A Case of Cutaneous Dermal and Follicular Mucinosis

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Background

Cutaneous dermal mucinoses are a complex and diverse group of connective tissue disorders characterized by accumulation of mucin in the skin and/or within the hair follicle. They can be divided into idiopathic or primary form, and a secondary form which can be associated with benign or malignant processes. The primary form usually presents in children and young adults and may resolve spontaneously. However, the secondary form affects older age group and can be associated with mycosis fungoides. The clinical presentation can be varied and may be difficult to diagnose without histological examination.
Case Report

We describe a 66-year-old Malay female with a 3-month history of a growth behind her right ear. The lesion was gradually increasing in size and slightly itchy. There was no associated symptoms such as pain, bleeding or discharge. No preceding trauma or injury was noted. She was otherwise systemically well.

Physical examination revealed a solitary, well demarcated 3.5cm x 2.5cm polypoid jelly-like raised plaque at the retroauricular region and adjacent to the hairline. There was no associated alopecia or other similar lesion elsewhere. Initial impression was a lymphangioma.
Figure 1. Clinical photograph showing a well demarcated, rather boggy, polypoid plaque at right retroauricular region adjacent to hairline.
Figure 2, 3. A 4-mm punch biopsy specimen of skin. At low power microscopy, histology shows the epidermis is flat with loss of rete ridge pattern. Within the dermis, there are large deposits of bluish stringy material which are positive on Alcian blue stain, which occupy the superficial dermis to the deep reticular dermis.
Figure 4. Excision specimen of skin. The dermis shows massive mucin deposition which are also seen within the follicular epithelium and sebaceous gland. The mucin deposits are further highlighted on Alcian blue stain. (x4)
**Figure 5.** Excision specimen of skin. A sparse perivascular and perifollicular infiltrate of lymphocytes and histiocytes is present. (x10)
Progress

A diagnosis of cutaneous dermal mucinosis with follicular mucinosis was made.

Our patient underwent complete excision by carbon dioxide laser with good cosmetic outcome. Full physical examination was unremarkable. Full blood count and peripheral blood film were normal. However, she declined further investigation to rule out any underlying systemic disease and malignancy. She remains well a year later with no recurrence of the lesion.
Conclusion

This case highlights the interesting presentation of the rare cutaneous dermal and follicular mucinosis. Although solitary and localised follicular mucinosis may be idiopathic, it is advisable to follow up such patient in view of its association with underlying malignancy especially mycosis fungoides.
References


