and the pathologic examination was consistent with nodular fasciitis; however, there were positive surgical margins and the lesion recurred within a month. A total excision was performed via a postauricular incision.

The lesion originated from the cartilaginous part of the inferior canal wall. It was resected with wide surgical margins, including its base at the cartilage. The ear canal was reconstructed with a posterior ear canal skin flap. Histopathological analysis of the mass showed a fibrous appearence with spindle cell proliferation. Extravasated red blood cells were present throughout the lesion and mitotic cells were present, but without signs of atypia (Figure 3). The pattern of the lesion resembled tissue culture fibroblasts in the focal areas. Foci of metaplastic bone and osteoclast-type giant cells were also observed. The cells were reactive for smooth muscle actin (Figure 4) and vimentin, and negative for desmin, S-100 protein, CD34, and myoglobulin (not shown). The patient was without recurrence 7 months after a total excision.

Discussion

NF was first described by Konwaler *et al.*[2] in 1955 as a "subcutaneous pseudosarcomatous fibromatosis". It is an uncommon pseudosarcomatous proliferation of myofibroblasts that is difficult to differentiate from its malignant counterparts.

NF is a reactive process of unknown pathogenesis rather than a neoplastic process. Trauma was once hypothesized to play a role in the pathogenesis of NF because of its histologic characteristics that often resemble organizing granulation tissue. A history of trauma can be found in 5-15% cases.[3,4]

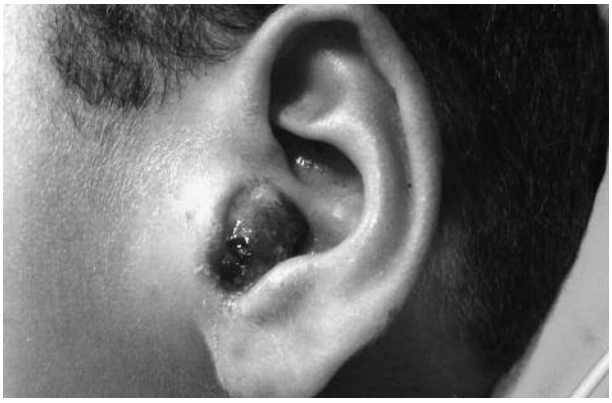

Figure 1
A mass protruding from the left external auditory canal

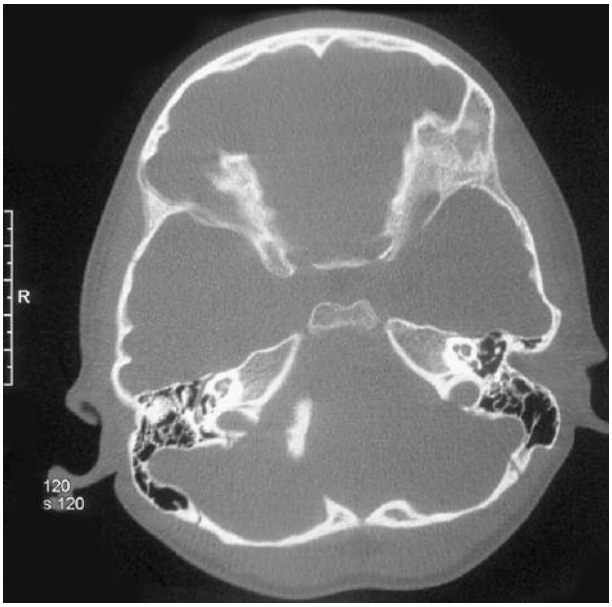

Figure 2
Axial CT scan showing a soft tissue mass filling the left external auditory canal.

