Fatal Epithelioid Sarcoma

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CASE SUMMARY

History

A 33 year-old lady had a history of mass over the front of her left ankle region in 1988 which was excised by surgeon. She was told that she had sarcoma and wide margin excision was done. In June 1993, she noticed a recurrence of mass at the excision site at the left ankle which was subsequently confirmed to be a recurrence of sarcoma. Re-excision with a wide margin followed by post-operative radiotherapy was done in August 1993. Afterwards, she was regularly followed up by the oncologist. In the recent one year, she noticed a non-itchy non-tender small lump at her scalp. She was relatively asymptomatic except the occasional episodes of spontaneous bleeding from the lump. Besides the past history of sarcoma, she enjoyed good past health all along.

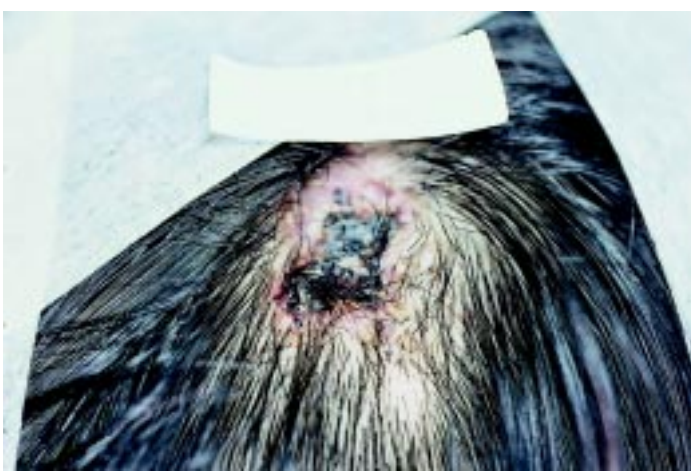

Figure 1: crusted, scabbed erythematous slightly pigmented nodule at vertex

Physical examination

There was a 1cm x 1.5cm erythematous, slightly pigmented nodule at her vertex. The surface of the nodule was crusted and scabbed (Figure 1). General examination revealed no regional lymphadenopathy. There was a long surgical scar at her left ankle, which marked the previous excision of sarcoma.

Investigations and diagnosis

The possible differential diagnoses include metastasis of sarcoma, infective granuloma or other benign skin appendigeal tumours on scalp. Incisional biopsy was done on the nodule which showed infiltrative fascicles of spindle cells that dissected between collagen in the periphery (Figure 2). In areas, there were slit-like spaces with red cells inside suggesting vascular differentiation. The tumour cells were mild to moderately pleomorphic and mitosis was readily found, some being atypical. Immunostaining showed a cytokeratin positive, CD34+ve and CD31-ve profile compatible with that of epithelioid sarcoma. The tumour was unusual in that the entire biopsy showed spindle cell morphology and lacks the characteristic nodular growth with granuloma-like pattern. However, review of the first recurrence in 1993 did show focal epithelioid granuloma-like area with central necrosis characteristic of this tumour (Figure 3).

Other systemic work up showed unremarkable complete blood picture, liver & renal function tests. Chest X-ray showed mild hazziness in the left lower zone.

Management and progress

The patient was urgently referred to surgeon and oncologist for wide margin excision and post-operative radiotherapy. However, the patient ran a rapid downhill course with vital organ metastasis and subsequently died six month later.

