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Case Report

A case of pleomorphic fibroma on the nose

一例鼻上的多形性纖維瘤

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Pleomorphic fibroma is a benign fibrous tumour characterised by cytological atypia. It typically presents as polypoid or dome-shaped mass on the trunk or extremities. Histopathologically, it is characterised by pleomorphic, hyperchromatic cells or giant multinucleated cells embedded in a collagenous stroma in the dermis. Herein, we describe a 40-year-old man who had a polypoid nodule on the right ala of the nose which had been present for 20 years. Histopathological features were consistent with those of pleomorphic fibroma.

多形性纖維瘤是一種良性纖維瘤，特點是有著細胞異型。其典型的臨床表現爲軀幹或四肢上的息肉狀或圓頂狀腫塊。組織病理學上，它的特點是眞皮層有多形性的深染細胞或多核巨細胞被嵌入在膠原基質內。在此，我們描述了一名四十歲的男子在其右側鼻翼有一個息肉樣結節達二十年之久。此結節的組織病理學特點與多形性纖維瘤吻合。

Keywords: Nose, pleomorphic fibroma

關鍵詞：鼻、多形性纖維瘤

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Introduction

Pleomorphic fibroma is a rare benign fibroblastic tumour characterised by pleomorphic, hyperchromatic cells or giant multinucleated cells embedded in a collagenous stroma.[1] Most of the described lesions were located on the extremities and the trunk in former studies.[1,2] Rarely, pleomorphic fibroma may occur in other sites, including the scalp,[3] tendon sheath,[4] and subungual space.[5] It is a rare fibroblastic tumour with 17 well-documented articles reported worldwide and there have been only two cases of pleomorphic fibroma which occurred on the nose.[1] Herein, we add a case of pleomorphic fibroma that arose on the nose, an uncommon site.

Report of a case

A 40-year-old Korean man presented with a 20-year history of a polypoid nodule on the right ala of his nose. This tumour was slow growing and asymptomatic. He had removed it three times by himself but it recurred each time afterwards. On physical examination there was a 0.8 cm x 0.8 cm sized flesh-coloured pedunculated polypoid nodule on the right ala of the nose (Figure 1). The lesion was clinically diagnosed as soft fibroma and shave biopsy was performed. Histological examination revealed a well-circumscribed, pedunculated lesion, covered with intact epidermis (Figure 2a). The papillary and reticular dermis showed a hypocellular neoplasm with haphazard arrangement of thick collagen bundles mixed with partially loose stroma, and also dilated blood vessels (Figure 2b). The tumour cells were spindle or irregularly shaped cells with indistinct borders and floret-like giant cells. The nuclei were large, pleomorphic and hyperchromatic, but no mitotic figures were found (Figure 2c). Immunohistochemical staining showed that the tumour cells stained positively for vimentin, smooth muscle actin, CD34 but not for S-100 protein (Figures 3a-d). These findings were consistent with those of pleomorphic fibroma. There has been no recurrence for six months since the shave biopsy.

Discussion

Pleomorphic fibroma is a cutaneous tumour first described by Kamino et al in 1989.[1] It is characterised by cellular atypia and pleomorphism, with mitoses on rare occasions, but the architectural features show a benign appearance.[2] Pleomorphic fibroma typically presents as a polypoid or dome-shaped mass on the trunk or extremities of middle-aged to older adults of both genders.[1] Rarely, it may occur in other sites and there have been only seven cases of pleomorphic fibroma which occurred on the head including the forehead,[2] retroauricular area,[6] eyelid[7] and nose.[1]

The pathogenesis of pleomorphic fibroma remains unclear. The immunoreactivity of vimentin and actin, along with the abundance of collagen suggests that pleomorphic fibroma is a tumour that is of fibroblastic or myofibroblastic origin.[8] Also, the fact that some of the cells were stained

