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Case Report

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# Occasional diagnosis of metastatic pleomorphic adenoma

## Abstract

**Background:** Pleomorphic adenoma is considered a biologically benign tumour. However, rare cases of metastases, showing the same histological benign features of the primary tumour, are reported, thus justifying the inclusion into the World Health Organization (WHO) definition "Malignant Mixed Tumour", even in the absence of histological atypias.

**Case report:** We report the case of a 45years-old woman affected by recurrent pleomorphic adenoma since about the age of 30years. During pre-surgical x-ray chest, performed as routine before excision of the last recurrence of pleomorphic adenoma, bilateral lung nodules were discovered, which ended up to be lung metastases.

**Discussion:** it had been proved by evidence that pleomorphic adenoma recurrences significantly increase the possibility both of malignant transformation of the disease and malignant behaviour recurrences that could develop distant localization, though histologically benign.

**Conclusion:** in management of primary pleomorphic adenoma radical surgical excision is crucial, to prevent first of all local recurrences and then distant spread.

**Keywords:** pleomorphic adenoma, malignant mixed tumour, lung diseases, neoplasm metastasis, x-ray, tumour, malignant, disease, radical, metastasizing, carcinosarcoma, pathologies, mesenchymal, salivary glands, metastases

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**Abbreviations:** WHO, world health organization; FNAB, fine needle aspiration biopsy; CECT, contrast-enhanced chest computed tomography; HU, hounsfield units; PET-CT, positron emission tomography – computed tomography

## Introduction

Pleomorphic adenoma is the most common benign neoplasm of salivary glands. It is considered a biologically benign tumour, but, occasionally, it acquires the capability of metastasizing, locally or even distantly. With regards to this, World Health Organization (WHO) classified three different pathologies, included in the definition of "Malignant Mixed Tumour": the Carcinoma Ex Pleomorphic Adenoma, the Carcinosarcoma, both characterized by a histologically malignant transformation of the epithelial component by itself or in combination with the mesenchymal-like one, respectively, and the Metastatic Mixed Tumour. It has been postulated that both Carcinoma ex Pleomorphic Adenoma and Carcinosarcoma probably results from the genetic instabilities and accumulation of key genetic alterations in long-standing pleomorphic adenoma.[1] The Metastatic Mixed Tumour is defined as a histologically benign Pleomorphic Adenoma, typically recurrent, that manifests local or distant metastasis. We present a case of recurrent pleomorphic adenoma developing lung metastases.

## Case presentation

A 45years-old woman with history of recurrent oral pleomorphic adenoma was referred to the Radiology Department to perform a routine chest x-ray pre-operative examination before a planned excision of a left submandibular swelling (Figure 1), present since about three years and diagnosed as a pleomorphic adenoma at a recent fine needle aspiration biopsy (FNAB). The patient had already undergone surgical excision of pleomorphic adenoma in the left part of the palate in 1980 followed, ten years later, by another excision of recurrent disease in the same site. She was free from comorbidities.

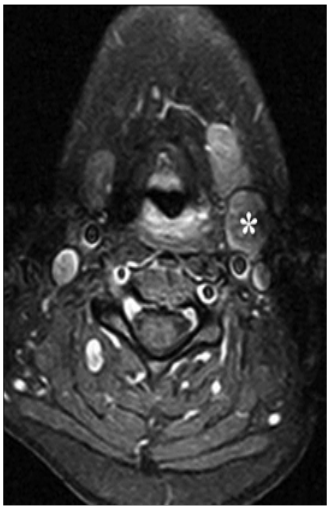

**Figure 1** Axial T1-weighted post-contrast, fat suppressed MR section showing a well-defined, contrast-enhanced, lesion (asterisk), with a maximum diameter of 2cm, between the sternocleidomastoid muscle and left submandibular gland. The lesion was diagnosed as pleomorphic adenoma at FNAB.