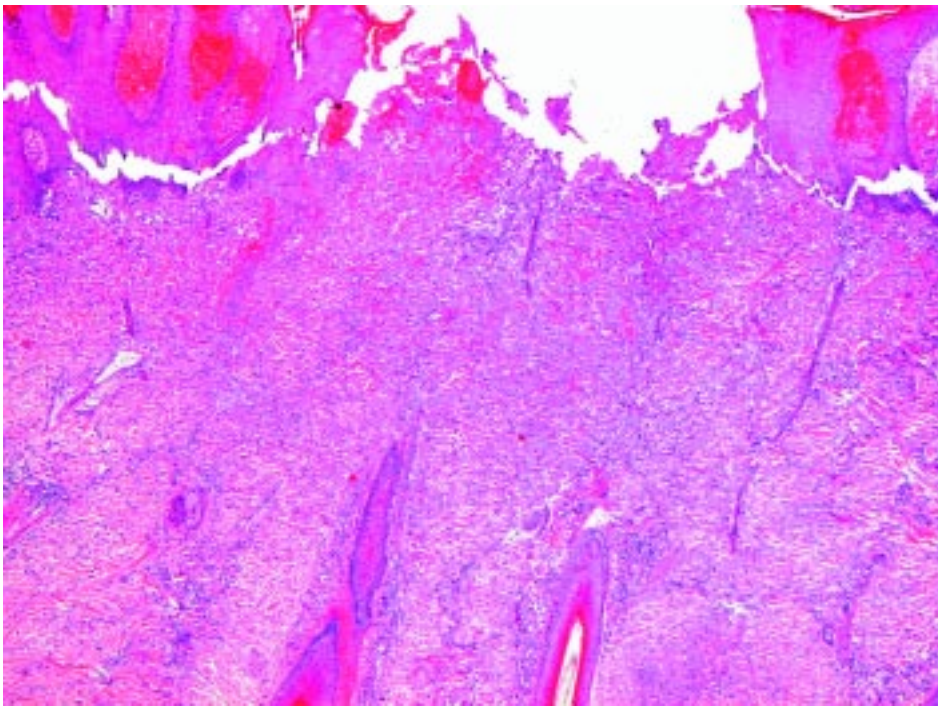

Figure 2: Low power view (15x) showing the diffuse infiltrative spindle cells in the dermis separating skin appendages and dissecting between collagen in the periphery (left lower field). (By courtesy of Dr. K. C. Lee, Department of Pathology, QEH)

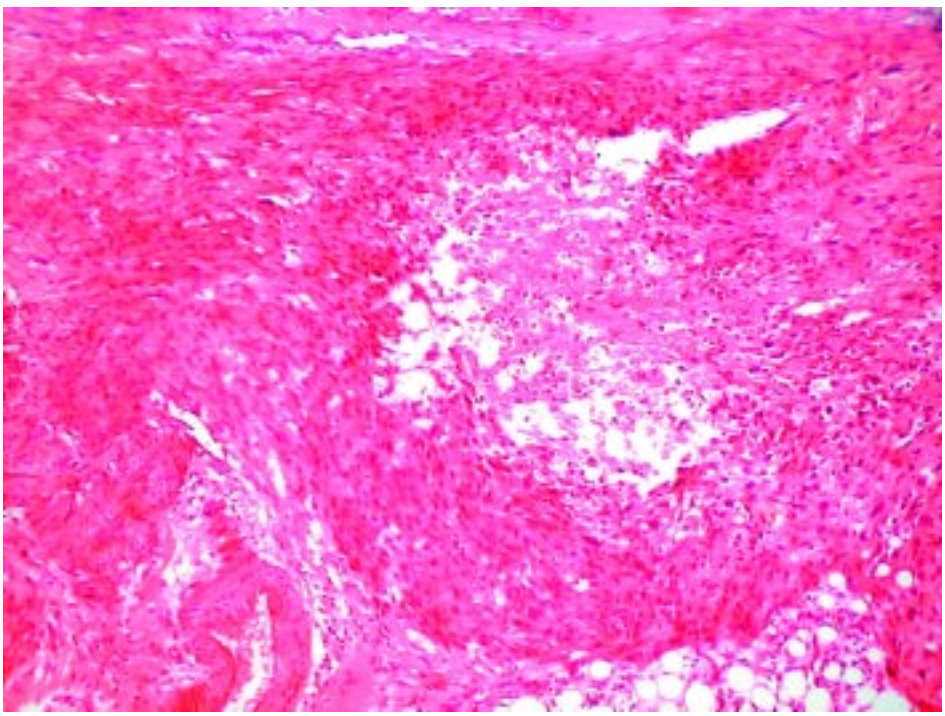

Figure 3: High power view showing the more characteristic epithelioid granuloma-like nodule with central necrosis in the first local recurrence. (By courtesy of Dr. K. C. Lee, Department of Pathology, QEH)

