

Spinal Cord Compression caused by Metastatic Epithelial Myoepithelial Carcinoma of the Parotid Gland

Pages with reference to book, From 249 To 250

Irshad N. Soomro, Akber S. Hussainy, Rashida Ahmed, Sheema H Hasan (Departments of Pathology, The Aga Khan University Hospital, Karachi.)

Khalid Chishti (Departments of Surgery, The Aga Khan University Hospital, Karachi.)

Margaret H Pui, Shahzad A Khan (Departments of Radiology, The Aga Khan University Hospital, Karachi.)

Epithelial myoepithelial carcinoma (EMC) is a tumour of intercalated duct origin mostly arising in salivary gland. It occurs mainly in the 6th to 8th decades with a female predominance¹. Its infiltrative nature and presence of dual glandular secretory and myoepithelial differentiation are the main criteria of diagnosis. The tumour arises de novo. Most of the reported cases in the literature originated from the major salivary glands, particularly parotid gland. The clinical course was variable with lymph node metastasis at presentation and 23.5% local recurrence in a series of 21 patients. Twelve of these cases were tested and yielded a diploid DNA histogram by means of single cell scanning Sytophotometry². Thus, the tumour was considered to have low malignant potential. However, some cases have been reported to run an aggressive course. Of seven cases, two were reported to metastasize and three recurred locally³. We recently encountered a case of EMC presenting with cord compression.

Case Report

A 65 year old lady presented with weakness of both lower limbs. She complained of back pain for four months.

On examination, she had tenderness of the left paraspinal, retroscapular and sternal regions. The muscle power was grade 3/5 in the left and 4/5 in the right leg. Sagittal and axial T1 and T2-weighted magnetic resonance images of the thoracic spine showed destructive lesions in the T3 and T4 vertebrae compressing the spinal cord. The intervening disc was also involved (Figure 1).

