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Myoepithelioma of the spine: first case in the literature

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Abstract. – Myoepithelioma is a very rare tumour.

This tumor type has been reported in the soft tissue, ear, sinonasal cavity, breast and lung. Although rare, myoepithelioma can occur in bone.

We present the first case of myoepithelioma in the spine, documenting the clinical, radiographic and pathological features.

Key Words:
Myoepithelioma, Spine.

Introduction

Myoepithelioma is a very rare tumour type; mixed tumours, parachordomas and myoepitheliomas are well-defined soft tissue malignancies characterized by cells of myoepithelial linage embedded in a hyalinized to chondromyxoid stroma. Thus, they are currently considered to belong to the same spectrum of differentiation[1].

Mixed tumor is the analogue soft tissue counterpart of the more common salivary-gland tumor[2-4] that accounts for 1.5% of salivary gland tumours[5,6].

Myoepithelioma has been reported in the soft tissue[7-9], ear[10], sinonasal cavity[11], breast[12] and lung[13].

Although rare, myoepithelioma can occur in bone of the extremities or ileum[2,5,6,14-16].

These tumors usually occur in adults with an average age of 35 years with no specific gender predilections[5,6,16]. We are not aware of any previous reports of a mixed tumor arising in the spine.

We present the first case of myoepithelioma in this location, documenting the clinical, radiographic and pathological features. Our patient gave consent for data concerning the case to be published.

Case Report

A 62-year-old man was attended at our Center for 10-month history of dorsal pain and asthenia. No neurological deficit was evidenced in the physical exam. Medical history revealed right lung segmental resection for a lesion described as mesenchimoma in 1992. During the follow-up the patient underwent chest radiographs and computed tomographic scan of the chest that showed a 3 cm pulmonary nodule in the left ilium.

Positron emission tomography (PET-TC) revealed a hypermetabolic lesion in the vertebral body of T11 with a maximum standardised uptake value (SUV of 7) with no hypermetabolic lesions reaching clinical significance in the lungs. A second PET-TC, repeated after 5 months, showed an increase in hypermetabolic captation with a SUV of 14.5 (Figure 1).

Computed tomography (CT) of the dorsal spine showed osteolytic process with a maximal diameter of 27 mm invading the right part of the vertebral body with erosion of posterior part of the body and the right pedicle (Figure 2).

A magnetic resonance image of the dorsal vertebra showed an hypointense signal lesion on T1-weighted and an hyperintense signal lesion on T2-weighted involved the right part of vertebral body and the right vertebral pedicle. The lesion produced a cortical breakdown with adjacent soft-tissue extension, occupation of the foramen of T11-T12, with compression of the dural sheath without contact with bone marrow. Moreover, there was a fluid-filled lesion of 3x4x10 cm in the context of the right paravertebral muscles (Figure 3).

A first computed-tomographic-guided biopsy performed in other Institute revealed bone marrow rich in plasmacells in aggregates but with a kappa/lambda ratio of 1:1. In bone marrow tissue were detected small areas of round to ovoid cells set in myxoid to oedematous stroma. Also a second biopsy performed was not enough for a definitive diagnosis.

Due to the impossibility to reach a diagnosis the patient was scheduled for frozen section

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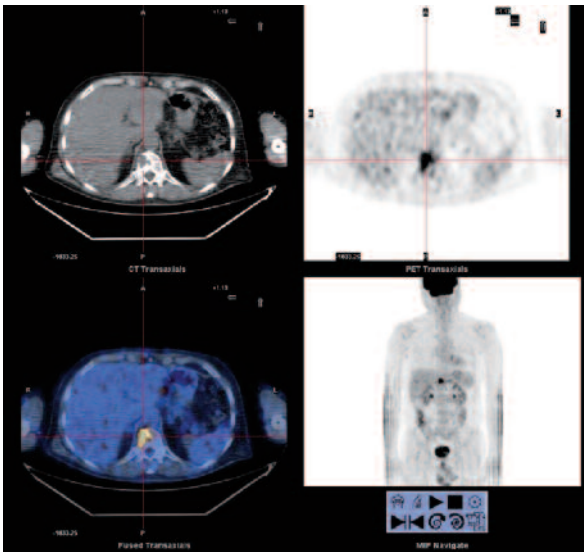

Figure 1. Positron emission tomography (PET-TC) revealed a hypermetabolic lesion in the vertebral body of T11 with a maximum standardised uptake value 14.5.