At chest x-ray two well-defined radio-opaque nodules in both the lower lung fields were depicted, the biggest one in the right lung, projectively in the retro-cardiac region, with a maximum diameter of about 3cm (Figure 2). Contrast-enhanced chest Computed

 

Tomography (CECT) was requested to better characterize the lesions; the question was to make a correct differential diagnosis between inflammatory or benign diseases, such as bronchial cyst or vascular malformation, or neoplastic lesions, of primary or secondary nature. Chest CT, performed with a 64-slice equipment after the intravenous administration of high concentrated contrast media (370mgI/ml) confirmed the presence of two pulmonary lesions, in the right and left inferior lobes, with a maximum diameter of 3.5cm, and 2cm, respectively. Both the lesions had well-defined, smooth margins, mean density of 12-18 Hounsfield Units (HU) and homogeneous contrast enhancement; they slightly displaced the adjacent blood vessels and sub-segmentary bronchi; there were neither evidence of infiltration of surrounding normal pulmonary parenchyma nor signs of cavitation, calcification or necrosis into the nodules (Figure 3). Successively, the patient underwent PET-CT examination that documented a high metabolic activity of both pulmonary lesions (Figure 4) and, therefore, a fine needle aspiration biopsy of the right lung lesion was done, in order to histologically define the mass.

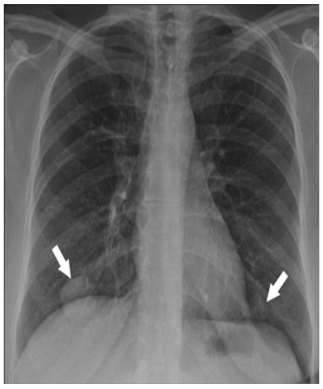

**Figure 2** Posterior-anterior chest-x-ray projections showing two well-defined radio-opaque nodules (arrows), projectively in both lower lung fields.

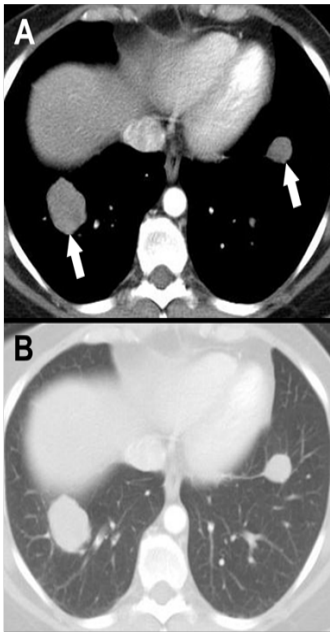


**Figure 3** Contrast-enhanced chest Computed Tomography (CECT) sections with mediastinal and lung parenchyma windows, respectively, showing in anterior-basal segment of the inferior pulmonary lobes two, well-circumscribed lesions (arrows in A), with a maximum diameter of 3.5cm, and 2cm, respectively, mean density of 12-18 Hounsfield Units (HU) and homogeneous contrast enhancement.

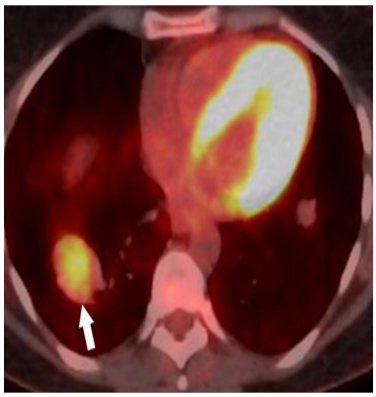

**Figure 4** PET-CT axial fusion image demonstrating the high metabolic activity particularly of the right pulmonary lesion (arrow, SUV 4.24).

The patient was then hospitalized and underwent, after a collegial decision between maxillofacial and thoracic surgeons, surgical excision of both the submandibular lesion and the one in the left inferior pulmonary lobe. The histological analysis of the lung samples showed a clear cells salivary gland tumour, with low-grade of malignancy.

Subsequent atypical resection of the right inferior pulmonary lobe was performed and at histopathology even the second pulmonary lesion turned out to be of the same histological type as the lesion in left lung. Up to date, the patient is free from local or distant recurrences, as documented by CT and MRI examinations performed six and twelve months after surgery, respectively.

## Discussion

Pleomorphic Adenoma is the most common neoplasm affecting the salivary glands, accounting for about 60% of all salivary neoplasm's, with a reported annual incidence of 2.4-3.05 cases per 100,000 people.[1] It is known as a benign tumour, with no cellular atypias, usually circumscribed by a fibrous pseudo-capsule of varying thickness, composed of a combination of proliferating glandular and myoepithelial cells and mesenchymal or stromal component.[2] The typical clinical presentation, as happened to the patient described in this report, is a slow-growing, painless, mobile, singular nodular mass.