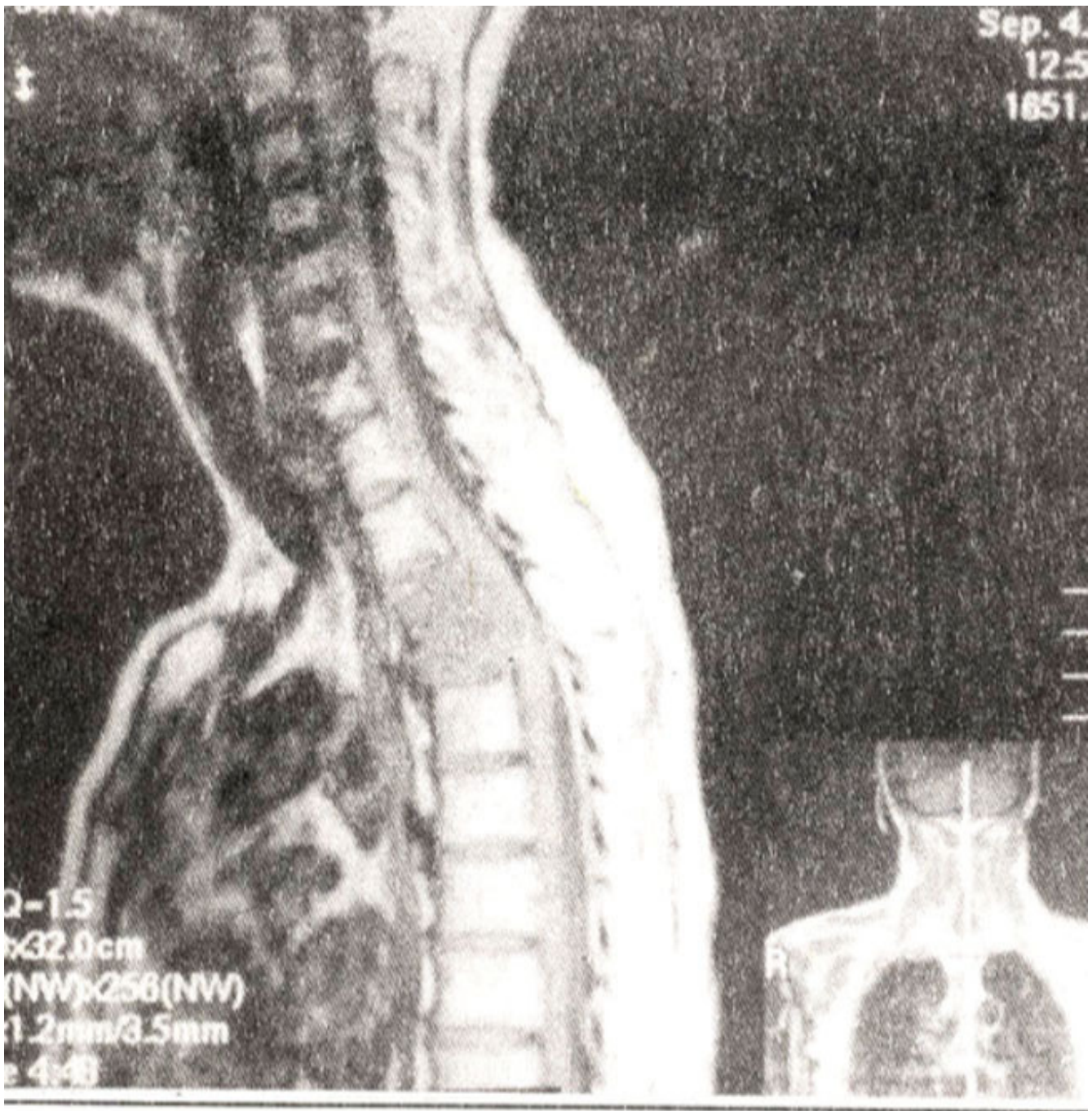


Figure 1.

Metastatic disease or tuberculosis was suspected. Decompressive surgery was performed with removal of T3 and T4 vertebral bodies and insertion of fibular graft between T2 and T5. Frozen section of the removed tissues revealed an infiltrative lesion of duct-like structures and fibromyxoid stroma. The differential diagnosis included chordoma and metastatic carcinoma. Because the clinical information of multiple lesions was not available, the diagnosis of chordoma was suspected. A review of permanent paraffin-embedded sections suggested metastatic disease. Upon inspection of the medical record, it became apparent that the patient had undergone removal of parotid gland lesion eighteen months previously. The lesion had been enlarging slowly over ten years and was considered to be a pleomorphic adenoma at another institution. The previous histological slides were obtained and reviewed. The lesion comprised infiltrative ductal structures of epithelial cells surrounded by myoepithelial cells. There was evidence of perineural invasion. Immunohistochemically, the epithelial

cells stained for cytokeratin markers, AE1/AE3 and CAM 5.2. The myoepithelial cells showed staining with antibody against smooth muscle actin (Figures 2 and 3).