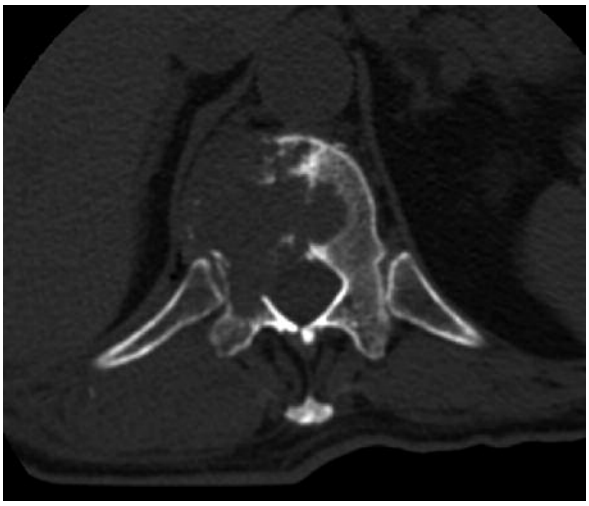

Figure 2. Thoracic axial CT showed a lytic lesion extending from the right side of corpus to the pedicle with cortical destruction measuring 27 cm.

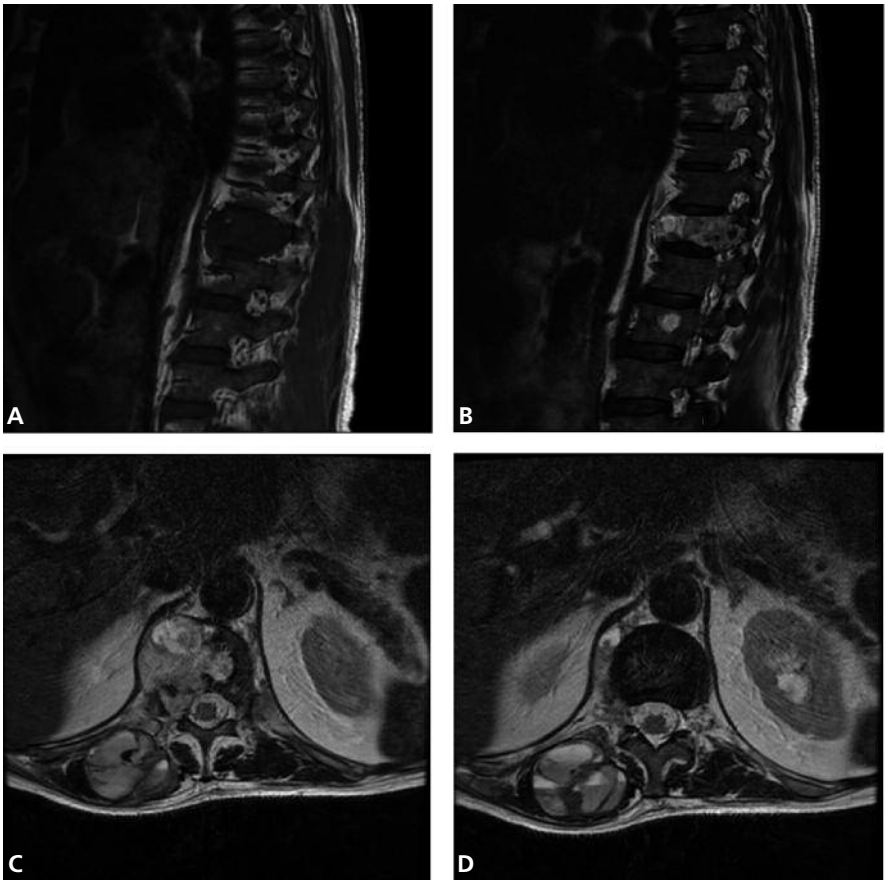

Figure 3. ***A,*** A sagittal magnetic resonance image of the dorsal spine showed hypointense signal lesion on T1-weighted and ***B,*** an hyperintense signal lesion on T2-weighted magnetic resonance image involving the right part of the eleventh vertebral body and the right vertebral pedicle. ***C-D,*** A transverse T2-weighted magnetic resonance image shows a fluid-filled lesion of 3x4x10 cm in the context of the right paravertebral muscles.

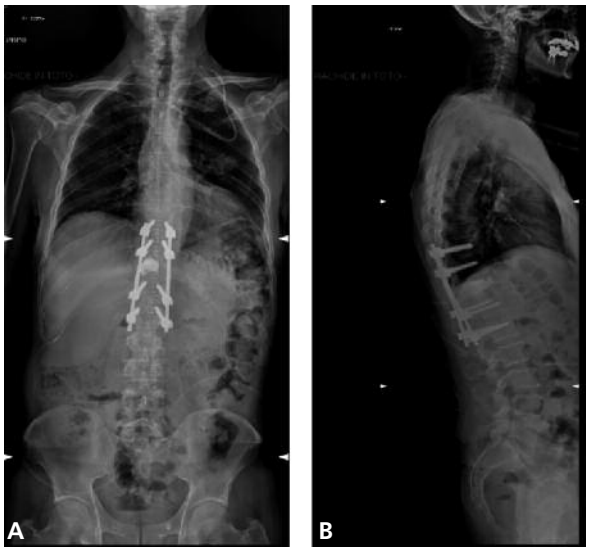

Figure 4. Anteroposterior ***(A)*** and lateral ***(B)*** post-operative X-rays of T11 laminectomy, curettage, vertebroplasty and T9-T12 osteosynthesis.

biopsy during surgical excision of the tumor. Two days preoperative selective arterial embolization was performed through the femoral artery with complete occlusion of XI left intercostal artery and XII right intercostals artery while from the X intercostals artery originated the Adamchiewicz artery. Through a posterior lumbar approach the patient underwent excision of paravertebral mass that was composed by blood and transpedicular biopsy of the vertebral body and frozen section that revealed showed plasma cells and