

原文題目(出處)：	Primary cutaneous osteosarcoma of the scalp: a case report and review of the literature ( J Cutaneous Pathol 2007;34: 61)
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內文：

➤ Introduction：

- ✚ Extraskkeletal osteosarcoma 是一種罕見的惡性腫瘤，在所有osteosarcoma裡大概只占了2~4%左右。從定義上來看，這種tumor並不是從骨頭起源的，但可能影響到periosteum, cortex和medullary canal。它比較常發生在中年至中老年的男性身上，位置在deep soft tissues of the thigh最常見。
- ✚ 大多數的Extraskkeletal osteosarcoma是de novo的，但是有接近10%的case是跟previous radiation or trauma有關的。
- ✚ 只有10%的Extraskkeletal osteosarcoma是superficially located

➤ Case report：

- ✚ age：84歲；sex：男性
- ✚ Solitary，rapidly growing，exophytic，nodular lesion located on the scalp (vertex region).
- ✚ Physical examination：reddish cutaneous nodule, firm in consistency, measured 2 cm in diameter, had been present for 3 months.
- ✚ Medical history was otherwise unremarkable.
- ✚ CT showed that the lesion extended in soft tissues to the level of the fascia but did not involve the underlyingbone.
- ✚ 因此這個tumor的clinical diagnosis是cutaneous carsinoma
- ✚ Histopathological examination：

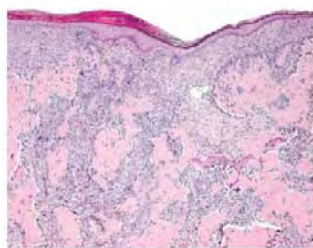


Fig. 1. Scanning magnification shows an atrophic epidermis overlying a dermal proliferation of atypical cells embedded within abundant extracellular matrix.

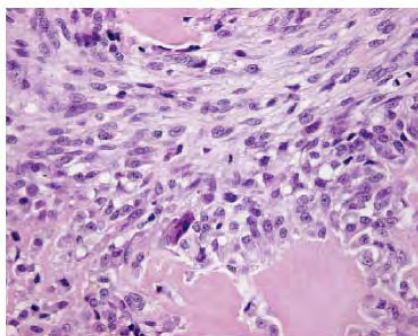


Fig. 2. Neoplastic cells were a mixture of predominantly spindle and epithelioid cells often exhibiting pronounced nuclear atypia and grew in large, cohesive sheets. Scattered osteoclast-like giant cells were distributed within spindle-cell areas.

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## 1. High mitotic activity

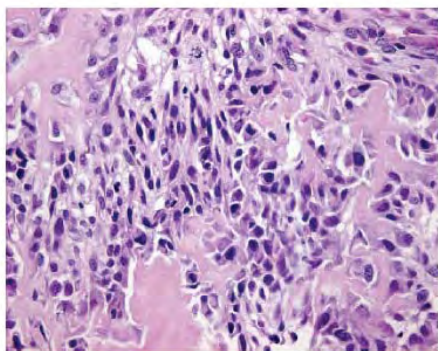


Fig. 3. A high mitotic activity, with numerous atypical mitoses, was noted in the superficial and deep portions of the lesion. Tumor cells merged gradually with areas of less mature osteoid.

2. Numerous atypical mitoses, was noted in the superficial and deep portions of the lesion.

3. The osteoblastic nature of tumor cells:  
 → their close apposition to trabeculae of tumor bone  
 → their entrapment in lacelike osteoid deposits.

The extracellular matrix was recognized as a dense, fibrillar eosinophilic substance deposited between groups of cells. More advanced signs of mineralization producing

clearly recognizable trabeculae of woven tumor bone were also noted the tumor bone trabeculae were haphazardly arranged, had irregular borders, and merged gradually with areas of less mature osteoid. Areas of necrosis were not observed.

✚ Immunohistochemical analysis :

Positivity of tumor cells for cimentin and osteonectin, whereas S-100 protein and cytokeratins were negative.

✚ Clinical examination and extensive total body radiologic workup were performed : no other bone lesions were demonstrated

→ Final diagnosis : primary cutaneous high-grade conventional osteoblastic osteosarcoma.