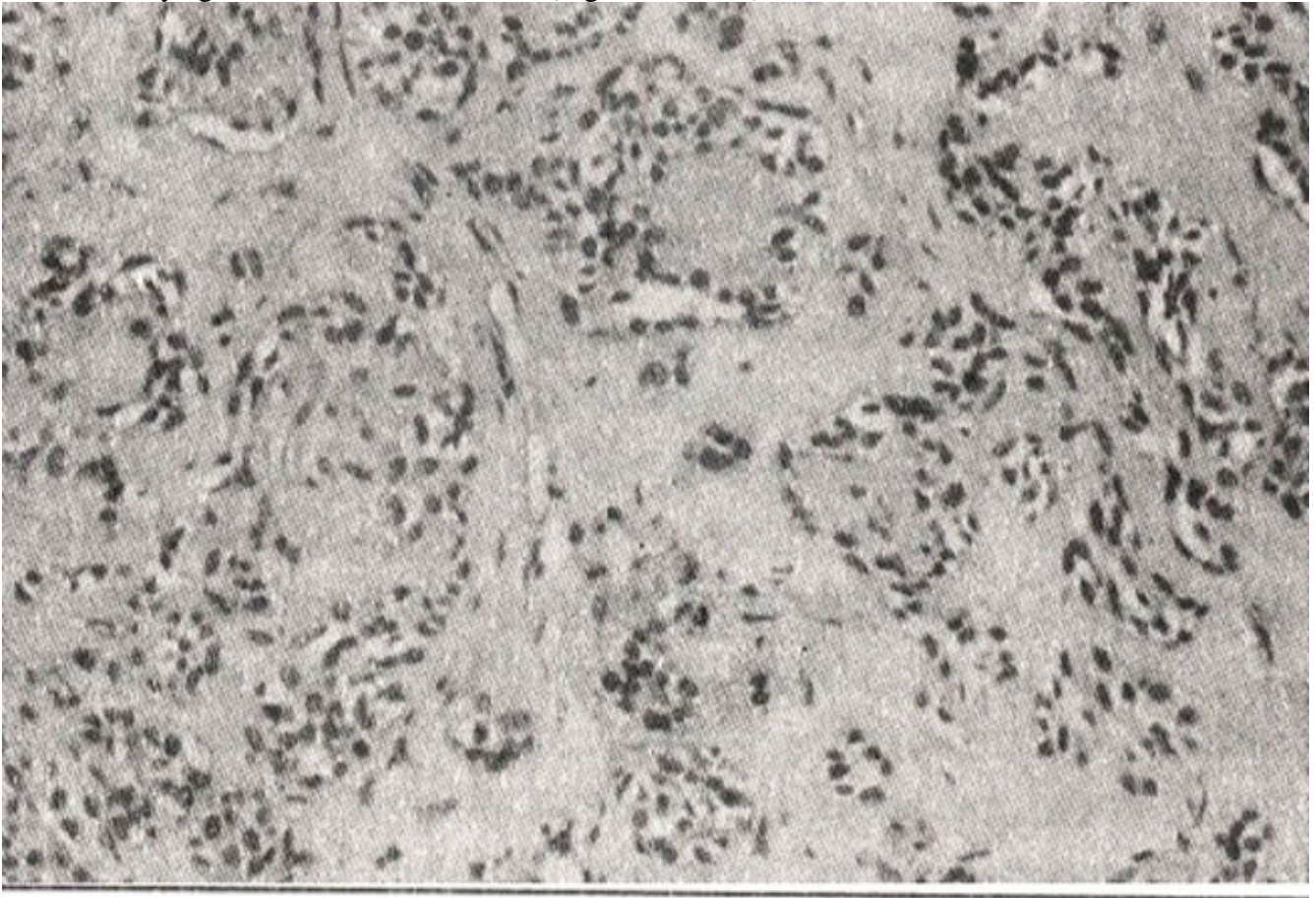


Figure 2.

