

Primary cutaneous osteosarcoma of the scalp: a case report and review of the literature

Abstract: We report on an 84-year-old man with a solitary, nodular lesion on the scalp. The patient had been previously submitted to electrodesiccations of the scalp due to multiple solar keratoses. Histopathologically, the lesion showed features of a high-grade conventional osteoblastic osteosarcoma involving the dermis. Computed tomography showed no involvement of the underlying bone tissues. Clinical examination and extensive total body radiologic workup revealed absence of bone lesions in any body site, thus suggesting a final diagnosis of primary cutaneous extraskeletal osteosarcoma. The clinico-pathological features of the case are discussed in light of the rare cases previously described in the literature.

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Extraskeletal osteosarcoma is a rare malignant neoplasm accounting for approximately 2–4% of all osteosarcomas.¹ By definition, the tumor does not arise from bone but may secondarily involve the periosteum, cortex, or medullary canal. It typically arises in male patients during mid and late adulthood in the deep soft tissues of the thigh.^{1–3} Other less frequent sites include the buttock, shoulder, trunk, and retroperitoneum.¹ The majority of extraskeletal osteosarcomas develop *de novo*, but up to 10% of cases have been associated with previous radiation or trauma.² Only 10% of extraskeletal osteosarcoma are superficially located, and the skin may be involved in the setting of widespread metastatic dissemination from a skeletal or extraskeletal osteosarcoma.^{4–9} The occurrence of an extraskeletal osteosarcoma arising primarily in the skin is an exceedingly rare finding.

We herein describe an 84-year-old patient developing a primary cutaneous extraskeletal osteosarcoma on the scalp and discuss its clinico-pathological features in view of the cases previously described in the literature.

Case report

An 84-year-old man presented with a solitary, rapidly growing, exophytic, nodular lesion located

on the scalp (vertex region). Physical examination revealed that the reddish cutaneous nodule had firm consistency and measured 2 cm in diameter. The lesion had been present for 3 months. The patient had been previously submitted to electrodesiccations of the scalp for multiple actinic keratoses. Medical history was otherwise unremarkable. Computed tomography showed that the lesion extended in soft tissues to the level of the fascia but did not involve the underlying bone. The tumor was excised with a clinical diagnosis of cutaneous carcinoma.

Histopathological examination showed an atrophic epidermis overlying a dermal proliferation of atypical cells embedded within abundant extracellular matrix (Fig. 1). 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 (Fig. 2). High mitotic activity, with numerous atypical mitoses, was noted in the superficial and deep portions of the lesion (Fig. 3). The osteoblastic nature of tumor cells was recognized by their close apposition to trabeculae of tumor bone or by their entrapment in lacelike osteoid deposits. The extracellular matrix was recognized as a dense, fibrillar eosinophilic substance deposited between groups of

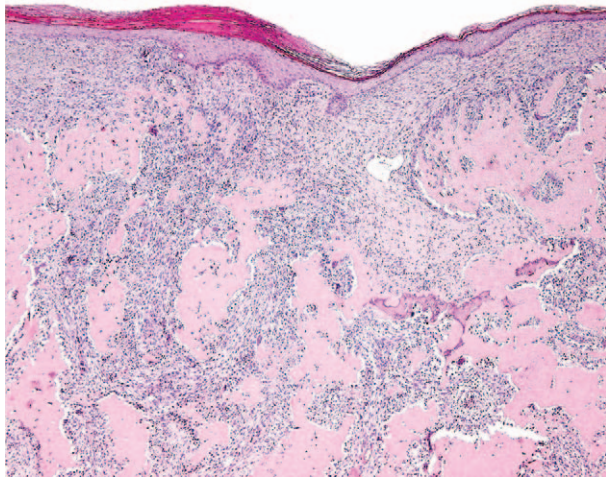


Fig. 1. Scanning magnification shows an atrophic epidermis overlying a dermal proliferation of atypical cells embedded within abundant extracellular matrix.

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 showed positivity of tumor cells for vimentin and osteonectin, whereas S-100 protein and cytokeratins were negative. Upon histopathological examination, clinical examination and extensive total body radiologic workup were performed. No other bone lesions were demonstrated, thus suggesting a final diagnosis of primary cutaneous high-grade conventional osteoblastic osteosarcoma. The patient did not receive adjuvant therapy and is alive and well with no recurrences after 6 months' follow up.

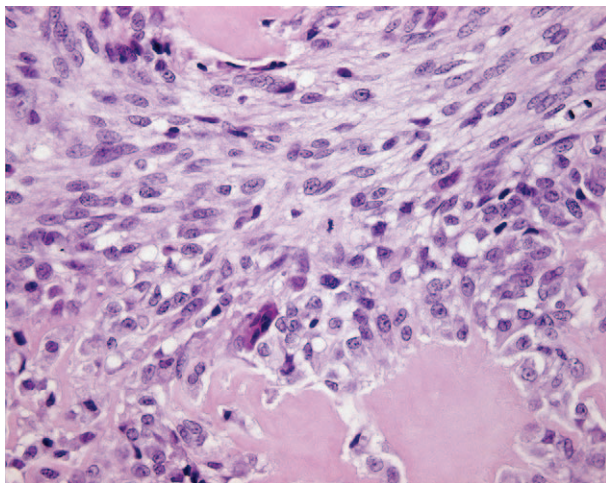


Fig. 2. Neoplastic cells were a mixture of predominantly spindle and epithelioid cells often exhibiting pronounced nuclear atypia and grew in large, cohesive sheets. Scattered osteoclast-like giant cells were distributed within spindle-cell areas.

