CASE REPORT

Malignant myoepithelioma of the humerus with a satellite lesion: a case report and literature review

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ABSTRACT. We report an extremely rare case of malignant myoepithelioma presenting as a cortical osseous lesion on the humeral shaft with a satellite lesion. A 21-year-old man presented with persistent pain of the right upper arm after local trauma that had occurred 2 months earlier. Radiological examination revealed an expansile osseous tumour based on the cortex of the humeral shaft as well as a satellite lesion. *En bloc* resection was performed. Microscopic examination with immunohistochemical staining was used to establish a diagnosis of malignant myoepithelioma. Osseous malignant myoepithelioma occurring in long tubular bones rather than in bony structures with salivary tissue is extremely rare. Here, we demonstrate radiological and pathological features of a malignant myoepithelioma that developed in the cortex of the humeral shaft and review previously reported cases.

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Myoepithelioma is a tumour composed of spindle, plasmacytoid, epithelioid and clear cells exhibiting myoepithelial differentiation, with island-like arrangements of sheets and cords [1]. These tumours frequently have abundant stroma, including acellular, mucoid or densely hyalinised myxochondroid stroma without ductal differentiation. Both benign and malignant forms of myoepithelioma can exist, although the malignant form is very rare compared with the benign form.

Myoepithelioma occurs most frequently in the parotid gland [2], although it can also occur in hard and soft palate areas containing salivary tissues [2–5]. Other than salivary glands, myoepithelioma has also been reported in the soft tissue [6–8], ear [9], sinonasal cavity [10], breast [11] and lung [12]. Bony involvement is extremely rare [13–15], and most of the cases occurred in the head and neck region.

This is the second case report of osseous malignant myoepithelioma of a tubular bone outside the head or neck region. In this study, we report the case of a 21-year-old man with right upper arm pain that was later revealed to be malignant myoepithelioma. We discuss the clinico-pathological and radiological features of the case and review previously reported cases. Our patient gave consent for data concerning the case to be published.

Case report

A 21-year-old man presented with a 2-month history of pain in his right upper arm following local trauma that occurred during military training.

Plain radiographs of the right humerus showed two eccentric osteolytic lesions with similar appearances

(Figure 1). The lesions were well defined, expansile, lobular, lytic and intracortical with medullary extensions through the inner cortical disruption. Shell-like periosteal reaction was noted, along with focal breakdown of the outer cortex in the larger lesion. Mild cortical thickening adjacent to the lesion was observed at the transition zones. There were no soft-tissue masses, and matrix calcification was not present. The proximal lesion was much larger than the distal lesion, and there was no direct continuity between the two lesions. MRI demonstrated that the lobular lesions were centred on the cortex and extended into the intramedullary portion (Figure 2). The lesions were isointense or hypointense on T_1 weighted images, intermediate (hypointense relative to fat) on T_2 weighted images and showed heterogeneous enhancement on Gd-enhanced T_1 weighted images. Additional MRI findings, such as adjacent soft-tissue extension related to focal cortical breakdown and medullary extension with surrounding marrow oedema resulting from irregular disruption of the inner cortical margin, were thought to suggest the locally aggressive nature of their growth patterns rather than a pathological fracture related to the trauma history. A positron emission tomography (PET)-CT scan revealed a mildly hypermetabolic lesion on the right humerus with a maximum standardised uptake value (SUV) of 2.93. No other hypermetabolic lesions reaching clinical significance were observed. Radiological differential diagnoses included osteofibrous dysplasia, adamantinoma, polyostotic fibrous dysplasia, osteoblastoma and metastases, although the last diagnosis was deemed least likely.

Incisional biopsy was performed, and the patient was diagnosed with malignant myoepithelioma. Following this, the patient underwent *en bloc* tumour resection of the humeral shaft with an autologous bone graft.

The humeral lesion was segmentally resected, and the cut surface had two separate lesions. The larger of the

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Figure 1. Anteroposterior radiograph of the right humerus reveals two well-defined, expansile, lobular, osteolytic lesions based on the lateral cortex with reactive sclerosis and focal cortical disruption (arrow).

two lesions exhibited thickened cortical bone and had a grey-to-bluish myxoid lobulated appearance. In addition, focal haemorrhage was present. A second smaller lesion that was separate from the larger lesion also exhibited a myxoid lobulated appearance (Figure 3). Histologically, the lesion was composed of epithelioid cells with round-to-ovoid nuclei with distinct and abundant pinkish cytoplasm. The cells were arranged in sheets in a cord-like fashion with squamous differentiation (Figure 4). Within the area, cells formed lobular arrangements within the myxochondroid matrix, which exhibited a bluish benign chondroid differentiation. Mitoses were frequently observed and definite marrow invasion was identified. Immunohistochemical staining

was positive for cytokeratin, p63, S-100 protein, desmin, vimentin and glial fibrillary acid protein (GFAP).

Discussion

Myoepithelioma is a very rare tumour type, accounting for 1.5% of both major and minor salivary gland tumours [13]. The mean age of myoepithelioma onset is in the fifth decade of life, and there are no specific gender predilections [13]. Malignant myoepithelioma, also known as myoepithelial carcinoma, was first described by Stromeyer et al [16] in 1975. During the past 40 years, this disease has been considered one of the variants of pleomorphic adenoma rather than a unique disease entity [2].

