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CORRESPONDENCE

Primary cutaneous osteosarcoma: a lesion arising *de novo* in the posterior triangle of the neck

Sir,

Osteosarcoma is characterised by the formation of malignant osteoid.[1] It most commonly occurs in children and young adults in their second decade, with a slightly higher incidence in males.[3] Osteosarcoma typically involves the long bone metaphysis, typically around the knee.[2] Risk factors for the occurrence of osteosarcoma include previous radiation therapy, Paget's disease of bone, and germ line abnormalities such as the Li–Fraumeni syndrome, Werner syndrome, Rothmund–Thomson syndrome, and familial retinoblastoma.[2]

Extraskeletal osteosarcomas are osteoid producing mesenchymal derived malignancies that arise in soft tissue, distinct from the bone or periosteum.[3] Skeletal and extraskeletal osteosarcomas commonly metastasise to the lung, liver, brain and bone, and metastasis to the skin is extremely rare.[4]

Primary extraskeletal osteosarcoma was first described by Wilson in 1941 and represents 1–2% of all soft tissue sarcomas.[1] Extraskeletal osteosarcoma most commonly arises in the muscle and subcutaneous tissues of the limbs and, in contrast to skeletal osteosarcoma, it tends to affect patients in their fifth and sixth decades.[3] The histological appearance is very similar to a primary osseous lesion, characterised by variable proportions of irregularly distributed malignant osteoid, chondroid elements and a spindle cell stroma.[5] Cellular pleomorphism, a high mitotic rate and the presence of multinucleated giant cells are typical.[2] Primary cutaneous osteosarcomatous lesions are a rare entity. Patients are most commonly in their fifth or sixth decades and lesions are often located on the lower limb.[6]

Primary cutaneous osteosarcomas of the head and neck are extremely rare and to our knowledge all have previously occurred at the site of previous radiotherapy or trauma.[7] Two of these tumours arose in areas where an actinic keratosis had previously been electrodessicated, one following traumatic injury and two following radiotherapy.[7,8] There have been a small number of cases of extraskeletal osteosarcoma arising *de novo* in the subcutaneous tissues of the head and neck.[9] However, of these only one displayed any gross or histological evidence of epidermal involvement and this was in association with a nearby nest of basaloid cells with peripheral palisading and clefting artefact, typical of basal cell carcinoma (Table 1).

Here we describe the case of a 70-year-old farmer presenting with a 3 month history of a rapidly growing, pedunculated lesion in the right posterior triangle, overlying nodal zone V, of his neck (Fig. 1). He recalled no history of a predisposing lesion or trauma at this site but described intermittent bleeding and crusting. The patient had a past medical history of gastro-oesophageal reflux disease (GORD) and last attended hospital due to a left slipped upper femoral epiphysis, aged 16 years. Otherwise he was fit and well. He was a non-smoker and lived an active lifestyle despite residual limp secondary to his adolescent hip disorder.

Examination revealed a 2.5 cm diameter, friable, brown lesion, with ulcerated areas and a collar of white nodular material. The lesion was overlying the posterior triangle and the middle third of the sternocleidomastoid muscle. There was no associated lymphadenopathy and no clinical indication of nerve or vessel infiltration. The working clinical diagnosis was squamous cell carcinoma or amelanocytic melanoma and surgery was planned.

The lesion was excised and at operation this was found to be adherent to underlying fascia which resulted in the greater auricular nerve being sacrificed. Urgent pathology was requested. Macroscopic examination described a friable brown multinodular lesion measuring 2.5 × 2.5 × 2 cm ulcerating the skin surface and infiltrating into the deep dermis. Histological examination revealed a cellular mitotically active epithelioid and spindled malignant tumour with zones of coagulative tumour necrosis. It was associated with osteoid matrix production. A panel of immunohistochemical markers including keratins, vascular and melanoma markers, desmin and smooth muscle actin were negative. Although at this site the morphological differential would include pleomorphic undifferentiated sarcoma with multinucleate giant cells, which can form peripheral shells of immature matrix, giant cells were not a conspicuous feature of this lesion and, furthermore, this tumour was associated with filigree calcifying osteoid throughout (see Fig. 2). There were no morphological or immunophenotypic features to suggest carcinosarcoma.