REVIEW ON EPITHELIOID SARCOMA

Epithelioid sarcoma (ES) is an uncommon soft tissue malignancy first reported by Enzinger in 1970.[1]

Clinical feature

ES commonly arises in the distal extremities of young adults as seen in our patient. The male to female ratio is 1.8:1.[2] The most common site is hand and forearm (58% of cases), followed by the lower extremities.[1] Significantly, ES is the most common soft tissue sarcoma of the hand.

The tumour usually presents as a solitary nodule of woody hard consistency in the deep soft tissue, the subcutis or the dermis. Twelve percent have epidermal involvement with ulceration. [1] It is commonly associated with tendon sheaths, joint capsules or neurovascular bundles. Locally recurrent disease tends to involve multiple sites. Pain is usually absent or mild, unless there is peripheral nerve involvement. Radiographic examination is non-specific, revealing a soft tissue mass which may contain fine flecks of calcification. Pressure erosion of the adjacent bone is a common finding.

Histology

The characteristic histological appearance consists of nodular proliferations of plump polygonal to round epithelioid cells which blend with a population of spindle-shaped cells. The polygonal cells surround areas of necrosis. The nodules may fuse into scalloped palisades and display a geographic growth pattern. These cells show large irregular nuclei with obvious nucleoli and variable number of mitotic figures. They tend to splay between collagen bundles and invade adjacent mesenchymal structures. A peripheral chronic inflammatory infiltrate is present. Seventy five percent of ES stain positively for cytokeratin and 50% positive for epithelial membrane antigen. [1] These markers help to differentiate ES from other sarcoma or melanoma. Characteristically, they are also positive for CD34, a marker originally thought to be specific for vascular endothelial cells. But now CD34 are found to be expressed in a number of soft tissue tumour including epthelioid sarcoma, dermatofibrosarcoma protuberants, solitory fibrotic tumour and some gastrointestinal stromal tumours.

Prognosis

The natural history of ES runs a long protracted course with local recurrences and possible metastasis that may occur years after initial presentation. Seventy seven percent of ES have repeated local recurrence. Forty five percent of cases metastasize, most commonly to regional nodes, followed by metastases to lung, scalp and skin. [1] ES may metastasize distantly, without first recurring locally or in regional lymph nodes.

The overall survival is around 70% at 5 years and 50% at 10 years.[2] The poor prognostic factors are tumour greater than 3cm; proximal or axial location of tumour; tumour in deep subcutaneous locations; focal necrosis, haemorrhage and vascular invasion present in histology. On the other hand, women and young patients have a more favourable prognosis.

Treatment

Wide surgical excision or amputation is the treatment of choice, in view of the high rate of local recurrence, poor response to chemotherapy, slow and insidious growth, and an unpredictable mode of spread. However, the extent of surgery does not necessarily correlate with survival or local recurrence, as 20% of cases recur even after amputation. These findings suggest that the tumour extends imperceptively beyond the margin of the operative site as it invades along fibrous structure. In advanced disease, both radiation and chemotherapy have been used but remissions were rare and there was no definitive report of cure.

Learning points:

Epithelioid sarcoma usually presents as slow growing solitary subcutaneous nodule on upper extremities of young men. Local recurrence or distant metastasis in 5 to 10 year period is the usual clinical course. The treatment of choice is surgical resection with wide margins.

References

1. Chase DR, Enzinger FM. Epithelioid sarcoma: diagnosis, prognostic indicatiors, and treatment. Am J Surg Pathol 1985; 9:241-63.
2. Bos GD, Pritchard DJ, Reiman HM, et al. Epithilioid sarcoma: an analysis of fifty one cases. J Bone Joint Surg[Am] 1988; 70:862-70.