round cells with plasmacytoid features morphologically consistent with plasmacellular dyscrasia. Then laminectomy of T11, curettage of the lesion, vertebroplasty and T9-T12 instrumentation was performed (Figure 4).

The tissue from the definitive curettage showed histologically a proliferation of neoplastic cells with epithelioid to plasmacytoid features, arranged in cords, focally embedded in a hyalinized to chondromyxoid matrix (Figure 5). Most neoplastic cells are immunopositive for EMA (Figure 6).

The immunohistochemical (IHF) profile documenting the expression of the following antigens: vimentin, epithelial membrane antigen (FOC), and CD 138. KI67 CLONE MIB 1 index was 12%. S-100 protein, glial fibrillary acid protein (GFAP), cytokeratin 7, cytokeratin 20, cytokeratin MNF116, CD 31, CD34, ERG, CD45 and other lymphoid markers and were all negative. Considering the diffuse immunohistochemical positivity for EMA and the morphological features the lesion was finally interpreted as a bone myoepithelioma.

One month after surgery a dorsal swelling in the lower left part of the wound appeared; the patient underwent new CT that documented the presence of hematoma. A second procedure was performed with debridment of the hematomas and post-operative selective embolization with a complete occlusion of X and XI intercostal artery. The histology report did not reveal pathologic cells.