## ➤ Discussion

在skin可以找到各種形式的Mature bone (osteoma cutis)，但是分化成骨頭的腫瘤在skin是很少的。

Cutaneous ossification分成primary form (osteoma cutis)，也就是並沒有pre-existing lesion或associated lesion；和secondary type (metaplastic ossification)，其ossification可能是由inflammatory (scleroderma, dermatomyostis, chronic venous insufficiency)，traumatic (injection sites, scars)，或是neoplastic的過程 (melanocytic nevi, basal cell carcinomas, pilomatrixomas, chondroid syringomas, dermatofibromas, carcinomas, and desmoplastic melanomas)。

在這些case中，我們可以發現在dermis看到small spicules或是large masses的bone tissue，這些bone tissue常常都是由membranous ossification形成的，所以並沒有cartilage precursor的出現。他們的共同點就是：cellular atypia, along with a high mitotic activity with atypical mitoses, and the disordered architectural pattern strongly suggested a malignant neoplastic process, either primary or metastatic。Osteosarcomatous components是malignant melanoma, metaplastic or biphasic sarcomatoid carcinomas or carcinosarcomas的成分。Malignant epithelial and mesenchymal elements都有osteoblastic differentiation的傾向，並且都是很自發性的表現出來。我們的case中並沒有epithelial component在深層的section中表現出來，因此我們排除了biphasic carcinosarcoma的可能性。基於以上的檢查和檢驗，我們發現這個case在dermis有malignant tumor，並且tumor cell有能力去製造osteoid和成熟的骨頭，因此我們推斷它是個primary或metastatic的osteoblastic osteosarcoma。而且

因為病人的年紀較大，所以cutaneous metastases from osteosarcoma相關的疾病包括了Paget's disease, fibrous dysplasia, chronic osteomyelitis, bone infarct, 和postirradiation。但是因為我們的lesion並沒有其他相關的distant bone或是soft tissue lesion, 因此我們覺得它是Primary cutaneous extraskeletal osteosarcoma。

Table 1, 是以前primary cutaneous extraskeletal osteosarcoma的literature review, 顯示出皮膚作為一個primary site是非常罕見的。

Table 1. Review of reported cases of primary cutaneous extraskeletal osteosarcomas

Author	Cases	Age (years)/sex	Site	Size	Therapy	Follow up	Status	Notes
Drut (1975)	1 case	70/female	Right thigh	-	-	-	-	Arsen in old burn scar
Fletcher (1987)	2 cases	-	Lower leg	-	-	-	-	Arsen in chronic tropical ulcers
Chung and Enzinger (1987)	2 cases	-	-	-	-	-	-	Tumors confined to the subcutis or dermis
Reyes (1989)	1 case	62/male	Temporoparietal region	5 × 3 cm	Surgery	No recurrences	DOC, 7 months	Previous radiotherapy for epibulbar basosquamous carcinoma
Kuo (1992)	1 case	51/female	Popliteal region	3 × 3 cm	Surgery + chemotherapy + radiotherapy	-	-	-
Kobos (1995)	1 case	78/male	Shoulder	1 × 1 cm	Surgery	Local recurrence after 9 months	DOC, 15 months	-
Kircik (1995)	1 case	83/female	Frontal and parietal region	-	Surgery + radiotherapy	Lung metastases	-	Previous trauma
Pillay (2000)	1 case	56/female	Scalp	8 × 8 cm	Surgery + chemotherapy	Metastases after 6 months	-	-
Santos-Juanes (2004)	1 case	96/female	Right temple	4 × 3 cm	Surgery	No recurrences	DOC, 24 months	Arsen under a previously electrodesiccated actinic keratosis
Massi (2006)	1 case	84/male	Scalp	2 × 2 cm	Surgery	No recurrences	Alive, NED, 6 months	Arsen on a previously electrodesiccated scalp due to multiple actini keratoses

DOC, dead of other causes; NED, no evidence of disease.

有一個report認為primary cutaneous extraskeletal osteosarcoma跟burn scars, previous radiation, electrodesiccation of actinic keratoses相關, 這個結果跟我們的case是很雷同的。

而scalp是比較常見到的lesion位置, 所有的病人也都是中老年的成年人。

Prognosis方面, 大部份都是不佳的, 病人會有一些local的recurrences, 而且常伴隨metastatic dissemination。

但是跟更深層的sarcomas來比較, 這些小型、在表淺地區的sarcomas應該是預後較佳的。

題號	題目
1	下面何者是最常見的postirradiation sarcoma? (A) Osteosarcoma (B) Fibrosarcoma (C) Chondrosarcoma (D) Ewing's sarcoma
答案(A)	出處: oral & maxillofacial pathology second edition P.578
題號	題目
2	關於Osteosarcoma, 下列何者為非? (A) Osteosarcoma is the most common type of malignancy to originate within bone (B) The majority of osteosarcomas demonstrate intramedullary origin (C) Osteosarcoma is a malignancy of mesenchymal cells that have the ability to produce osteoid or immature bone (D) Osteosarcomas of the jaws are uncommon
答案(A)	出處: oral & maxillofacial pathology second edition P.574