

### Otophyma is rosaceous lymphoedema (elephantiasis)

Dear Sirs

The report of otophyma presented by Daniels and Haddow<sup>1</sup> appears to represent a case of lymphostatic elephantiasis due to rosacea. Although rarely reported in recent English literature, as pointed out by the authors, otophyma is not such an uncommon phenomenon as case reports do exist in the older literature. Indeed, a recent review of the literature demonstrated that rosacea is one of many causes of ear enlargement.<sup>2</sup> In fact, there appears to be two pathways to localised ear enlargement: (1) lymphoedema due to chronic inflammatory dermatoses (rosacea, psoriasis and 'eczema'), infection (erysipelas and pediculosis), trauma (frost bite) or primary (congenital) lymphoedema; and (2) vascular malformations with tissue hypertrophy.

Lymphoedema appears to be the underlying mechanism of otophyma in many cases. Histologically, lymphoedema is a fibro-inflammatory process characterised by dermal oedema, progressive fibrosis, lymphocytic and plasma cell inflammatory infiltrates, stasis (in the form of increased numbers of thick-walled blood vessels and dilated lymphatic vessels), epidermal acanthosis, and, in some cases, papillomatosis and hyperkeratosis.<sup>3</sup> Rather than being non-contributory to the patient's assessment, we believe that the four initial excision biopsy specimens of 'fibroepithelial polyps' described by Daniels and Haddow<sup>1</sup> were diagnostic of the pathology in question, to wit, lymphoedema presenting as 'lymphoedematous fibroepithelial polyps'. The fifth biopsy's description (scarred superficial dermis, mild telangiectasis and dermal mucin deposition) also implicates lymphoedema.

Once lymphoedema has been identified as the underlying mechanism of ear enlargement; the next question to address is the aetiology of the otophyma. Based on the description of lymphocytic infiltrates surrounding prominent folliculosebaceous units,<sup>1</sup> rosacea may well be the underlying cause, although the history of hand dermatitis raises the possibility of psoriasis, which can also affect the ears. Another factor to consider is infectious otitis externa, which could cause as well as exacerbate lymphoedema.

In conclusion, rosacea is one of many causes of elephantiasis of the ear (otophyma). Re-evaluation of the biopsies in the case report in question should demonstrate the pathological findings of lymphostatic elephantiasis, i.e.: increased number of fibrocytes associated with delicate bundles of collagen oriented parallel to the skin surface throughout at least the upper half of the dermis; dilated lymphatic and vascular spaces in the upper dermis; thick-walled venules oriented vertical to the skin surface; lymphocytic and plasma cell infiltrates; and 'knobby' protuberances covered by hyperkeratotic, acanthotic epidermis. Such findings indicate a case of chronic, inflammation-induced lymphoedema.

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### References

- 1 Daniels K, Haddow K. Otophyma: a case report. *J Laryngol Otol* 2007, 1–3 [Epub ahead of print]
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- 3 Ackerman AB, Bennin B, Chongchitnant N, Guo Y, Reichel M, Sanchez JL *et al.* Lymphedema. In *Histologic Diagnosis of Inflammatory Skin Diseases: an Algorithmic Method Based on Pattern Analysis*, 2nd edn. Baltimore: Williams & Wilkins, 1997: 826–8

### Author's reply

Dear Sirs

We thank Tran, Mazza and Carlson for their interest in our article.

Tran, Mazza and Carlson state that lymphoedema may be the end result of various processes, including rosacea. Although we did not state that lymphoedema was the pathological process involved in our reported case, we do agree that the findings could be consistent with lymphoedema; however, they were non-specific.<sup>1</sup> Our case report did not dispute this fact. However, the term 'fibroepithelial polyp' only describes the pathological findings, which were relatively non-specific and did not identify the aetiology (which had not yet been identified).

As Tran, Mazza and Carlson state, various processes can result in the development of lymphoedema; however, there is no recommendation on how to differentiate these at a histopathological level. This is where clinical history is important. Our patient, as mentioned, did have a previous history of hand dermatitis; however, there was no suggestion of psoriasis. In addition to this, he had a history of otitis externa.<sup>1</sup> Although it was impossible to diagnose with 100 per cent certainty the exact aetiology of our patient's otophyma, it was thought to be a consequence of rosacea rather than otitis externa. The reasoning for this was that the position of the otophyma, obstructing the external auditory canal, had led to the development of otitis externa, rather than the reverse.

There is no mention of the significance of the underlying aetiology and the implications this may have for treatment. However, as with the majority of conditions, ideally, the cause should be treated when possible.

Unfortunately, we are not currently in a position to re-evaluate the pathology samples. However, should this opportunity arise, we would aim to identify some of the other histopathological changes described, in order to try to confirm lymphostatic elephantiasis.

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#### Reference

- 1 Daniels K, Haddow K. Otophyma: a case report. *J Laryngol Otol* 2007, 1–3 [Epub ahead of print]