Read Item - Angioscarcoma of the Scalp

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Date: 29/05/2000
Publisher/Journal:
Keywords: Angiosarcoma loss

Abstract: Doctors resource on Angioscarcoma of the Scalp

Angiosarcoma Of The Scalp

Introduction

Idiopathic cutaneous angiosarcoma of the head and neck is a distinct highly malignant neoplasm of blood vessel endothelium that behaves aggressively and often metastasises to regional lymph nodes, and to lung. Local recurrence of the disease is common as the tumour margins are difficult to define. The prognosis of recurrent disease is dismal.

Clinical Features and Investigation

Angiosarcomas commonly present as single or grouped bluish, red nodules on the face and scalp. There may be some thinning of the hair over the tumour nodules, but alopecia is not marked. Less well differentiated tumours present as diffuse indurated plaques or flat infiltrating macules over which there is a cicatricial alopecia. This form is known as the malignant bruise. Nodules may develop from within the areas of macular erythema and the surface may be verrucous.

The tumours are progressive and extend to involve large areas of the face neck and scalp. There may be gross oedema of the eyelids and large ulcers may bleed profusely. Involvement of the skull produces erosion of the bone and seeds distant metastases. The average survival from diagnosis is under 2 years.

Histology shows a poorly circumscribed dermal tumour that infiltrates subcutaneous fat. Angiomatous and solid patterns occur within the tumour. In the angiomatous regions a meshwork of anastomosing dilated vessels extends between the collagen bundles. The channels are lined by atypical swollen endothelial cells that are plump and resemble hob nails. The solid areas contain poorly differentiated spindle and epithelioid cells. The histological picture may closely resemble Kaposi's sarcoma.

Immunohistochemical stains may be required to distinguish a poorly differentiated angiosarcoma from a spindle cell SCC or amelanotic melanoma.

Management

The prognosis of this condition is poor and the 5 year survival has been estimated at 15%. Surgical excision with a wide margin (2 to 3 cm) of normal skin is required. Even so local recurrence is common. It has been recommended that once the lesion is excised, closure or grafting should be delayed 24 to 48 hours until a rapid paraffin section can be histologically examined to ensure the margins are clear. A Moh's type examination of the margins is best. Post-operative radiotherapy should be given as it has been shown to increase the duration of disease free survival. Large lesions can be treated with interferon-alpha 2b in an attempt to shrink them preoperatively. Massive doses may be required.

Key Points

A highly aggressive malignancy of endothelial cells presenting as a single or multifocal, bluish or violaceous nodule, plaque or macule that may ulcerate and bleed. Sometimes the changes are subtle and the appearance may resemble a bruise.