Most cases of myoepithelioma occur in the parotid and mandibular glands, and less frequently in the hard and soft palates. Myoepithelioma in soft tissue is rare, constituting only 14.4% of reported cases [17]. Likewise, the involvement of bony structures is extremely unusual. The first case report of myoepithelioma involving a bony structure in English literature was in the maxillary sinus of a 67-year-old man in 1998. In this case, the source of the myoepithelioma was identified as the accessory salivary glands in the maxillary sinus [18]. Most intraosseous myoepithelial tumours occur in the maxilla. Ferretti et al [13] reported three intraosseous myoepitheliomas arising from the maxillae of a 14-year-old girl, an 18-year-old man and a 14-year-old boy. Further, in 2008, a case of intraosseous myoepithelioma from the maxilla was reported in a 54-year-old woman [14]. Alberghini et al [15] reported a case of primary malignant myoepithelioma involving the distal femur of a 55-year-old man. This was the first such case report of an osseous malignant myoepithelioma at a site other than the head and neck region, such as the maxilla. MRI revealed an intramedullary osteolytic lesion with multifocal cortical disruption and extension into the surrounding soft tissue. It was initially diagnosed as a sarcoma with myxoid stroma at the time of biopsy; however, a final diagnosis of malignant myoepithelioma was made after reviewing the resected specimen. In our review of the literature of osseous myoepitheliomas, all except one case were located in the maxilla, where accessory salivary glands can exist.

Pleomorphic adenoma or salivary gland mixed tumour, which is in a similar spectrum to myoepithelioma, also rarely occurs in bony structures. In 2001, McGough et al [19] reported the case of a mixed tumour arising in the left tibia of a 42-year-old woman; the tumour eventually metastasised to the lung. The patient's past medical records revealed no previous salivary gland lesion. In 1992, a multicentric mixed tumour, initially involving the metacarpal bone, radius, ulna and other metacarpal bones, was reported [20].

Pathologically, it is difficult to differentiate myoepithelioma from pleomorphic adenoma, as most of their histological features are similar. This is especially true for lesions with hyalinised stroma of the chondroid matrix. Therefore, differentiation of myoepithelioma from pleomorphic adenoma is possible only when there is either no or minimal ductal differentiation present. Likewise, it is very important to demonstrate myoepithelial

Figure 2. T_2 weighted (a) and fat-suppressed enhanced T_1 weighted (b) coronal images demonstrate the presence of a locally aggressive tumour with adjacent soft-tissue extension via focal outer cortical disruption (a, white arrow) and medullary extension through the breakdown of the inner cortex (a, black arrows). Heterogeneous enhancement and adjacent marrow oedema are also noted. (c) Axial T_1 weighted image reveals the epicentre of the tumour as the lateral cortex with cortical expansion and extension into both the adjacent soft tissue and medullary cavity.

Figure 3. Cut surface of the humerus depicts two separate lesions. Each lesion shows thickened cortical bones and lobulated greyish myxoid tumour tissue involving the medullary cavity.

Figure 4. Photomicrograph (haematoxylin and eosin, 400 $\times$) demonstrates squamous differentiation with myoepithelial cells.

differentiation immunohistochemically and/or ultra-structurally. In the present case, we were able to reveal myoepithelial differentiation using immunohistochemical staining. Specifically, cytokeratin, p63, GFAP and S-100 protein stained positively in epithelial cells [2].

Differentiation of myoepithelioma from pleomorphic adenoma is important in several aspects because myoepithelioma is considered to be at one end of the disease spectrum of pleomorphic adenoma. Specifically, myoepitheliomas have similar, but not identical, biological behaviours and can also reveal malignant transformation.

Radiologically, the case presented in this study manifested as an expansile osseous lesion based on the cortex of humeral metadiaphysis with local aggressiveness and smaller satellite lesions. These features shared little radiological similarity to previously reported cases, having extensive medullary osteolysis of the femur and adjacent soft-tissue invasion [15]. We considered various cortical lesions in the differential diagnosis of our case. Tumours or tumour-like lesions, which are centred on the cortex of long tubular bones, cause cortical destruction, and may have multiplicity, including osteofibrous dysplasia (so-called ossifying fibroma), adamantinoma, osteoblastoma, polyostotic fibrous dysplasia, metastases and haemangioendothelioma [21]. Radiologically, these lesions may take on various appearances, all of which share some similarities. Therefore, histological examination is frequently required for correct diagnosis. Nevertheless, each type of lesion has a few characteristic radiological features. Osteofibrous dysplasia involves the anterior cortex of the tibia in 90% of cases and is characterised by anterior bowing and an adjacent sclerotic band [21]. Likewise, adamantinoma exhibits very similar radiographic findings, but with a tendency towards local aggressiveness such as medullary extension or satellite lesions in direct continuity with the major lesion. Osteoblastoma produces highly variable radiological features. It is characterised by a high potential for calcification or ossification, and may show local aggressiveness without metastasis. Fibrous dysplasia can be differentiated from other lesions by a ground-glass appearance, additional opacification and deformities such as anterior bowing. Haemangioendothelioma involves multiple sites in 20–50% of all cases, and its appearances are either well-defined or ill-defined lytic lesions with rare reactive sclerosis.

In conclusion, we present the case of a 21-year-old man with malignant myoepithelioma based on the cortex of his humerus. We documented the clinical and characteristic pathological features of the bony myoepithelioma and discussed the radiological features with their differential points. Although cases like this are extremely rare, and this case possessed a few radiological findings incompatible with other cortical tumours of long tubular bones, immunohistochemical analysis clearly demonstrated myoepithelial differentiation and was thus crucial for making the final diagnosis.

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