

CASE AND RESEARCH LETTER

[Translated article] Malignant Chondroid Syringoma of the Face With Distant Metastasis



Siringoma condroide maligno de localización facial con metástasis a distancia

To the Editor,

Malignant chondroid syringoma (MCS), which is also known as malignant cutaneous mixed tumor, is an extremely rare malignant neoplasm that originates in the adnexa. Approximately 50 cases have been reported in the literature to date.¹ In contrast with its benign counterpart (benign mixed tumor), which shows a predilection for men and generally affects the head, MCS is more common in women and mainly affects the limbs and trunk.^{2,3} Only 10 cases of MCS affecting the head have been reported to date.^{1–10} Below, we report the case of giant facial MCS with distant metastasis.

A 77-year-old man with myasthenia gravis and arterial hypertension under treatment presented with a painless tumor on the right cheek that had first appeared 2 years earlier. The lesion had grown abruptly in the previous 3 months. The patient reported difficulty chewing and reduced salivation. Physical examination revealed a firm multilobulated skin-colored subcutaneous tumor nodule measuring 11 cm × 6.7 cm × 3.5 cm with multiple telangiectasias adhering to deeper planes. The nodule affected the malar and lower right maxillary areas of the face (Fig. 1A and B). Enlarged lymph nodes were palpable in the ipsilateral axilla. Computed tomography of the head revealed a heterogeneous multilobulated lesion with hypodense areas measuring 10 cm at its widest diameter on the right side of the face. The lesion was in contact with the masseter and affected the ipsilateral parotid gland. Histology of an incisional biopsy specimen (hematoxylin–eosin) revealed tumor proliferation constituted by epithelial cells. These formed solid nests, cords, and gland-like structures, the main finding being foci of necrosis and other hypercellular foci with marked anisokaryosis, evident nucleoli, and several mitotic figures (Fig. 2). The surrounding stroma contained mainly



Figure 1 Firm multilobulated subcutaneous lesion adhering to deeper planes on the malar and lower maxillary area of the right half of the face. Frontal view (A) and lateral view (B).

sclerohyaline areas, myxoid areas, and other chondroid lesions. Immunohistochemistry revealed positive findings for carcinoembryonic antigen (CEA), epithelial membrane antigen (EMA), protein S-100, and cytokeratin and a KI-67 result of 25%. Based on these results, we diagnosed the patient's condition as MCS. Positron-emission tomography revealed hyperdense and hypermetabolic nodular lesions compatible with metastasis to the brain, liver, right axillary lymph nodes, and soft tissue in the right arm. The patient died of respiratory causes during admission.

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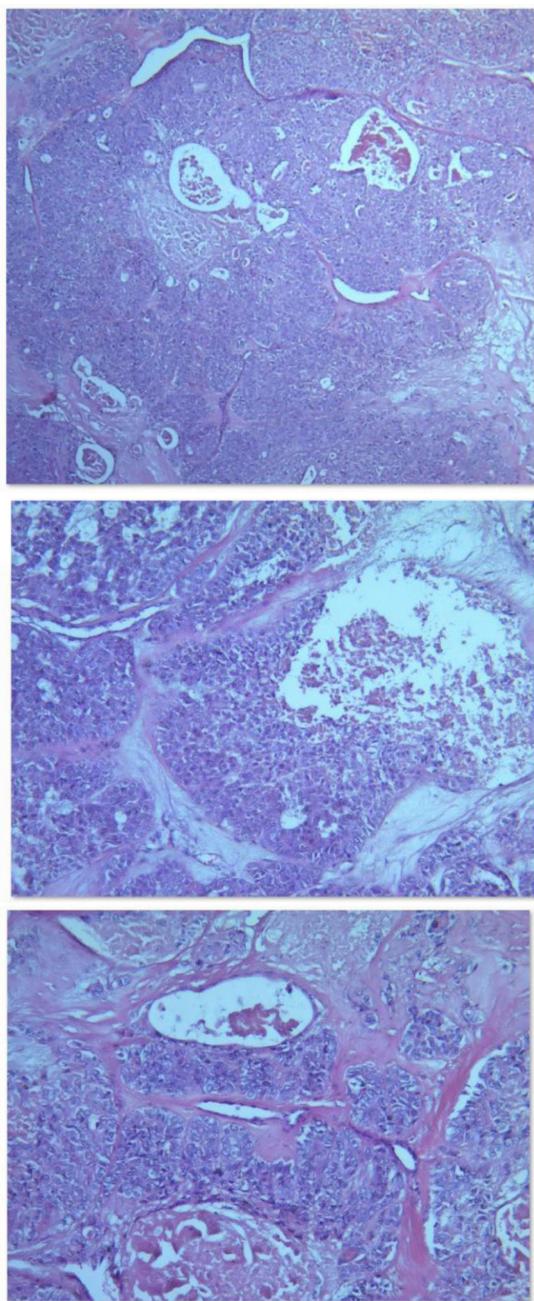