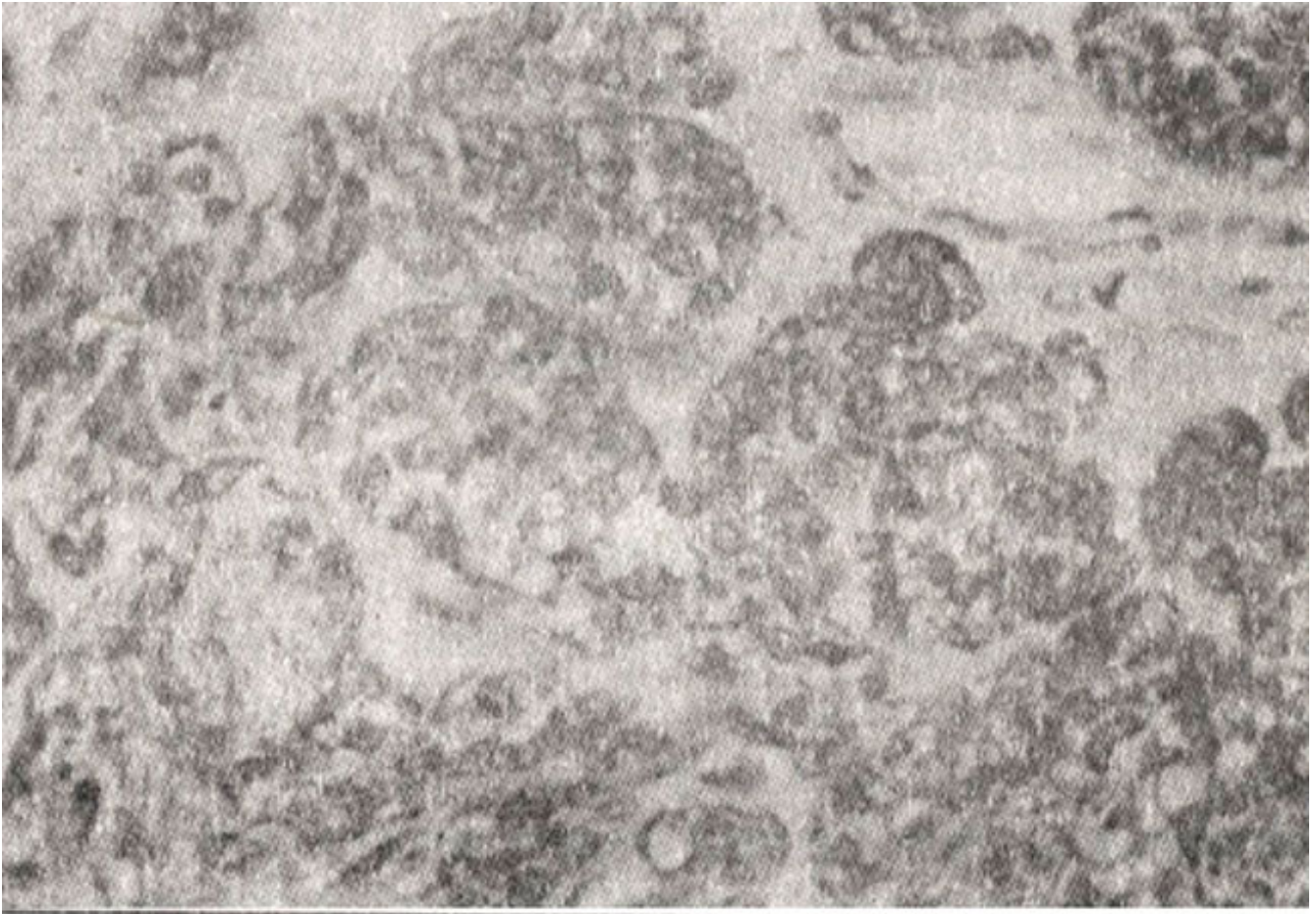


Figure 3 (a)

A diagnosis of metastatic EMC was made. The clinical course was rapid deterioration with recurrent spinal cord compression, widespread bone and lung metastases.

