Introduction

Nodular fasciitis (NF), a benign reactive myofibrolastic proliferation, is an uncommon lesion in the external auditory canal (EAC) [1-3]. NF has been first described by Konwaller et al. in 1955 as “subcutaneous pseudosarcomatous fibromatosis (fasciitis)” due to the possible clinical and histological confusion with a sarcoma. The lesion is self-limited and proper preoperative diagnosis is essential to avoid unnecessary aggressive treatment with dysfunctional and inesthetic outcome. The largest series has been reported by Thompson et al., which included 50 cases of NF of the auricular region identified in the files of the Otorhinolaryngologic-Head and Neck Tumor Registry of the Armed Forces Institute of Pathology [3]. There are some additional case reports [4-10] and a case series in English literature [11].

Case Report

An 8 years old male patient referred to outpatient department with a 2 weeks history of rapidly growing, hemorrhagic, painless mass protruding from the right ear canal. An incisional biopsy was already performed and the diagnosis was reported as “myxoma”.

Examination revealed a fungating, hemorrhagic polypoid lesion protruding from the right EAC. In a computerized tomography (CT) scan of the temporal bones (TB), the mass was confined to the right EAC without any invasion to the surrounding structures. Magnetic Resonance Imaging (MRI) showed an homogenous, well-defined soft tissue mass with a diameter of 1.5 cm (Figure 1). There was no evidence of involvement of the parotid gland, temporomandibular joint, masseter, pterygoid muscles, great vessels or lymph nodes. Audiological evaluation revealed a 50 dB conductive hearing loss in the right ear and normal hearing on the other side. Incisional biopsy was repeated under neuroleptic anesthesia. Histological diagnosis was reported as nodular fasciitis.

The patient was operated under general anesthesia. The lesion was totally removed through a retroauricular approach together with the partial cartilaginous EAC. The tumor was originated from the posterior wall of the cartilaginous portion of the canal. The tympanic bone and the middle ear were normal. The tympanic membrane was normal also.
Histologically the lesion was characterized by a mixture of plump myofibroblasts and fibroblast-like spindle cells, arranged in short, loosely textured bundles. Neither cytologic atypia nor atypical mitoses were observed. At immunohistochemistry the tumor cells were positively reacted to smooth muscle actin, suggesting myofibroblastic differentiation (Figures 2-3). Final pathologic diagnosis was confirmed as nodular fasciitis. The surgical margins were tumor free.

Figure 1. T2 weighted MRI scan (in coronal and axial planes) of the lesion (arrow)

Figure 2. Short and irregular spindle cell fascicles with scattered inflammatory cells and erythrocytes. HEx100
The healing of external auditory canal was completed at postoperative sixth week and the control audiometry was normal. No recurrence was detected up to 28 months follow-up.

Discussion
NF is a benign, self-limited, fibroproliferative disease thought to be a reactive process rather than a true neoplasm [1]. It is most commonly seen in the upper extremities. A total of 10% to 20% of lesions occur in the head and neck region with the predominant sites being the neck and face. Its cause is unknown; however, an episode of trauma can be recalled in less than 15% of cases [12]. The age at diagnosis in most individuals is between 20 to 50 years, with male predominance [2]. Although NF of head and neck is common, particularly its localization in the EAC is unusual in children.

The largest series of NF involving the external ear region has been reported by Thompson et al. It included 50 cases identified in the files of the Otorhinolaryngologic-Head and Neck Tumor Registry of the Armed Forces Institute of Pathology. They were 22 females and 28 males with ages ranging from 1 to 76 years (mean: 27.4 years). Only 6 among these cases had a tumor located in the EAC [3]. Ninety-eight percent of the patients presented clinically with a mass lesion; one patient showed only a conductive hearing loss. Five patients reported pain and bleeding from the ear. The duration of symptoms ranged from a few days to 36 months, with a mean of 5 months. Tumor size ranged from 0.4 to 8 cm with a mean of 1.9. The sites of origin were mainly the postauricular (19 cases) and preauricular areas (18 cases) while 6 cases only were reported in the EAC. NF can arise in the dermal, subcutaneous or fascial layers. The subcutaneous

Figure 3. Smooth muscle actin positivity of neoplastic cells. SMAx100
Nodular Fasciitis of the External Auditory Canal

tissue is the most common site of origin throughout the body, except for the head and neck region, which, interestingly, shows a predominance of the dermal origin. In our case, the lesion was subcutaneous.

All of the lesions in Thompson et al. series (9) were surgically excised; a local recurrence developed in 9.3% of the patients. However, the specific origin (dermal, subcutaneous or fascial), the anatomic site, bleeding and/or ulceration, patients’ age, neural entrapment, skeletal muscle entrapment/atrophy and size of the lesion were not correlated with an increased chance of recurrent or residual disease. There is a higher local recurrence likelihood in the auricular region than in other areas (9.3% vs. 2%-1%, respectively), that has been attributed to the difficulty in obtaining a complete surgical excision because of the anatomy of the auricular region.

At the review of English literature there are some additional case reports and a case series about NF of auricular region (4-11).

The surgeon should plan a careful and conservative resection of this benign tumor, as this lesion is self-limiting. Proper preoperative assessment is essential to avoid unnecessary aggressive treatment, with possible negative functional and aesthetic outcome.

Although CT and MRI are often used in the evaluation of nodular fasciitis, there is no specific finding. On CT and MRI, NF is seen as a relatively well-defined, soft tissue mass of superficial location. Deep-seated lesions, mostly of intramuscular type, tend to be large and have ill-defined margins. Although this lesions are histologically benign, the deep located lesions can invade and destroy the adjacent structures. In our case, the radiologic margins of the lesion was well defined. Various signal intensities have been reported for MRI of NF, reflecting the variable cellularity, the amount of collagen of cytoplasm and water content in the extracellular space and the vascularity of the lesion.

Macroscopically, NF is a nonencapsulated, tan to gray-white, round or oval mass. Microscopically, it is characterized by plump, immature appearing fibroblasts and a variable amount of collagen fibers. The fibroblasts are arranged in short irregular bundles and fascicles and are accompanied by an attenuated reticulin meshwork. Mitotic figures without atypical mitoses are common. Some reports show that rapid growth along with the histologic characteristics of high cellularity and increased mitotic activity of NF can lead to initial misdiagnosis of sarcoma (2,6,12). The differential diagnosis of nodular fasciitis includes neurofibroma, neurilemoma, fibrous histiocytoma, fibromatosis, desmin tumors, myxofibrosarcoma, myxoid liposarcoma, spindle cell lipoma and other soft tissue sarcomas.

The patient presented here was an additional case to the rarely seen auricular NF group. In rapidly growing, hemorrhagic neoplastic lesions; this rare entity should be included in the differential diagnosis. Our surgical experience although limited to this only patient highlights the importance of determining the origin of the lesion, that was the cartilage of the posterior EAC.

Preservation of the tumor-free surrounding structures achieves satisfactory anatomical and functional result.

References


