Case Report

Simultaneous proximal-type epithelioid sarcoma of the chest wall and primary pulmonary adenocarcinoma: a case report and literature review

Yanhai Wang¹, Limei Xie¹, Zhe Wang², Quanwei Zhang³, Yong Wang⁴, Jing Wang⁵

Departments of ¹Ultrasound, ²Pathology, Shengjing Hospital of China Medical University, Shenyang, China; Departments of ³Surgery, ⁴Radiology, ⁵Ultrasound, Liaodong Branch Hospital, Shengjing Hospital of China Medical University, Panjin, China

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Abstract: Epithelioid sarcoma (ES) is a rare and aggressive soft-tissue neoplasm that is divided into distal-type ES and proximal-type ES. Due to this tumor's infrequent occurrence, uncertain differentiation, and atypical histological pictures, diagnosis of epithelioid sarcoma in its early stages has become extremely difficult. This present study reports, in detail, a rare case of proximal-type ES of the chest wall with primary pulmonary adenocarcinoma, simultaneously, in a 54-year-old man. To the best of our knowledge, this is the first case of simultaneous proximal-type epithelioid sarcoma of the chest wall with primary pulmonary adenocarcinoma.

Keywords: Epithelioid sarcoma, proximal-type epithelioid sarcomas, chest wall, primary pulmonary adenocarcinoma, double cancer

Introduction

Epithelioid sarcoma (ES), first described by Enzinger in 1970, is a rare aggressive soft-tissue neoplasm [1]. ES is divided into two recognizable clinicopathological entities, classic ES (also called distal-type ES) and proximal-type ES (PES). Due to this tumor’s infrequent occurrence, uncertain differentiation, and atypical histological pictures, diagnosis of epithelioid sarcoma in its early stages has become extremely difficult. In this study, we report a rare case of PES of the chest wall with primary pulmonary adenocarcinoma, simultaneously, in a 54-year-old man. This report includes clinical symptoms, physical signs, ultrasonic and CT findings, pathological features, treatment, and a review of the published literature. A current review of English-language literature showed no more than 10 cases PES of the chest wall, from 1972 to 2016, according to PubMed [2-4]. Among these papers, none of them had detailed documentation. To the best of our knowledge, this present study is the first case of simultaneous proximal-type epithelioid sarcoma of the chest wall with primary pulmonary adenocarcinoma.

Case presentation

A 54-year-old man, with no previous medical history, was admitted, due to a rapidly increasing mass on the left anterior chest wall, with thoracodynia, cough, and fever. Within a month, the mass developed in a rapid manner, from approximately 4 cm in diameter to 10 cm. He experienced about 10-kilograms of weight loss. Five days before admission, he suffered from a high fever. He was a farmer with 30 years of smoking history. He had no significant history of asbestos exposure or radiation exposure. No personal or family history of cancer was recorded. Upon physical examination, a large tender mass with poor mobility was palpated in the deep of his left nipple (Figure 1). A 10.1 × 7.8 × 5.6 cm solid irregular heterogeneously hypoechoic mass, with discontinuous calcifications under the nipple, was shown during ultrasonic examination. Centric blood flow was seen within the mass (Figure 2A, 2B). Thoracic computed tomography (CT) demonstrated a 7.9 × 5.0 cm heterogeneous smooth soft-tissue mass that had directly invaded the fifth anterior rib. Volume rendering technique (VRT) showed that the fifth rib of the left side was deficient (Figure 3A,
A nodule 1 cm in diameter was found in the dorsal segment of the left lower lobe (Figure 4). The patient underwent a biopsy of the mass, guided by ultrasound, which demonstrated a poorly differentiated carcinoma, perhaps breast cancer. A rare malignant tumor, breast cancer with lung metastasis was suspected. Patient underwent a left “breast cancer” radical resection, left chest wall reconstruction, dissection of the left axillary lymph node, and resection of left lower lobe of the lung. The resected mass of “breast cancer” was 10 × 5 × 5 cm in volume and solid, but very soft. It was yellow-white in color, with focal necrosis (Figure 5). Unexpectedly, the pathology of the chest wall mass was not breast cancer at all. It was composed of...
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round large volume cells with cytological atypia, part of the nucleoli at the bottom and part at the top. This was not typical for any kind of cancer previously seen (Figure 6). While the pathology of left pulmonary nodules was primary adenocarcinoma, it was quite different from that of the chest wall mass (Figure 7). According to histological characteristics, an accurate diagnosis could not be made. Thus, immunohistochemistry was tested to ensure accurate diagnosis. We confirmed positive immunohistochemical stains for calretinin, cytokeratins (CKs), epithelial membrane antigen (EMA), and Vimentin. Stains were negative for thyroid transcription factor-1 (TTF-1), Napsin, CD68, CD30, CD34, CD56, MelanA, S-100, SMA, Myogenin, HMB-45, SALL4, CD117, CK20, MPO, LCA, and ALK (Figure 8A-C). Our final diagnosis was double cancer, proximal-type epithelioid sarcoma of the chest wall and primary pulmonary adenocarcinoma. No chemotherapy or radiation therapy was performed. Two months after the operation, patient recovered rapidly and no discomfort was indicated. Unfortunately, four months after the operation, he felt a headache and right limb hemiplegia. CT of the head showed that the left hemisphere was occupied by a low density area, perhaps metastasis (Figure 9). Six months after surgery, he died due to hernia cerebri.