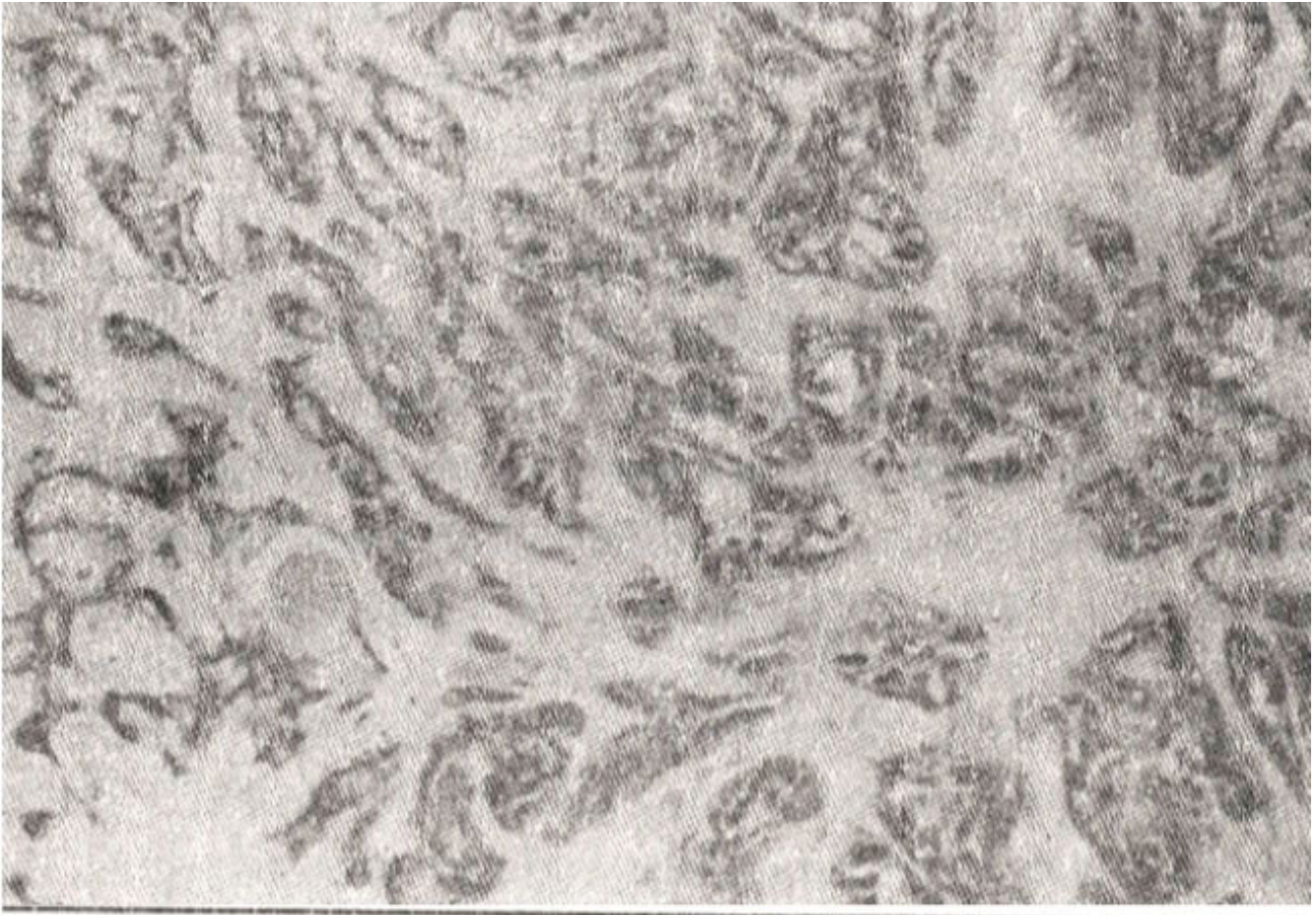


Figure 3 (b).