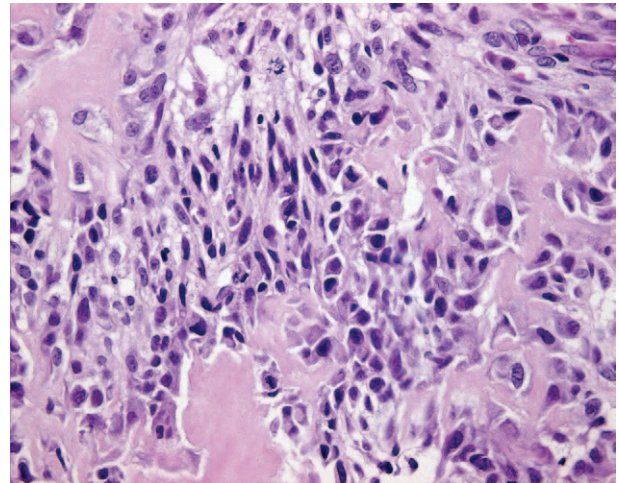


Fig. 3. A high mitotic activity, with numerous atypical mitoses, was noted in the superficial and deep portions of the lesion. Tumor cells merged gradually with areas of less mature osteoid.

Discussion

Mature bone may be found in the skin in the various forms of osteoma cutis, but true neoplasms with differentiation toward bone are exceedingly rare in the skin. Cutaneous ossification may occur as a primary form (osteoma cutis), where there is an absence of a pre-existing or associated lesion, and a secondary type (metaplastic ossification), where the process of ossification develops in association with or secondary to inflammatory (scleroderma, dermatomyositis, and chronic venous insufficiency), traumatic (injection sites, scars), or neoplastic processes (melanocytic nevi, basal cell carcinomas, pilomatrixomas, chondroid syringomas, dermatofibromas, carcinomas, and desmoplastic melanomas). In these cases, small spicules or large masses of bone tissues are seen in the dermis and/or subcutis and usually develop by membranous ossification without the presence of a cartilage precursor. In the current case, 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 have been described in malignant melanoma¹⁰ as well as, relatively more commonly, in the setting of so-called metaplastic or biphasic sarcomatoid carcinomas or carcinosarcomas. In such metaplastic carcinomas, both malignant epithelial and mesenchymal elements, including signs of osteoblastic differentiation, are simultaneously present.¹¹ Although the chance of sampling error always has to be taken into careful consideration, in our case no signs of epithelial component were demonstrated in serial deeper sections, thus ruling out the possible diagnosis of a biphasic carcinosarcoma.

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

Author	Cases	Age (years)/sex	Site	Size	Therapy	Follow up	Status	Notes
Drut (1975)	1 case	70/female	Right thigh	–	–	–	–	Arisen in old burn scar
Fletcher (1987)	2 cases	–	Lower leg	–	–	–	–	Arisen 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 epiauricular 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	Arisen under a previously electrodesiccated actinic keratosis
Massi (2006)	1 case	84/male	Scalp	2 × 2 cm	Surgery	No recurrences	Alive, NED, 6 months	Arisen on a previously electrodesiccated scalp due to multiple actini keratoses

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

In the current case, the overall cyto-architectural picture was in line with a malignant tumor involving the dermis and composed of tumor cell having the ability to produce osteoid and mature bone; thus, the hypothesis of an osteoblastic osteosarcoma, either primary or metastatic, was considered. Given the patient's age, the possibility of cutaneous metastases from osteosarcoma arising in association with pre-existing bone diseases, such as Paget's disease, fibrous dysplasia, chronic osteomyelitis, bone infarct, or postirradiation, was intriguing. Indeed, skin metastases from skeletal and extraskelatal osteosarcomas have been seldom reported. However, in the context of the clinical setting (absence of underlying or distant bone or soft tissue lesions), we interpreted the lesion as a primary cutaneous extraskelatal osteosarcoma.

Review of the literature showed that skin as a primary site is a rare event (Table 1).¹²⁻¹⁹ There are some hints to extraskelatal osteosarcomas confined to the dermis and subcutaneous tissue in two of 88 cases reported by Chung and Enzinger,² in two cases reported by Fletcher¹³ in association with chronic tropical ulcers, but the first case of primary cutaneous osteosarcomas with complete clinicopathological data was described by Kuo in 1992.¹⁵ Interestingly, there is one report suggesting a possible association with burn scars,¹² previous radiation,¹⁴ and electrodesiccation of actinic keratoses,¹⁹ as in our case. However, the causal relationship between these conditions and the development of the tumor has still to be demonstrated with certainty. The scalp seems to be a common site for occurrence, and all patients are in their early or late adulthood. The prognosis of soft tissue extraskelatal osteosarcoma is generally poor, with patients experiencing local recurrences often followed by metastatic dissemination. Concerning cutaneous osteosarcoma, the number of cases described so far is too low to draw definitive conclusions, although it is conceivable that the small, more superficial sarcomas may behave better than their deeper counterparts.

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