Figure 2 A, Solid area of the tumor with foci of necrosis and bleeding (hematoxylin–eosin [H–E], 50 \times). B, Epithelial cell nests with central necrosis and atypia surrounded by stroma with a mucinous appearance (H–E, 100 \times). C, Epithelial cell cords and nests surrounded by fibrous stroma, with foci of tumor necrosis (H–E, 100 \times).

First described by Hirsch and Helwig in 1961, chondroid syringoma is a benign tumor that originates in the eccrine or apocrine sweat glands.⁴ It is very uncommon, accounting for only 0.01% of all primary skin neoplasms. It presents as a solitary, slow-growing, painless intradermal or subcutaneous nodule that affects the head and neck.^{1,2} It more commonly affects middle-aged men. The size of the tumor can range from 2 mm to more than 1 cm; tumors measuring more than 3 cm are more likely to be malignant.^{1–3}

In contrast with its benign counterpart, MCS mainly affects women, has no predilection for age, and is more commonly observed on the limbs or the trunk.^{1,3} The tumor progresses slowly and is characterized by a final burst of rapid growth that can result in local destruction, metastasis, and death.⁵ The most common site of distant metastasis is the lung, followed by bone and the brain.^{2,4} In the case reported here, the tumor affected an elderly man, was located on the head and neck, and initially progressed slowly with a burst of growth later in its course. Local invasion was detected at diagnosis, as was metastasis to lymphatic tissue, the brain, liver, and soft tissue.

MCS can develop *de novo* or, more rarely, from an incompletely resected benign syringoma. Recurrence of the lesion should alert to the possibility of malignancy.^{1,2} In the present case, the tumor developed *de novo*, and the patient reported previous removal of facial lesions.

While some authors have pointed to local injury as playing a role in the etiology and pathogenesis of the tumor, no clear risk factors for either benign or MCS have been identified to date.^{1,4}

Histologically, chondroid syringoma is characterized by both epithelial and mesenchymal components. The mesenchymal component may have myxoid, chondroid, osteoid, adipose, and fibrous characteristics and contain epithelial cell nests that differ depending on whether the eccrine or apocrine glands are affected.¹ The criteria for malignancy include cytological atypia, increased mitotic activity, tumor necrosis, capsular invasion with compromise of deep structures, and, possibly, satellite nodules.^{3,7} Immunohistochemistry reveals the simultaneous presence of luminal cells that were positive for cytokeratin, CEA, and EMA and myoepithelial cells that were positive for cytokeratin, protein S-100, and vimentin.¹ Given its uncharacteristic clinical presentation, chondroid syringoma can only be diagnosed based on histopathology.

The differential diagnosis includes epidermal cysts, dermoid cysts, pilar cysts, pilomatrixoma, trichoepithelioma, schwannoma, and neurofibroma.^{1,2}

Chondroid syringoma is treated by complete excision. Control of the disease and prevention of recurrence are necessarily based on a wide free margin.^{2,4} Local radiotherapy is often unsuccessful, although bone metastases do respond to it. Combination chemotherapy has not been reported to be beneficial in patients with metastasis.^{2,4}

Given the high rate of recurrence of MCS (49%), the patient should be followed up closely. More importantly, follow-up should be long-term, since there have been reports of recurrence with dissemination to the lymphatic system and distant metastasis up to 12–20 years after the initial removal of the tumor.¹

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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