In our case, the identification of pulmonary lesions was an accidental finding, during a routine chest x ray, preliminary to surgical excision of a pleomorphic adenoma relapse. The radiological appearance was not sufficient to make the differential diagnosis between benign lesions and metastasis so a CT scan was required. The contrast media uptake hinted to a solid nature of the lesions, excluding the possibility of bronchial cysts. However, the patients had no history of malignant tumours that could lead us think of metastasis. Only the history of recurrent pleomorphic adenoma could have led us closer to the appropriate diagnosis, even if a core biopsy was nevertheless required to get the conclusive diagnosis.

Local recurrence of Pleomorphic Adenoma after surgery has been described in 1 to 5% of cases, malignant degeneration has been reported in 2-9% of cases, while metastasizing pleomorphic adenomas without histological evidence of malignancy have rarely been reported[3]. In these cases, both the primary tumour and the metastasis are composed of a typical mixture of benign-appearing epithelial and mesenchymal components. The histology of primary pleomorphic adenoma has not been demonstrated to be predictive regarding its likelihood to metastasize.[3,4] In a literature review by Nouraei et al.,[5] between

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1953 and 2005, only 42 patients with metastasizing pleomorphic adenoma were reported. About 80% of these patients had had at least one local recurrence of primary tumour prior to the detection of a distant metastasis, likely it happened to our patient. Lung is the third most common site for distant metastases (36%), after bone (45%) and head and neck (43%). Furthermore, metastasectomy demonstrated to confer statistically significant survival advantage.[5]

It is reported a strong association between the incomplete surgical excision of primary tumour and recurrences, first, and development of distant metastases later. Unfortunately, no information was available regarding the surgical excision that our patient underwent about 30 and then 20years earlier. The possible reason for recurrences or persistence of pleomorphic adenoma may include either the tumour's ability to invade the capsule that could have variable thickness or the presence of tumour micronodules bulging through the capsule that could escape surgical enucleation. It has also been postulated that previous radiotherapy of primary site or surgical manipulation could be responsible for increased vessels permeability, thus making possible distant spread of the disease. Notwithstanding, the reasons of the great variance between histologically benign hallmark of metastasizing pleomorphic adenoma and its biologically malign behaviour are not completely understood. The most important step in initial management of primary pleomorphic adenoma is radical, meticulous, surgical excision that is considered to prevent local recurrences and distant spread.

For benign salivary glands tumours, traditional management is surgical dissection, even though there is currently no agreement on its extent to obtain adequate margins. Intra-operative tumour spillage carries an increase in the rate of recurrence over a prolonged period and therefore long-term clinical observation follow-up is recommended. In such cases the possibility of an adjuvant RT should be discussed in a multidisciplinary team setting, but the use of RT is controversial and is generally not recommended especially in younger patients due to the risk of radiation-induced tumours.

Recurrent benign parotid tumours should be treated with surgical resection, tailored to tumour size and to its histological type. Careful pre-operative ultrasound marking may be helpful. Even in these cases the possibility of post-operative RT to reduce other relapses should be discussed.[6]

To our knowledge there are no current guidelines that state the necessity and timing of imaging follow up of patients with recurrent pleomorphic adenoma, especially with regards to distant localization, given its rarity. In our experience a clinical follow up is crucial and should be supported by ultrasound examination if local recurrence is suspected. To conclude, distant metastases, despite being rare, cannot be excluded in case of neck, bone or pulmonary distant lesions discovery, especially in patients with no history of malignant tumours.

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## Conflict of interest

Author declares that there is no conflict of interest.

## References

1. Rodríguez FJ, Mateos MM, Martínez TFJ, et al. Metastatic benign pleomorphic adenoma. Report of a case and review of the literature. *Med Oral Patol Oral Cir Bucal*. 2008;13(3):E193–E196.
2. Tumors of the salivary glands. *International Agency for Research on Canc*er. 2009. p. 208–281.
3. Marioni G, Marino F, Stramare R, et al. Benign metastasizing pleomorphic adenoma of the parotid gland: a clinicopathologic puzzle. *Head Neck*. 2003;25(12):1071–1076.
4. Wenig BM, Hitchcock CL, Ellis GL, et al. Metastasizing mixed tumor of salivary glands. A clinicopathologic and flow cytometric analysis. *Am J Surg Pathol*. 1992;16(9):845–858.
5. Nouraei SA, Ferguson MS, Clarke PM, et al. Metastasizing pleomorphic salivary adenoma. *Arch Otolaryngol Head Neck Surg*. 2006;132(7):788–793.
6. Sood S, Mcgurk M, Vaz F. Management of salivary gland tumours: United Kingdom National Multidisciplinary Guidelines. *J Laryngol Otol*. 2016;130(Suppl 2):S142–S149.