Metastatic pleomorphic adenoma to the supraspinatus muscle: a case report and review of a rare aggressive clinical entity

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ABSTRACT

We report a case of a 65-year-old female with a recurrent right parotid pleomorphic adenoma (PA) 24 years after initial surgical excision. Positron-emission tomography (PET) and computed tomography (CT) demonstrated an unusual suspicious FDG-avid erosive rim enhancing mass centered in the right supraspinatus muscle. Cytology from CT-guided aspiration of the mass was consistent with a histologically benign PA, and the patient was diagnosed with metastatic pleomorphic adenoma (MPA). The patient later developed diffuse pulmonary metastases and died within 3 months. MPA, although rare, is recognised as a potentially lethal malignant complication of recurrent or longstanding benign PA. As no biochemical or genetic parameters are predictive of malignant change, patients presenting with recurrent PA should be considered for screening for metastatic disease.

CASE REPORT

We present the case of a 65 year old female with a prolonged history of recurrent pleomorphic adenoma. Initial presentation was in 1988 with a right parotid mass, which was excised with clear surgical margins and confirmed histologically a pleomorphic adenoma (PA). In 2006 she represented with increasing difficulty swallowing, and imaging confirmed local recurrence with extension from the right parotid space into the right parapharyngeal and carotid spaces. Mandibulotomy and deep parotidectomy with adjuvant chemotherapy and radiotherapy (discontinued against medical advice prior to completion) were performed, and a percutaneous gastrostomy feeding tube was inserted radiologically. Positron emission tomography (PET) in 2008 demonstrated fluoro-deoxyglucose (FDG) uptake in right prevertebral spaces, which was attributed to post-surgical changes considering the benign nature of pleomorphic adenoma.

She represented in 2012 with slurred speech, deviation of the angle of the mouth to the right, and an aspiration pneumonia. Contrast-enhanced computed tomography (CT) demonstrated a 6.6 cm solid and cystic mass extending trans-spatially from the right parotid space, consistent with recurrence of the PA with superimposed post-
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Etiology & Demographics

Pleomorphic adenoma (PA), the most common tumour of the salivary glands, mainly occurs in the parotid superficial lobe (85%), and less frequently in submandibular, lingual and minor salivary glands [1]. Also known as benign mixed tumours, PAs comprise varying proportions of epithelial, myoepithelial and stromal components, and are encapsulated with a delicate pseudocapsule. Incidence of PA varies from 2 to 3.5% with a female preponderance, most commonly presenting in the 3rd to 6th decade [2]. The only known risk factors include neck irradiation, and a potential link with simian virus [3].

While PA is considered pathologically and clinically benign, it can undergo malignant transformation in situ, termed carcinoma ex-pleomorphic adenoma (CEPA), with an incidence of 2 to 7% of PAs [4,5]. A much rarer phenomenon can also occur whereby a PA demonstrates clinically malignant behaviour while remaining histologically benign, termed metastatic pleomorphic adenoma (MPA). MPA is recognised as a histological sub-type of PA by the World Health Organisation [6], and is considered clinically distinct from CEPA (although some authors consider metastatic pleomorphic adenoma (MPA) an intermediate link in transformation from PA to CEPA) [7].

The exact incidence of MPA has not been conclusively established, although it is certainly very rare. For example, a 2007 review paper documented 52 cases in the literature over 50 years [8]. In the majority of these cases there was an association between MPA and longstanding or recurrent PA (with an average presentation-to-metastasis latency of 16 years). An association with previous incomplete surgery was also reported, suggesting that incomplete enucleation or rupture of the thin capsule is related to later metastatic behavior [8]. Seeding to the lungs by aspiration of PA [9], and a lymphatic metastatic route have both been proposed [10]. Despite these associations, however, the mechanisms underlying the metastatic behavior of MPA are far from clear, and no histological or molecular genetic alterations correlate with risk of subsequent development of MPA from the familiar benign PA [11].

Clinical and imaging findings

Radiological characterization of a metastatic pleomorphic adenoma (MPA) will invariably begin with identification of a PA, which usually presents as a slow-growing, painless parotid mass [12]. PA in the deep lobe of the parotid gland may present as an oral retro-tonsillar mass or parapharyngeal space tumour. Rapid enlargement, multiple symptoms or facial nerve involvement should raise concern for malignant change [12].