Following discussion at the national sarcoma multidisciplinary team meeting, a bone scan, local and staging computed tomography (CT) scans were arranged. The bone scan highlighted an area of increased uptake in the left hip, which was consistent with osteoarthritic degeneration secondary to slipped upper femoral epiphysis. Increased uptake in the lumbar spine and right knee were consistent with degenerative change. In particular, no abnormal uptake was seen in the cervical spine. No alternative primary sites or metastatic lesions were detected and the patient underwent 2 cm wider local excision, with resection of the underlying sternocleidomastoid muscle and reconstruction using a cervicofacial flap. Histological examination of the wider excision specimen revealed no involvement of the underlying muscle and confirmed complete clearance. The post-operative period was uncomplicated and the patient was discharged 4 days following surgery with radiotherapy commenced once wound healing was achieved. At the time of writing this article, 5 years post-operatively, the patient had no further recurrence of disease at the neck; however, he had developed jaundice and been recently diagnosed with hilar cholangiocarcinoma with liver metastases.

This is the first described case of primary cutaneous osteosarcoma, with epidermal involvement, arising spontaneously in the head and neck. Primary cutaneous osteosarcoma of the head and neck is extremely rare and those few cases previously described have had a history of previous excision of lesion, radiotherapy or injury.[5] As in this case, previous cases have been described in patients beyond the fifth decade. Some reports suggest a slight male preponderance.[3]

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Table 1 Primary extraskeletal osteosarcoma of head and neck – no cutaneous changes/epithelial components

Author	Age/sex	Location	Previous trauma	Macroscopic appearance	Cutaneous involvement	Size	Histological description	IHC	Management	Outcome
Nishimura (1972)	–	–	–	–	No	–	–	–	–	–
Binkhuysen (1986)	–	–	Previous liposarcoma	–	No	–	–	–	–	–
Reyes (1989)	62/M	Tempoparietal region	Previous radiotherapy for BCC	–	–	5×3 cm	–	–	Surgery	–
Kircik (1995)	83/F	Frontal and parietal region	Yes	–	–	–	–	–	Surgery and radiotherapy	Lung metastasis
Lima (1998)	78/M	Frontal region, subcutaneous	–	Subcutaneous tissue involving fascial planes	No	8×6×5 mm	Osteoblastic: malignant osteoid deposited around osteoblasts, pleomorphic cells and malignant giant cells, spindle cells arranged in bundles	Cytokeratins negative	Resection and radiotherapy	Local recurrence at 4 months, death 10 months
Ayad (1999)	55/F	Buccal region	Multiple SCC and radiotherapy	–	No	–	–	–	–	–
Pillay (2000)	56/F	Scalp	No	Exophytic	–	8×8 cm	Osteoblastic/fibroblastic	–	Surgery and chemotherapy	Metastases after 6 months
Santos-Juanes (2004)	96/F	Right temple	Previous electrodessicated actinic keratosis	Cutaneous, exophytic lesion	Yes	4×3 cm	Osteoblastic/telangiectatic: cytoplasmic eosinophilic cells with nuclear pleomorphism, conspicuous chromatin abnormalities, prominent nucleoli, and many mitotic figures. Multinucleate tumour giant cells present. Malignant cells producing osteoid	Vimentin positive. S100, cytokeratins (AE1/AE3), desmin, muscle-specific actin, and EMA all negative. CD34 showed positivity in vessels	Resection	No recurrence or metastasis, but died of pneumonia 24 months after original diagnosis
Hatano (2005)	25/M	Right jaw	No	Subcutaneous tumour with calcified central portion, no intramuscular elements	No	15×13 mm	Rounded or spindle-shaped cells in the stroma with osteoid, some of which were calcified	Vimentin positive. Cytokeratin (AE1/AE3) and EMA negative	Pre-operative chemotherapy followed by wide resection	Treated with chemotherapy, wide resection, disease free (16 months post-op at time of writing)
Massi (2006)	84/M	Scalp (vertex region)	Previous electrodessicated scalp due to multiple actini keratoses	Cutaneous, exophytic lesion	Yes	2×2 cm	Osteoblastic: spindle and epitheloid cells exhibiting pronounced nuclear atypia. Osteoclast-like giant cells distributed within spindle-cell areas associated with osteoid production	Vimentin and osteonectin positive. S100 and cytokeratins negative	Resection	No recurrences after 6 months follow-up
Saito (2008)	18/M	Left submandibular region	No	Subcutaneous/ intramuscular, calcified lesion	No	23×18 mm	Chondroblastic: malignant spindle cell tumour with osteoid production	Vimentin and S100 positive	Resection, selective neck dissection and radiotherapy	Massive local recurrence 3 months post-operatively; 5 months post-operatively death from acute intracranial haemorrhage

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