Two months later, the CT scan of the chest with contrast medium documented a 1 cm lesion suggestive for local recurrence in T11 vertebral body; clinically there was also an ischemic complication of the wound and blood lost (Figure 7).

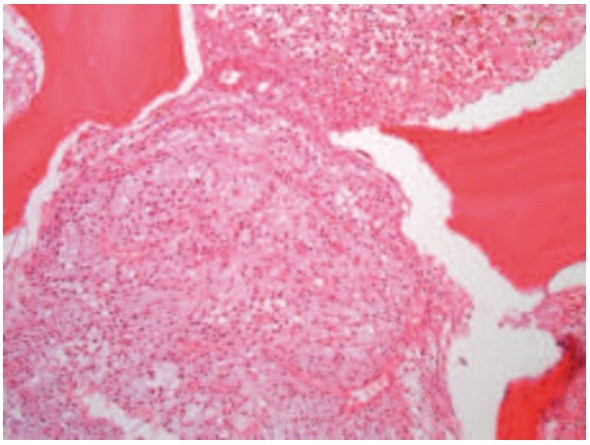

Figure 5. Neoplastic cells with epithelioid features arranged in nests and cords, focally embedded in a hyalinized to chondromyxoid matrix (hematoxylin and eosin,10x).

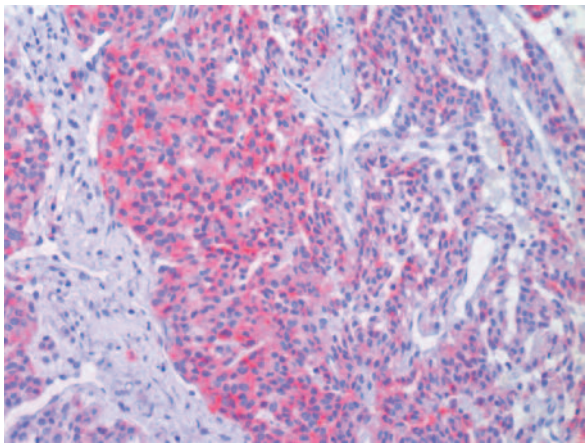

Figure 6. Most neoplastic cells are immunopositive for EMA immunostaining (20x).

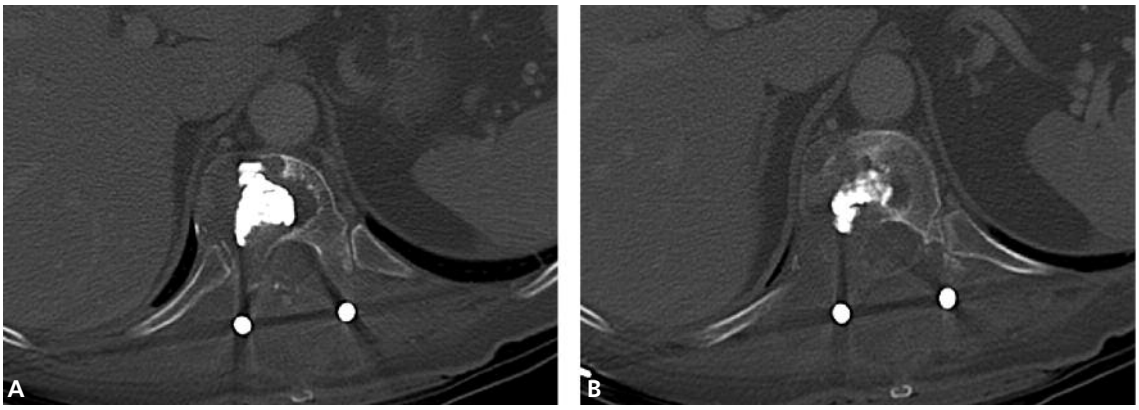

Figure 7. Thoracic axial CT showed a 3 cm lesion suggestive for local recurrence.

