Purpose: Inflammatory myofibroblastic tumours (IMT) are a subcategory of inflammatory pseudo-tumours (IPT). They arise most commonly in the abdominopelvic region, lung and retroperitoneum, but virtually any anatomical site may be involved. Predominantly children and adolescents are affected and there is a tendency for local recurrence. In the literature up to the present, 20 patients have been reported with an IPT/IMT of the breast. We would like to present another patient with this unusual tumour entity of the breast and discuss the literature.

Patient and Examinations: A 23-year-old woman presented with a painless lump in her left breast. There was no history of breast cancer in her family. Sonography showed a hypoechoic heterogeneous solid mass with irregular margins. A core needle biopsy revealed a tumour of high cellularity and a densely collagenous background. Immunohistochemically, the spindle-shaped cells were immunoreactive to smooth muscle actin and ALK-1 protein. Additional FISH analysis proved ALK rearrangements on chromosome 2p23 leading to the diagnosis of an IMT. Wide surgical excision was performed with no evidence of local recurrence after 12 months.

Conclusion: Three of the above mentioned 20 patients with IMT/IPT of the breast developed a recurrent tumour, none presented with distant metastasis. A significant recurrence rate of 15% leads to a clinically and sonographically close follow-up in these patients.

Zusammenfassung


Patientin und Untersuchungsbefunde: Eine 23-jährige Patientin mit inflammatorischem myofibroblastischen Tumor der linken Mamma – Fallbeschreibung und Literaturübersicht

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Key words

- breast tumor
- inflammatory myofibroblastic tumor
- ALK gene

Schlüsselwörter

- Brusttumor
- inflammatorischer myofibroblastischer Tumor (IMT)
- ALK-Gen

received 18.9.2013
revised 1.12.2013
accepted 3.12.2013

Bibliography

DOI http://dx.doi.org/10.1055/s-0033-1360185
Geburtsh Frauenheilk 2014; 74: 167–170 © Georg Thieme Verlag KG Stuttgart · New York · ISSN 0016-5751

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23-Year-Old Female with an Inflammatory Myofibroblastic Tumour of the Breast: A Case Report and a Review of the Literature

23-jährige Patientin mit inflammatorischem myofibroblastischen Tumor der linken Mamma – Fallbeschreibung und Literaturübersicht

23-Year-Old Female with an Inflammatory Myofibroblastic Tumour of the Breast: A Case Report and a Review of the Literature
Introduction

Inflammatory myofibroblastic tumours (IMT) are inflammatory pseudotumours (IPT). The latter is a generic term to describe various neoplastic and non-neoplastic entities that share a common histological appearance. Cytologically, this consists of bland spindle cell proliferation with a prominent, usually chronic inflammatory infiltrate [1]. Over the last two decades, IMT has emerged as a distinctive neoplasm of intermediate biological potential [2]. They arise most commonly in the abdominopelvic region, lung and retroperitoneum, but virtually any anatomical location can be involved. Accompanying symptoms can be fever, weight loss and pain as well as anaemia, thrombocytosis and polyclonal hypergammaglobulinaemia. Predominantly children and adolescents are affected but it may appear in either sex at any age. Local recurrence rate varies between <2 and 25% depending on whether the tumour is confined to the lung or an extrapulmonary lesion. Up to the present time 20 patients have been described with an IMT/IPT of the breast (Table 1). All these cases occurred spontaneously without any apparent prior injury. The first case of a posttraumatic IPT has recently been reported by Vecchio et al. in a 22-year-old male [3]. We present another female patient with this unusual tumour entity of the breast and discuss the unique appearance and clinical behaviour of IMTs among the group of spindle-cell breast lesions.

Patient and Examinations

A 23-year-old healthy female was examined because of a painless lump in her left upper breast. There was no history of breast cancer in her family. Physical examination revealed a solitary, hard, non-mobile mass measuring approximately 2.0 cm in size. Lymph nodes were not palpable in the axilla. Sonography showed a hypoechoic heterogeneous lesion of 2.0 cm in diameter with irregular margins and indistinct acoustic shadowing (Fig. 1). Power Doppler was negative. Due to the young age of the patient and dense breast tissue we added contrast-enhanced dynamic MR-mammography. MRI confirmed a strong contrast enhancing mass in the left breast corresponding to the ultrasound findings without demonstrating any further lesions (Fig. 2). Both imaging modalities were classified as BIRADS IV and a core needle biopsy was performed. Microscopy showed a well-defined tumour with high cellularity. Histology showed aside from spindle cells that the lesion was composed of a dense mixed inflammatory cell infiltrate of plasma cells and lymphocytes, little cellular atypia and rare mitoses. Immunohistochemistry yielded strong smooth-muscle actin (SMA) reactivity within the spindle cells with a slight heterogeneous pattern of distribution (Fig. 3). Other than that the tumour was composed of macrophages intensely expressing CD14.