On contrast-enhanced computed tomography (CT), PAs are typically smooth, homogeneous, well-circumscribed tumours, with higher attenuation than surrounding parotid parenchyma, demonstrating poor and delayed enhancement [13]. Imaging findings can vary with tumour size. Small PAs are more circumscribed and homogeneous with earlier enhancement, whereas larger PAs have more lobulated contours with poor enhancement, often with necrotic, calcified or haemorrhagic areas [14].

On magnetic resonance imaging (MR), PAs are usually well-circumscribed T1 hypointense and T2 hyperintense, often with a T2 hypointense rim representing the fibrous capsule [15]. Again larger PAs yield a more heterogeneous signal and irregular border. An ill-defined border, and heterogeneous signal intensity can be suggestive of carcinomatous transformation. Recurrent PA can appear as subcutaneous T2 hyperintense lesions in the operative bed, subcutaneous fat, and/or spaces adjacent to the parotid in patients with prior parotidectomy [16]. On US a PA is generally a smooth round hypoechoic mass with a distinct often lobulated border [17]. Percutaneous ultrasound-guided biopsy is feasible, with low tumour seeding rates, and low risk of facial nerve injury provided meticulous technique used [18].

In recurrent or longstanding PA, a high index of suspicion for MPA is advisable, and additional imaging to rule out metastases by full body CT, MR, and/or positron emission tomography (PET) imaging can be considered. To our knowledge, this is the first reported case of MPA metastasizing to the rotator cuff muscles. Examples of recently reported sites of metastases in MPA include the mandible [19], a solitary kidney mass [20], pulmonary nodules [21,22], intra-oral deposits [23], intra-vascular endothelial deposits [24], liver [25], sphenoid sinus [26], scapula [27], calvarium [28], ribs [29], and mediastinal lymph nodes [30]. Overall, the most commonly reported sites of metastases are bone (45%), followed by head and neck (43%) and lung (36%) [8]. Interestingly, metastases do not always arise concurrently with local recurrence of the primary PA, as reported in a case of

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pathological fracture due to a glenoid MPA [31], and a metastatic supraclavicular lymph node [32].

**Treatment and prognosis**

Metastatic pleomorphic adenoma (MPA), because of its association with longstanding or recurrent pleomorphic adenoma (PA), can be prevented by adequately treating the initial presentation PA, which involves surgical excision [8]. Historically PAs were removed by enucleation, which resulted in recurrence rates of 20 to 45% presumably due to disruption of the thin capsule [33]. Superficial or total parotidectomy (with facial nerve sparing) has reduced recurrence rates to 5% [33]. If a MPA is diagnosed sufficiently early, a surgical metastectomy does yield a statistically significant survival advantage over non-operative treatment [8]. Treatment of the local recurrence is no means standardized and would generally involve wide surgical excision [34] and/or radiotherapy [35].

Prognosis with MPA certainly justifies its classification as a malignancy, as mortality due to disseminated metastatic disease is reported as high as 50% over 5 years. There is decreased survival if the MPA occurs within 10 years from initial PA, or with multiple metastatic sites [8]. In a 2005 editorial article, Bradley [36] advised that because no molecular or histopathological parameters can distinguish MPA from PA, it should be considered a low-grade, potentially lethal malignant disease.

**Differential Diagnoses**

There is considerable overlap between the radiologic appearance of parotid masses, and biopsy is often required for definitive histological or cytological diagnosis. The differential for benign entities includes Warthin’s tumour and monomorphic adenoma, both of which can have large cystic or necrotic areas, which aids in their differentiation from pleomorphic adenoma (PA). A Warthin’s tumour typically demonstrates more rapid enhancement and washout pattern than a monomorphic adenoma, which tends to be a smaller < 3cm and more circumscribed lesion [15,38]. Lymphadenopathy is also an important differential, and PAs will have a higher T2 signal intensity than adjacent intraparotid lymph nodes [15]. It is not possible to reliably distinguish between benign and malignant parotid lesion on PET imaging, as benign lesions often demonstrate high uptake of FDG [39].

