A 25 year old Sudanese male presented with acquired asymptomatic single unilateral lesion which started as a small pigmentation on the left side of the scalp by time increased in color, thickness, losing the hair on top of it and extended to the area behind the left ear. The history was of three years duration, patient gave no history of local trauma. The patient is healthy other wise and there is neither history of any systemic illnesses nor for medication.

On Examination there was defined hyper pigmented plaque with mammillated verrucous surface located on the left occipital area measuring 5x6 Cm in diameter and extending to the retro auricular area (where the skin is normally thin with minimal adnexa) with less thickness and less verrucous surface measuring 2X2 cm in diameters. There was no regional lymph nodes enlargement.

What is your diagnosis?

A verrucous lesion 7x10 cms located in the left retroauricular area and extending to occipital region of the scalp.

Three mm skin biopsy was taken from the lesion on the occipital area for H&E and PAS which revealed pseudoepitheliomatous hyperplasia (hyperkeratosis and papillomatosis) of the epidermis.

The dermis showed arborization of the blood vessels with swollen endothelial cells and lymphoid follicle like lymphocytic infiltration around the newly formed blood vessels and eosinophilic infiltration which became in a lesser degree away from the follicles. PAS stained the basement membrane of the newly formed blood vessels.

Immunophenotyping showed CD45RO+ve, CD3+ve, CD4+ ve, CD5 + ve, CD7+ ve, CD8- ve, CD15- ve, CD19- ve, CD20- ve.

The biopsy confirmed the clinical impression of subcutaneous angiolymphoid hyperplasia. Both clinical and histopathological changes were correlated.

H&E section, x10 magnification showing hyperplastic epidermis overlying superficial and deep patchy perivascular infiltrate.

Dilated blood vessels & eosinophils
The diagnosis is:
Angiolymphoid hyperplasia with eosinophilia

Discussion:
We are reporting a rare case of angiolymphoid hyperplasia with acquired asymptomatic unilateral slowly progressing skin lesion in a young healthy Sudanese male, even this disease is more in middle-aged to elderly white women according to the reported cases. The lesion in our patient is affecting the head, retro-auricular area and the scalp which is similar to the site of predilection in other reported cases. Our patient gave neither history of local trauma nor drug intake while there have been several proposed theories related to the cause of the disease including environmental factors, such as insect bite or parasites, trauma, hyperestrogen states and immunological mechanisms. Some authors have also suggested that it may be secondary to an inflammatory vascular reaction, a neoplastic process or a neo-vascular formation from pre-existing blood vessels. Our patient had no regional lymph nodes enlargement, although regional lymphadenopathy has been reported in approximately 15% of cases and no report of metastasis are known.

All patient’s lab reports were within normal including blood eosinophils which is reported to be increased in the blood and skin lesion and the eosinophilia may be reflecting one of the theories of the cause of the disease which is insect bite or parasites. Our skin biopsy showed the classical follicle-like lymphocytic infiltration which was of T helpers according to the Immunophenotyping. Newly formed blood vessels lined with swollen endothelial cells were obviously seen in our patient’s biopsy stained by H&E and PAS stain. Scanty eosinophilic infiltrate was seen. Although the disease is benign yet its treatment is difficult. In our patient the surgical excision was not recommended because of the site and the size of the lesion. So it was decided to treat him with interferon which is a naturally occurring cytokines that has antiviral, antibacterial, anticancer, anti-inflammatory, immune modulatory effects and in vitro studies demonstrated a direct antiproliferative effect on human dermal microvascular endothelial cells. Intra-lesional interferon 2a was started weekly and after three months the size of the lesion decreased. And when biopsied at that stage the biopsy showed marked fibrose and less lymphocytic infiltration. Cryotherapy was followed in order to achieve further improvement of the thicknees of the lesion.
References: