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Clinical Records

Solitary adult myofibroma of the pinna

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Abstract
Solitary myofibroma is a recently described benign neoplasm of the skin or superficial soft tissue and it represents the adult counterpart of infantile myofibromatosis. This new clinicopathological entity is being recognized increasingly. A case of solitary myofibroma occurring in the pinna of a 50-year-old woman is presented. Such a lesion occurring in the pinna of an adult has not been reported in the literature.

Key words: Myofibroma; Ear, external

Introduction

Infantile myofibromatosis was first described in 1954 by Scout as 'congenital generalized fibromatosis' (Walsh *et al.*, 1996) and since then numerous examples of this entity have been reported in the medical literature. Even though it has been variously described as generalized hamartosis, multiple congenital mesenchymal hamartoma, multiple vascular leiomyoma, or metastatic congenital fibrosarcoma in the literature, the term infantile myofibromatosis has been preferred. This is due to the microscopic resemblance to smooth muscle tissue, the electron microscopic findings and the need to clearly distinguish this process from the locally more aggressive desmoid type of infantile fibromatosis.

Myofibroma presents as small, solitary or multiple (myofibromatosis), dermal, subcutaneous or sometimes deep intramuscular nodules, usually in small children; rarely, similar lesions have been noted in adults (Chung and Enzinger, 1981; Daimaru *et al.*, 1989). The terms infantile myofibroma and infantile myofibromatosis have been employed to describe the solitary and multicentric forms, respectively. The solitary forms are, at least, twice more common than the multiple forms. The occurrence of such lesions in adults has only recently been recognized.

Case report

A 50-year-old female presented to the Department of Otolaryngology and Head and Neck Surgery of Kasturba Hospital, Manipal, with a four-month history of a painful swelling on the helix of the left pinna. This was of insiduous onset and progressive in nature, and there was no past history of trauma. She was a known diabetic on treatment. On examination, a tender nodule was noted which was 'raised scar-like' with stretching of the overlying skin giving it a 'shiny' appearance. The nodule appeared firm and fixed to the underlying cartilage. The blood parameters were normal and the blood sugar was under control. A provisional diagnosis of an inflammatory nodule secondary to localized pericondritis was made as the lesion was tender and the patient was a diabetic. A course of injectable antibiotics - ciprofloxacin and metronidazole was tried for one week with no response. Under cover of antibiotics, an excision biopsy of the nodule was performed with primary reconstruction.

Histopathological examination revealed a picture of a myofibroma with haemangiopericytoma-like pattern and haphazardly arranged fascicles of long spindle-shaped cells with elongated nuclei around dilated vascular spaces (Figure 1). The resected margins were not free of tumour. Immunohistochemistry was not carried out.

On review two months later, the patient reported recurrence of pain in the area of the scar. No nodule formation was seen even though the scar was found to be tender. Wider excision (Figure 2) of the tender area was done, including underlying cartilage, as the previous

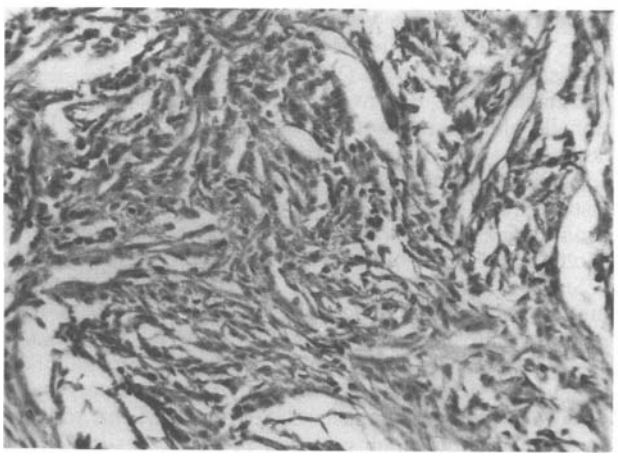

Fig. 1
Histological section of myofibroma showing haemangiopericytoma-like pattern with spindle cells around dilated vascular spaces (H & E; × 400).