Table 1  IP/IMT in the literature compared to our case; NA, not available.

<table>
<thead>
<tr>
<th>No. of cases and reference</th>
<th>Age/gender</th>
<th>IP/IMT</th>
<th>ALK positive/negative</th>
<th>Follow-up</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>16 [4]</td>
<td>range 13–86 years, female</td>
<td>7/9</td>
<td>5 cases –</td>
<td>maximum 9 years</td>
<td>3 recurrences</td>
</tr>
<tr>
<td>1 [5]</td>
<td>60, female</td>
<td>IMT</td>
<td>–</td>
<td>24 months</td>
<td>no recurrence</td>
</tr>
<tr>
<td>1 [3]</td>
<td>22, male</td>
<td>IP</td>
<td>–</td>
<td>NA</td>
<td>no recurrence</td>
</tr>
<tr>
<td>1 [7]</td>
<td>46, female</td>
<td>IMT</td>
<td>+</td>
<td>5 years</td>
<td>no recurrence</td>
</tr>
<tr>
<td>Present case</td>
<td>23, female</td>
<td>IMT</td>
<td>+</td>
<td>12 months</td>
<td>no recurrence</td>
</tr>
</tbody>
</table>

Fig. 1  Sonogramm shows a 2 cm, irregular shaped lesion with indistinct dorsal echoing.

Fig. 2  Subtraction image of T1-weighted contrast-enhanced breast MRI in transverse orientation demonstrating the single nodule in the left upper breast with strong contrast medium enhancement and a central necrosis.
and CD68 as well as CD3-positive T-lymphocytes with an admix-
ture of scattered CD-20 positive B-lymphocytes. Immunohisto-
chemistry for the pan-cytokeratin markers AE1/3 and E-Cadher-
in was negative. S100-staining displayed several dendritic cells.
Furthermore the ALK protein (anaplastic lymphoma kinase) was
moderately positive on staining. FISH analysis using a dual colour
probe specific for the ALK gene found a split of the red/green sig-
nals, demonstrating rearrangement of this gene on chromosome
2p23 (Fig. 4). Altogether the findings were consistent with the
diagnosis of a myofibroblastic tumour belonging to the category
B3 lesions.
The mass was entirely excised with free margins of at least
10 mm after repeated surgery because of too narrow margins.
Macroscopically the nodule was firm, circumscribed and yellow
on cut section with a central necrosis (Fig. 5).
A survey for distant metastasis including chest x-ray and sonog-
raphy of the liver showed normal results. The postoperative
course was uneventful and the 12-months follow-up examina-
tion exhibited no evidence of local recurrence.

Discussion

Inflammatory myofibroblastic tumours of the breast are a very
rare condition, which clinically and on imaging easily mimic ma-
lignancy [8]. Recommended treatment is complete surgical exci-
sion as recurrence rates of up to 25% are experienced [1]. The
World Health Organization continues to classify inflammatory
myofibroblastic tumours as a distinct borderline lesion. Still it
has not been conclusively decided whether it is reactive or neo-
plastic in nature. Remarkably, about 50% of IMTs are positive for
rearrangements involving the ALK gene proven by FISH analysis.
This property favours the idea of a neoplastic nature of this le-
sion. Clonal abnormalities of ALK were first described in anaplas-
tic large cell lymphoma (ALCL), which is a true neoplasm [9]. In
ALCL patients ALK-positivity is accompanied by a less aggressive
clinical course [10, 11]. Only recently, the first 46-year-old wom-
an with an ALK overexpressing IMT of the breast was reported of
[7]. She was free of recurrence five years after diagnosis. Whether
there is a more favourable outcome of patients with an ALK-pos-
itive IMT is not known yet. Although studies suggest a very low
risk for metastasis, its influence on recurrence remains question-
able [12, 13]. Larger numbers of patients with an ALK positive
IMT of the breast are needed to judge its impact on metastasis
and recurrence. The translocation of the ALK gene leads to a con-
stitutive tyrosine kinase activation. Similar to ALCL, ALK expres-
sion is more common in younger patients with IMT, but is not
confined to this population. Distant metastasis of IMT is rare, oc-
curring in <5% of cases. So far, no patient with an IMT/IPT of the
breast has developed distant metastases. But three (15%) of the
20 reported cases showed local recurrence during follow-up ex-
aminations. Time until recurrence varied between three months
and nine years. This fact emphasises the need for a close follow-
up in these patients. We recommend sonography every six
months as the nodule was well depicted on ultrasound and re-
currence rarely developed within months. Mammography is not
appropriate at this young age. MR-mammography on a regular basis seems overdone.

In summary, according to the literature IMT of the breast is adequately treated with wide local excision. There is no need for radiation and chemotherapy. A survey to exclude distant metastasis is advisable.

**Conflict of Interest**

None.

**References**