Discussion

This case report describes spinal cord compression caused by bone metastases from EMC. In the past this tumour was considered to be benign and was called clear cell adenoma. Its malignant potential has only recently been known⁴. Several authors reported this lesion to be low-grade malignancy^{2,4}. However, biologically aggressive EMC has been reported. Fonseca and Soares described 22 cases with tumor recurrence in 50% and mortality of 40%. Nuclear atypia, clear cell type and aneuploid DNA content was considered indicators of poor prognosis. Our patient appeared to have a low-grade malignancy of more than ten years. This case was misdiagnosed as pleomorphic adenoma which can look like a cellular infiltrative lesion. The presence of perineural invasion raised the suspicion of EMC. Histologically, EMC comprises of small ducts with a double cell lining surrounded by a basement membrane. The inner cells are epithelial and outer cells myoepithelial; the latter usually processing clear cytoplasm. There is variable degree of intervening hyalinized stroma⁴. These features, when seen in salivary gland and supported by immunocytochemistry, pose little diagnostic problem. To knowledge, clinical presentation of spinal cord compression by EMC has not previously been described. Chordoma was initially suspected. Both EMC and chordoma are formed by ductal structures. Immunohistochemically, both tumours are positive for cytokeratins and S-100. The differentiating feature is the presence of smooth muscle actin in the basement membrane of EMC ductal cells and not in chordoma. Further clinical information enabled us to include smooth muscle actin in the panel which strongly decorated the ductal structure of the EMC in our patient. Metastatic EMC should be distinguished from metastasizing malignant mixed tumour of the salivary

gland. Carcinomas are known to develop in long-standing pleomorphic adenomas. The carcinomatous element displays both cytomorphic abnormalities and infiltration of the surrounding tissues. There is variable proportion of myxoid, chondroid and hyalinized stroma between the tumour cells. The carcinomatous component in a series of 37 cases included 13 ductal, 10 undifferentiated, 9 terminal ductal, 3 myoepithelial and 2 unclassified lesions. The differential diagnosis of metastatic mixed tumour includes chordoma and chondrosarcoma⁷. EMC arising in a pleomorphic adenoma has recently been described⁸. Although the existence of this has been disputed by some⁹. The initial histology, metastatic disease and immunohistochemistry clearly ruled out malignant mixed tumour in our patient. This case shows convincing evidence of slow biologic aggressiveness of EMC. Complete clinical information and a close liaison between clinician and histopathologist are crucial for its diagnosis.

Acknowledgement

We would like to express thanks to Professor Sebastian Lucas at the Department of Histopathology, UMDS, St. Thomas' Hospital, London.

References

1. Corio RL, Sciubba JJ, Brannon RB. et al. Epithelial myoepithelial carcinoma of intercalated duct origin. A clinicopathologic and ultrastructural assessment of sixteen cases. *Oral. Surg. Oral. Med. Oral. Pathol.*, 1982;53:280-87.
2. Hamper K, Briggmann M, Koppermann R, et al. Epithelial myoepithelial duct carcinoma of salivary gland: A follow up and cytophotometric study of 21 cases. *J. Med. Oral. Pathol.*, 1989;18:299-304.
3. Collina G, Gale N, Visona A, et al. Epithelial myoepithelial carcinoma of the parotid gland: A clinicopathologic and immunohistochemical study of seven cases. *Tuinoi*, 1991;77:257-63.
4. Simpson RH, Clarke Ti, Sarsfield PT, et al. Epithelial myoepithelial carcinoma of salivary glands. *J. Clin. Pathol.*, 1991;44:4 19-23.
5. Fonseca!, Soares J. Epithelial myoepithelial carcinoma of the salivary glands: A study of 22 cases. *Virchows Arch.*, 1, 1993;422:389-96.
6. Tortoledo ME, Luna MA, Batsakis JO. Carcinomas ex-pleomorphic adenoma and malignant mixed tumours. Histomorphologic indexes. *Arch. Otolaryngol. Head Neck Surg.*, 1984;110:172-76.
7. Ellis GL, Auclair PL. Atlas of tumour pathology. Tumour of the salivary gland. Editor Juan Rosai. Washington, D.C., Armed Forces Institute of Pathol., 1994.
8. Mc-Cluggage WG, Primrose Wi, Toner PG. Myoepithelial carcinoma (malignant myoepithelioma) of the parotid gland arising in a pleomorphic adenoma. *J. Clin. Pathol.*, 1998;51:552-56.
9. Jones TJ. Epithelial - myoepithelial carcinoma. *J. Clin. Pathol.*, 1998;51:944.