Malignant masses include mucoepidermoid, adenoid cystic carcinoma, malignant lymphoma, and rarer entities such as acinic or squamous cell carcinoma, pleomorphic sarcoma (malignant fibrous histiocytoma), and metastatic disease [15]. A mucoepidermoid carcinoma is the most common parotid malignancy, and demonstrates predominantly low signal intensity on T2, with T1 and T2 hyperintense mucin-containing cysts. Adenoid cystic carcinoma, the second most common parotid malignancy demonstrates a predominantly low T2 signal with a large T1 hyperintense area of haemorrhagic necrosis, and can be difficult to distinguish from PA [15]. Malignant lymphoma often presents with multiple well-defined lobulated lesions, with low signal on both T1 and T2 [15]. Magnetic resonance imaging (MR) is superior to computed tomography (CT) in distinguishing between benign and malignant parotid tumours and high-grade malignancies have ill-defined infiltrating margins post-contrast administration [37].

**REFERENCES**


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Figure 1: A 65-year-old female with a prolonged history of recurrent pleomorphic adenoma presented with slurred speech and deviation of the right angle of mouth. She was subsequently diagnosed with metastatic pleomorphic adenoma to the right supraspinatus muscle.

FINDINGS: Axial contrast-enhanced CT thorax in the venous phase demonstrates: (A) a solid and cystic mass in the right parotid space extending into the right carotid and parapharyngeal spaces; (B) There is bony destruction of the right jugular foramen and occlusion of the jugular bulb, and (C) marked fatty atrophy of the right tongue due to encasement of the right hypoglossal nerve. A hypodense rim-enhancing mass was identified also centered in the right supraspinatus with infiltration into the adjacent scapula and infraspinatus muscle (D).

TECHNIQUE: A-D: Axial contrast-enhanced computed tomography (CT), SIEMENS Emotion 6-slice, slice thickness 2.5 mm, 100 mL Omnipaque 350, 130kV, 120mAs.


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Figure 2: A 65-year old female with a prolonged history of recurrent pleomorphic adenoma presented with slurred speech and deviation of the right angle of mouth. She was subsequently diagnosed with metastatic pleomorphic adenoma to the right supraspinatus muscle.

FINDINGS: (A) Fusion positron emission tomography (PET) and computed tomography (CT) demonstrates a 5 cm hypodense soft tissue mass centred on the supraspinatus muscle with erosive infiltration of the scapula and infraspinatus. The mass demonstrates significant fluorodeoxyglucose (FDG) uptake to a maximum SUV of 5.2. (B) Coronal PET maximum intensity projection demonstrates an isolated 5 cm rounded focal region of increased uptake over the right shoulder concerning for a metastatic deposit. No other sites of abnormal FDG uptake are evident.

TECHNIQUE: GE DISCOVERY ST PET-CT. 362 MBq of FDG was administered. Imaging was acquired at 66 minutes. Axial contrast-enhanced computed tomography (CT) slices were acquired at 3.75 mm thickness (100 mL Omnipaque 350, 140kV, 120 mAs).

Figure 3: A 65-year old female with a prolonged history of recurrent pleomorphic adenoma presented with slurred speech and deviation of the right angle of mouth. She was subsequently diagnosed with metastatic pleomorphic adenoma to the right supraspinatus muscle.

FINDINGS: Fine needle aspiration histology from the right supraspinatus mass demonstrates myoepithelial cells and fibromyxoid stroma consistent with benign pleomorphic adenoma.

TECHNIQUE: Hematoxylin-eosin stain, magnification x 20.
Etiology: MPA is a clinically malignant but histologically benign sub-type of pleomorphic adenoma (PA). It is associated with recurrent or longstanding PA, but etiology is unknown.

Incidence: 52 cases reported in the literature to 2007.

Gender ratio: Unknown. There is a slight female preponderance for PA.

Age predilection: MPA occurs in recurrent or incompletely treated PA. Age predilection for PA is from 3rd to 6th decade.

Risk factors: Recurrent and incompletely treated PA. Known risk factors for PA include a history of neck irradiation.