Through an anterior approach the patient underwent T11 corporectomy, reconstruction with titanium expandible cage and surgical debridment of the posterior wound (Figure 8).

Blood cultures isolated Sthaphylococcus Aureus treated with two-months levofloxacin 500 mg twice day and Rifampicin 600 mg once day and in the posterior wound was positioned VAC therapy maintained for 1 month.

After one month of VAC therapy treatment a vascularized cutaneous flap was positioned; after 3 weeks the patient underwent 10 sessions of hyperbaric therapy with complete healing of the posterior wound and then the patient was discharged from the hospital.

At the final F.U., 12 months after first surgery, the patient is completely pain free, there is no evidence of Local Recurrence in the spine and the skin is completely healed.

Discussion

Mixed tumours, parachordomas and myoepitheliomas are well-defined bone malignancies characterized by prominence of myoepithelial cells embedded in a hyalinized-to-chondromyxoid stroma. Thus, they are currently considered to belong to the same spectrum of differentiation[1,14]. Myoepitheliomas are distinguished from mixed

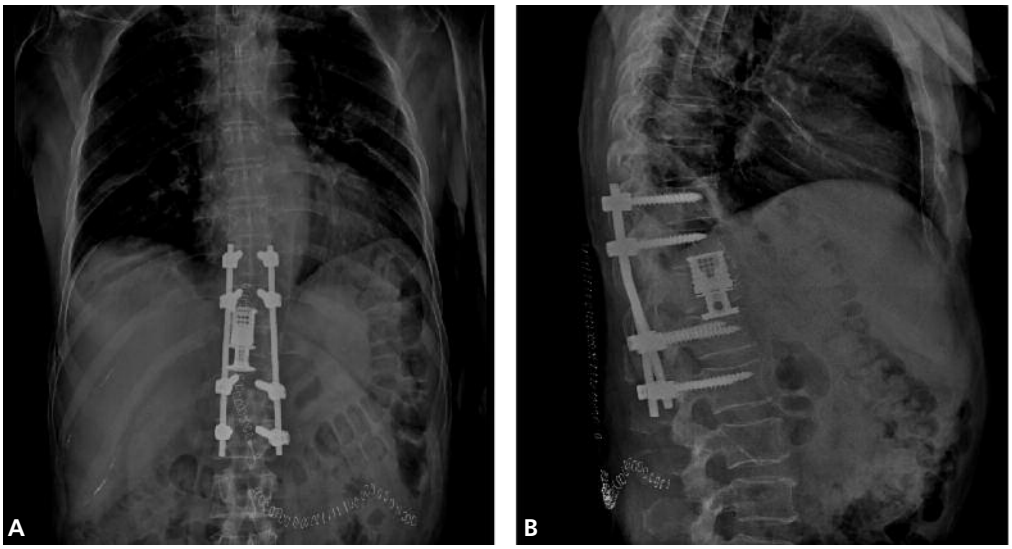

Figure 8. Anteroposterior ***(A)*** and lateral ***(B)*** post-operative X-rays of T11 corporectomy and reconstruction wit expandible cage.

Table I. Summary of published cases on myoepithelioma of the bone.