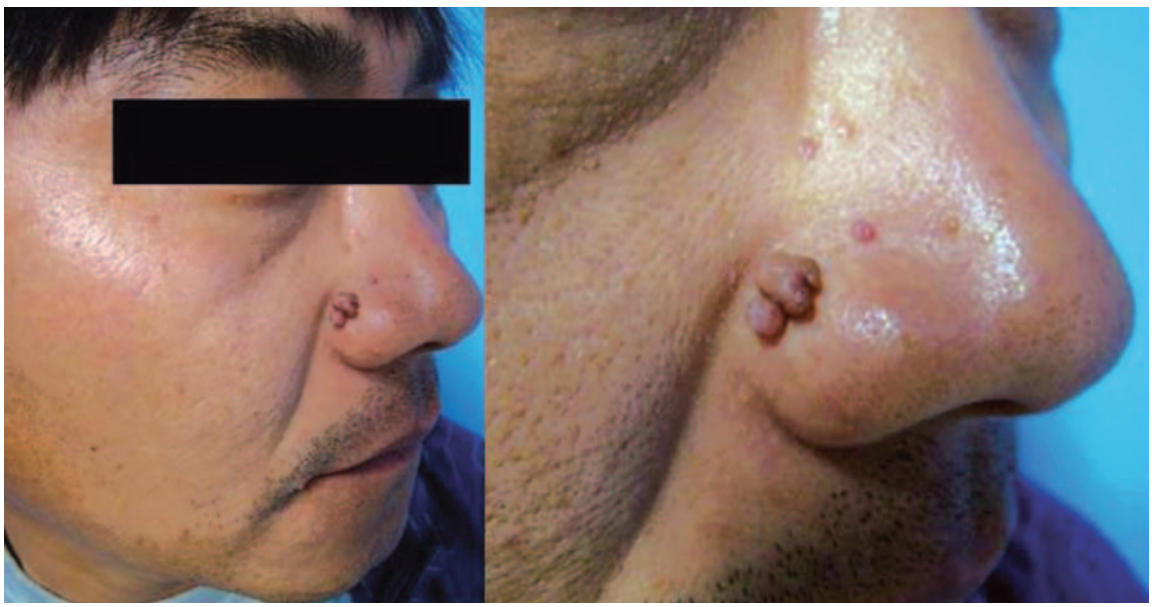

Figure 1. 0.8 cm x 0.8 cm sized flesh-coloured pedunculated polypoid nodule on the right ala of the nose.

with anti-factor XIIIa and CD34 due to the presence of reactive dermal dendrocytes suggests the tumour also has dermal dendritic cell origin.[9,10] The negative result for S-100 protein rules out the possibility of a melanocytic or neural origin.[1] Our immunohistochemical findings which show vimentin+/actin+/CD34+/S-100- were consistent with previous reports. Recently, some studies suggest that there is a link between pleomorphic fibroma and sclerotic fibroma. Martin-Lopez described the term 'pleomorphic sclerotic fibroma', proposing that pleomorphic fibroma, sclerotic fibroma, and pleomorphic sclerotic fibroma form a spectrum.[11]

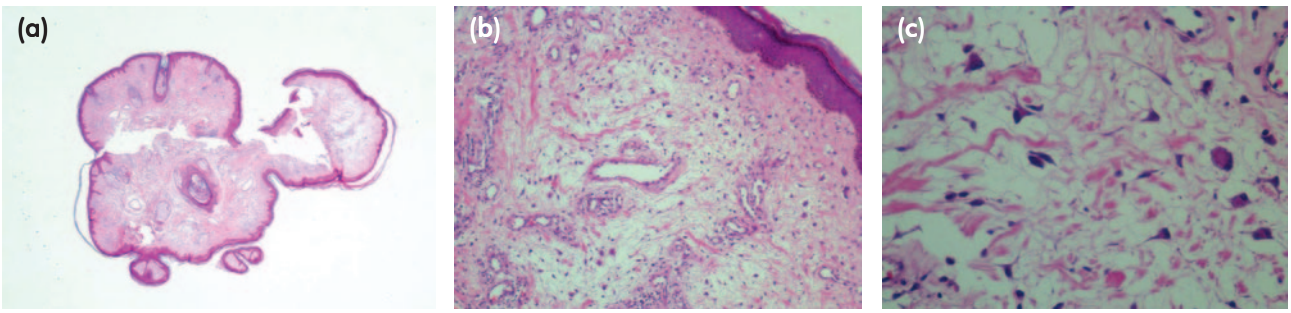

Figure 2. (a) A well-circumscribed polypoid tumour demonstrating an intact epidermis and haphazardly arranged thick collagen bundles mixed with partially loose stroma (H&E x10). (b) Atypical spindle-shaped cells and giant multinucleated cells embedded in haphazardly arranged collagen bundles in the dermis (H&E x40). (c) Atypical spindle cells showing pleomorphic, hyperchromatic nuclei without mitotic figures (H&E x100).