Treatment: Superficial or total parotidectomy of the local PA recurrence +/- radiotherapy. Metastectomy confers survival benefit.

Prognosis: Prognosis with MPA is poor, with a 50% 5-year mortality.

Imaging findings: MPA usually presents with a local PA recurrence and metastatic disease.

- The most common sites of metastases are lung and bones.
- CT: PA is well-circumscribed with higher attenuation than surrounding parenchyma, with delayed enhancement. Larger PAs can have lobulated contours or infiltrative margins.
- MR: PAs are typically T1 hypointense and T2 hyperintense, with a T2 hypointense rim (fibrous capsule). Signal heterogeneity can vary. Recurrent PA can appear as T2 hyperintense lesions in the operative bed.
- US: PA is a round hypoechoic mass with a distinct often lobulated border. US-guided biopsy is feasible with low risk of seeding.

Table 1: Summary table outlining metastatic pleomorphic adenoma (MPA). CT = computed tomography, MR = magnetic resonance imaging, US = ultrasound.

<table>
<thead>
<tr>
<th>BENIGN:</th>
<th>CT</th>
<th>MR</th>
<th>US</th>
<th>PET</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pleomorphic adenoma (PA)</td>
<td>Smooth, homogeneous, well-circumscribed tumours, with higher attenuation than surrounding parotid parenchyma and delayed contrast enhancement.</td>
<td>Well-circumscribed T1 hypointense and T2 hyperintense mass, often with a T2 hypointense rim representing the fibrous capsule.</td>
<td>Rounded hypoechoic mass with a distinct often lobulated border.</td>
<td>Often FDG-avid and therefore not possible to distinguish from malignant lesions on PET.</td>
</tr>
<tr>
<td>Warthin tumour</td>
<td>Similar CT appearance to pleomorphic adenoma (PA). Small usually &lt; 3 cm ovoid enhancing circumscribed mass with solid and cystic components. More rapid enhancement and washout than a PA. Can be multifocal and bilateral.</td>
<td>Usually T1 hypointense. Cystic areas may be T1 hyperintense due to proteinaceous debris or haemorrhage. T2 signal is variable.</td>
<td>Well-defined ovoid hyperchoeric mass, with anechoic cystic components.</td>
<td>Lower FDG uptake than PA, but still not possible to reliably distinguish from malignant lesions on PET.</td>
</tr>
<tr>
<td>Lymphadenopathy</td>
<td>Multiple intra-parotid enhancing lymph nodes.</td>
<td>Intra-parotid nodes have lower T2 signal intensity than PA.</td>
<td>Multiple hypoechoic hypervascular nodes.</td>
<td>No FDG uptake.</td>
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| MALIGNANT:                   |                                         |                                        |                                         |                                          |
| Mucoepidermoid carcinoma     | Heterogeneously enhancing, often with cystic components and calcification. High grade have infiltrative margins. Intra-parotid lymph nodes may be seen. | Both low- and high-grade are T1 hypointense. Low grade will have T2 hyperintense mucinous cystic areas. High-grade has fewer cystic areas and more infiltrative margins. | Predominantly hypoechoic lesion with anechoic areas. | Marked FDG uptake. |
| Adenoid cystic carcinoma     | Low grade is typically a homogenously enhancing well-circumscribed mass, difficult to distinguish from PA. High grade will have infiltrative margins. | Predominantly T1 hypointense but there may be high T1 signal areas of haemorrhagic necrosis. High-grade are typically T2 hypointense and infiltrative, with perineural spread to CN7. | Not characteristic. MR is recommended. | Marked FDG uptake. |

Table 2: Summary table outlining benign and malignant differential diagnoses of a parotid pleomorphic adenoma (PA).
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ABBREVIATIONS

CEPA = Carcinoma ex-pleomorphic adenoma
CT =Computed tomography
FDG = Fluoro-deoxyglucose
MPA = Metastatic pleomorphic adenoma
MR = Magnetic resonance imaging
PA = Pleomorphic adenoma
PET = Positron-emission tomography
US = Ultrasound

KEYWORDS

Pleomorphic adenoma; metastatic; rotator cuff metastasis; parotid tumours

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