Case report	Location of the tumor	Treatment	IHF profile	Oncologic outcome
Rose et al[15]	Metacarp	Resection		Multiple bone involvement and lung metastasis. AWD at 15 years
De Pinieux et al[18]	Cuboid		CK+, EMA+, Vimentin+, S100-P+, CEA+, SMA+, NSE+	
McGough et al[2]	Tibia	Curettage. Resection and chemotherapy for local recurrence	The IHF profile revealed CK+, EMA+, Vim+, S100-P+, SMA+, HHF-35+, GFAP+.	RL at 2 years and lung metastasis at 3 years. DWD at 5 yrs
Kamiyama et al[19]	Ilium	Resection	CK(AE1/AE3)+, CK14+, EMA+, S100-P+, GFAP+, CEA+	CDF at 14 months
Alberghini et al[14]	Distal femur	Resection and prosthetic reconstruction	Vim+, CK(AE1/AE3)+, EMA+, HHF35+, SMA+, Calponin+, Caldesmon+, p63	Lung metastasis. AWD at 1 yrs
Park et al[5]	Humerus lesion with a satellite lesion	Marginal excision and adjuvant radiotherapy	CK+, p63+, S100-P+, Desmin+, Vimentin+	
Rekhi et al[16]	Ilium	En bloc resection	EMA+, CK(MNF116)F+, S100-P+, GFAP+, CK5/6+, Calponin+, p63+	CDF at 5 months
Current case	Spine	Debulking and vertebrectomy	vim+, FOC+, EMA+ and CD 138+	NED 1 RL at 5 months

AWD (alive with disease), DWD (died with disease), RL (local recurrence), CDF (continuously disease free), NED (no evidence of disease)

tumours by the absence of a definite ductal component, and from parachordomas by the presence of an intense and diffused cell cytoplasmic vacuolization[14]. Half of the reported cases have been considered malignant based both on clinicoradiological findings and histopathological features of local aggressiveness[5,14,17] as well as on pulmonary involvement[2,5,15,17].

Mixed tumor is the most common salivary-gland tumor and is most frequently found in the parotid gland[2,3,4]. Most intraosseous myoepithelial tumours occur in the maxilla[6]. Ferretti et al[6] reported three intraosseous myoepitheliomas arising from the maxillae of three young patients with a mean age of 15 years old. Previous reports have documented the bones as ectopic sites for malignant mixed tumor[2,15].

We describe the clinicopathological features of an examples of primary myoepitheliomas arising in the vertebral bone, a previously unreported location for these exceedingly rare tumors.

Some Authors reported cases of myoepithelioma occuring in the bone treated in different way and with different oncologic outcome (Table I).

Rose et al[15] described a case of metacarpal myoepithelioma treated with resection; the patient had multiple bone involvement, lung metastasis and was alive with disease at 15 years. McGough et al[2] reported a case of tibial myoepithelioma treated with curettage followed by resection and chemotherapy; the patient had local recurrence at 2 years, lung metastasis at 3 years and died less than 5 years after surgery. Kamiyama et al[19] described a case of myoepithelioma in the ileum treated with resection; the patient is free of disease at 14 months. Alberghini et al[14] described a case of distal femur myoepithelioma treated with resection and reconstruction; the patient was alive with disease at 1 year with lung metastasis. Park et al[5] described a case of myoepithelioma in the humerus with a satellite lesion that underwent marginal excision and adjuvant radiotherapy; the oncologic outcome was not available for this patient. Rekhi et al[16] reported a case of en bloc resection for a myoepithelioma located in the ileum; the patient was free of disease at 5 months (Table I).

In the current case the lesion was located in T11 and was treated in the first step with debulking,

then after 2 months the patient underwent corporectomy for local recurrence and the patient is free of disease at 12 months from the first surgery.

Myoepithelioma is an extremely rare entity, thus the immunohistochemical analysis was crucial to make the final diagnosis.

Musculoskeletal myoepithelioma is considered as a tumor with uncertain behavior. On CT and MRI, the tumour showed features of malignancy, i.e. cortical bone destruction and extension into adjacent soft tissues.

However, in view of a marginal excision, adjuvant radiotherapy was offered for better loco regional clearance, as rarely incompletely excised benign tumors have been known to recur or metastasize[2,15,16].

This case confirmed that myoepithelioma has an high risk of recurrence, while the metastatic potential is unknown. We recommend to resect myoepithelioma of the bone possibly en bloc and to avoid intralesional procedures such as debulking or curettage.

Radio and chemotherapy could be considered for local and systemic control in case of local recurrence or evidence of metastasis.

Conflict of Interest

The Authors declare that there are no conflicts of interest.

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