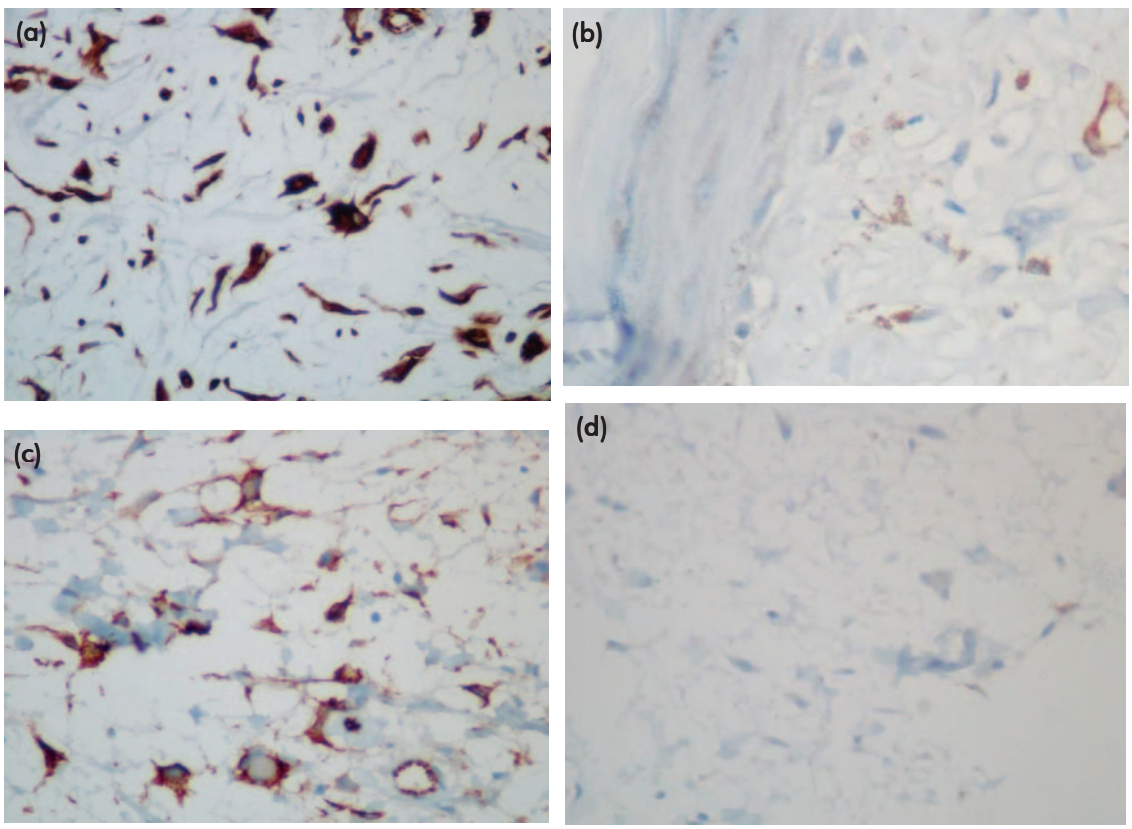

Figure 3. In immunohistochemical staining, atypical tumour cells staining positive for vimentin (a: x400), smooth muscle actin (b: x400), CD34 (c: x400) but negative for S-100 protein (d: x400).

In our case, the main differential diagnoses include soft fibroma, intradermal naevus, neurofibroma and skin appendageal tumours. Pleomorphic fibroma differs from these tumours by its atypical cellularity, like pleomorphic, hyperchromatic nucleus with rare mitoses. Histopathologically, when a tumour shows prominent cellular atypia, pleomorphic fibroma should be differentiated from dermatofibroma with monster cells (DFMC) and atypical fibroxanthoma (AFX). DFMCs are more cellular and have areas with typical histological findings of dermatofibroma. In contrast to that, pleomorphic fibroma expresses CD34 positivity.[10] AFXs present as rapidly growing, highly cellular lesions composed of many types of cells including pleomorphic, spindle-shaped cells, with numerous mitotic figures which may be atypical.[10] Other entities to be considered are benign nerve sheath tumours with atypical features, including neurofibromas with atypia and schwannomas with ancient change.[2] Benign nerve sheath tumours are positive for S-100 protein. These atypical cellular changes are believed to be degenerative and probably have occurred secondarily to ischaemia.[2]

Despite its nuclear atypia, pleomorphic fibroma is considered a benign tumour based on the absence or relative scarcity of mitotic figures. There has been one report of local recurrence of a pleomorphic fibroma, recurring after shave biopsy and electrodesiccation.[1] No reports of metastatic spread were found. However, there has been one report of myxofibrosarcoma arising from myxoid pleomorphic fibroma.[12] It is unclear whether it was a true malignant transformation or incidental change. Thus, studies of the prognosis of pleomorphic fibroma are needed. In our case, follow-up examination done six months after shave biopsy showed no evidence of tumour. Although local recurrence of pleomorphic fibroma is uncommon, the tumour recurred three times after removal by the patient. Further follow-up for the development of new lesions is needed.

In conclusion, we report a case of pleomorphic fibroma on the nose, which is a rare benign fibroblastic tumor occurring on unusual sites. Although pleomorphic fibroma follows a benign clinical course, there has been one case of malignant transformation which developed to myxofibrosarcoma. So, it is important to be aware of pleomorphic fibroma's histological features and immunohistochemical findings to avoid potential misdiagnosis as a benign tumour like soft fibroma or skin appendageal tumour. Also, pleomorphic fibroma needs to be differentiated from tumours that show atypical cellularity like DFMC and AFX which require complete surgical excision. Careful evaluation of histological findings in combination with clinical presentation will allow accurate diagnosis.

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