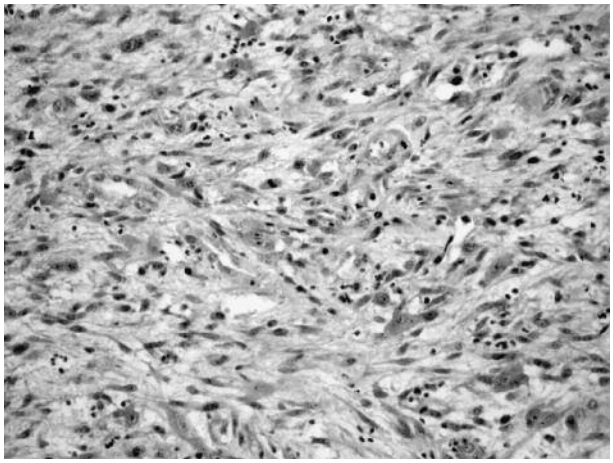

Figure 3
Spindle cells show mitotic activity without nuclear atypia. Also shown are extravasated erythrocytes.

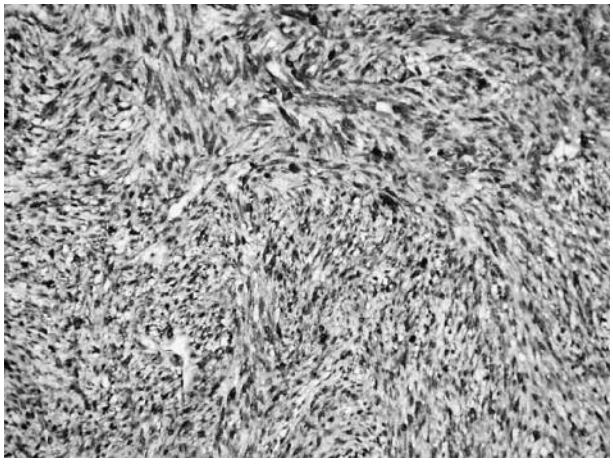

Figure 4
Cells staining intensely for smooth muscle actin

NF is most commonly seen in the upper extremities. A total of 10-20% of these lesions are located in the head and neck region, with involvement of the ear being extremely rare. In children, however, the head and neck region is the most common site, and recurrence is more common than in adults.[5] Thompson *et al.*[6] reported that ear lesions represented only 1.5% of all cases of nodular fasciitis diagnosed between 1970-1990, and that the frequency of the disease corresponds to 1.9% of all auricular lesions (reactive, benign, or malignant lesions). Although NF can occur in any age group, young adults between the age of 20 and 40 years are most commonly affected, with a slight male predominance. Typically, the lesion is located in the subcutaneous, fascial, or intramuscular regions.[1]

The diagnosis of NF is often a challange due to its sarcoma-like features. It is estimated that up to 20% of these lesions are misdiagnosed as sarcomas.[7] The differential diagnosis includes fibrosarcoma, leiomyosarcoma, and malignant fibrous histiocytoma because of its rapid growth, rich cellularity, and abundant mitotic figures. However, the histologic pattern, absence of nuclear atypia, and atypical mitosis separate NF from malignant tumours. More specifically to the ear, clinical misdiagnoses might include

necrotizing fasciitis, perichondritis, rhabdomyosarcoma, or fibrous dysplasia. However, an accurate diagnosis of NF can be made through its unique histopathologic and immunohistochemical features.[6,8]

The lesion is curable with adequate surgical treatment. The local recurrence rate (9.3%) in the auricular region is higher than in other regions (1-2%).[1,6] The increased recurrence rate is mostly because of the difficulty in obtaining complete surgical excision because of the anatomy of the auricular region and/or the external auditory canal. Thompson *et al*.[6] reported that recurrences were often described within a few months of the initial presentation. Due to an incomplete excision, our patient presented with residual disease rather than with a recurrence.[6] Abdel-Aziz *et al*.[9] reported recurrences in two of six cases located in the EAC. Our patient is without recurrence 7 months after the total excision.

Conclusion

Because it has histologic features that closely mimick a malignant lesion, nodular fasciitis must be considered in the differential diagnosis of a mass originating from the EAC to avoid overly aggressive and functionally debilitating treatment.

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