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FIG. 2
Diagram of left pinna showing the site of adult myofibroma and wedge resection.

resected specimen was reported to have positive margins. This specimen, on histological examination, was reported as myofibroma with adequate margins of resection. On follow-up after one year, there was no complaint of pain, or any evidence of recurrence of the lesion.

Discussion

The commonest sites in the head and neck for myofibroma are the skull, scalp, forehead and oral cavity (Chung and Enzinger, 1981; Sleeman and Eveson, 1991; Speight *et al.*, 1991; Vigneswaran *et al.*, 1992). Isolated cases have been reported in the ear canal (Hogan and Salassa, 1992) and nose (Walsh *et al.*, 1996).

Clinically, the lesions present as painless, firm, occasionally bluish, cutaneous or subcutaneous nodules. Laaff described a tumour macroscopically resembling basal cell epithelioma in a 17-year-old female which could not be histologically distinguished from an infantile myofibroma (Laaff *et al.*, 1990). A similar lesion has been described by Wolfe in a 49-year-old woman (Wolfe and Cooper, 1990).

Smith *et al.* (1989) presented 34 cases with acquired skin tumour which histologically were identical to infantile myofibromatosis. Of the 34 cases, 26 were over 14 years of age with a median age of 36. The lesions were essentially benign and none of the lesions recurred after surgery or showed evidence of metastasis. He proposed this to be of vascular origin. Beham reported 11 new cases in 1993 in the age range of 13–64 years. They presented as solitary painless nodules of variable duration in the skin or oral cavity (Beham *et al.*, 1993).

Calonje and Fletcher (1993), stressed the need for accurate recognition and classification of soft tissue tumours to provide appropriate clinical care based on reasoned prediction of the behaviour of a given lesion. He characterized all dermal connective tissue lesions including adult myofibroma. Histologically, the lesion displays a distinct zoning phenomenon with a biphasic pattern. At the periphery, the tumour consists of plump spindle cells that often show a pronounced eosinophilia and are arranged in short bundles forming a fascicular or whorled pattern. More centrally and blending with the peripheral spindle-cell areas are frequently differentiated cells that are round or polygonal, arranged around small vessels forming haemangiopericytoma-like areas (Beham *et al.*, 1993; Walsh *et al.*, 1996). The characteristic zonation of the cells is often less marked in adult lesions compared to the infantile forms and a haphazard arrangement of the fascicular and pericytic areas have been described in some cases. Hyalinization of the spindle-cell areas, especially at the periphery, has also been described (Wolfe, 1990; Beham *et al.*, 1993). The lesion is often surrounded by compressed atrophic muscle fibres which may also be invaded by proliferating fibroblasts. This microscopic invasion explains the possibility of recurrence (Miettinen and Weiss, 1996).

Kutzner *et al.* (1993) identified four tumour variants: (1) a leiomyoma type of hamartomatous composition, (2) spindle-cell type in hyalinized whorls, (3) haemangiopericytoma or glomus type with endothelium lined vascular spaces and (4) a biphasic type having myofibroblasts which may show spindle-cell or undifferentiated mesenchymal cell differentiation. Immunohistochemically, the spindle-cells are desmin negative but muscle actin positive (HHF-35 and IA4). The rounded cells are both desmin and actin negative (Hogan and Salassa, 1992).

Myofibroma can be misdiagnosed on histopathology as fibrosarcoma, infantile fibromatosis, fibrous histiocytoma, nodular fascitis, infantile haemangiopericytoma, leimyoma, leimyosarcoma and neural tumours (Chung and Enzinger, 1981; Hartig *et al.*, 1993).

It is necessary for otolaryngologists to be aware of this lesion which can occur in adults and is similar to infantile myofibromatosis. This rare entity can present at various sites in the head and neck where it is more common than at other sites. We have presented this case, as the pinna is a prior unreported site, and the mode of presentation as a tender nodule is contrary to previous cases reported in literature.

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