Discussion

Proximal-type epithelioid sarcomas were first described in 1997 by Guillou L [5]. Unlike classic-type epithelioid sarcomas, PES usually arises more proximally, that is, in the chest wall, inguinal region, thighs, perineum, etc. Although CT or MR findings are nonspecific, PES usually shows a multinodular configuration and admixes with regional lymph node metastasis [2]. PES shows a multinodular pattern of growth, consisting of large epithelioid cells with marked cytological atypia, vesicular nuclei, and prominent nucleoli [1]. Diagnosis of ES can be suggested based on its cytomorphology and immunohistochemistry [6, 7]. Surgical resection is still the main form of treatment. Surgery with a wide margin is a good and safe technique for treatment of primary chest wall tumors, with acceptable morbidity and mortality [8]. A multidisciplinary approach may be crucial for optimal management of large soft tissue sarcomas [9]. Prognosis for PES is poor and distant metastasis eventually occurs in up to 60% of cases [10, 11].
This study presented a rare case of PES, coexisting with primary adenocarcinoma. This case was extremely rare, not only because of the rare PES, but also because of the coalescence of two histologically different cancers. To the best of our knowledge, this was the first reported case of PES in the chest wall coexisting with adenocarcinoma of the lung. Initially, it was thought to be tuberculosis. According to lung CT and biopsy of the mass, rare male breast cancer with pulmonary metastasis was considered. After the operation, however, a reasonable diagnosis could not be made, according to cytomorphology. We had to test immunohistochemistry in order to ensure a true diagnosis.

There are very few papers documenting, in detail, ultrasonic features of PES. In our case, an ultrasound of PES of the chest wall showed a large heterogeneously hypoechoic mass with an irregular shape and central internal color Doppler, very similar to breast cancer. Fractures of ribs may be seen as discontinuous calcification within the mass. CT and Volume rendering technique (VRT) offered a fast 3-dimensional view of the mass, its surrounding structures, and degree of invasion of periphery tissues.

PES should be differential from tuberculosis of the chest wall, breast cancer, and other poorly differentiated soft tissue carcinomas. Radiographic and CT scan features of rib tuberculosis have been described in the literature as rib erosions. Destruction with adjacent abscess and use of US contributes to assess rib destruction and associated soft tissue abscesses [12]. Tuberculosis of the chest wall maybe show abscesses as hypoechoic areas, with varying degrees of internal heterogeneity [12]. PES may declare inhomogeneous hypoechoic without abscess, perhaps contributing to differential diagnosis. Breast cancer usually appears to be an irregular hypoechoic mass with penetrating arteries, with or without calcification, very similar to PES. Breast cancer, however, is commonly seen in females and tumors most likely originate from mammary glands rather than the chest wall. The above is helpful in differentiating between breast cancer and PES. It remains difficult to diagnosis if the mass is large in vol-

Figure 8. A. Malignant cells expressing CK; B. Malignant cells expressing EMA; C. Malignant cells expressing vimentin.

Figure 9. The left hemisphere was occupied by low density area.
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...volume, with surrounding tissue invasion, or in male patients. Malignant pleural mesothelioma (MPM) is a relatively common tumor of the pleura. Pleural effusion, pleural thickening, ipsilateral volume loss, local invasion, lymphadenopathy, and metastatic disease are the most common imaging manifestations of MPM. Asbestos-related pleural disease may also be recorded. Although individual imaging findings may not be specific, presence of one or more of these features should raise suspicion for diagnosis of MPM [13]. Because of the limited number of cases, more research is necessary.

ES is characterized by nodular aggregates of epithelioid cells, which are immunoreactive to cytokeratins (CKs) and epithelial membrane antigens (EMA). PES is composed of sheets of larger and more atypical cells with variable rhabdoid morphology. The histological similarity of ES to a wide range of non-neoplastic and neoplastic lesions creates a special diagnostic challenge for cytopathologists [7]. This current reported case was characterized by atypically round large volume cells, with obvious nucleoli, not typical for any kind of cancer. A correct diagnosis was not made until immunohistochemistry was tested.

Another characteristic of this present study case was that it involved double cancer. To the best of our knowledge, there have been no other reports of simultaneous epithelioid sarcomas of the chest wall and pulmonary adenocarcinoma. Double cancer was not known until the pathology was examined. It is unknown whether there is some relationship between them.

In summary, this study presented a rare case of simultaneous PES with primary pulmonary adenocarcinoma. PES of the chest wall may be manifested as a “breast cancer” in ultrasound and radiology. PES should be considered when diagnosis of chest wall tumors is difficult, according to conventional diagnostic methods. Immunohistochemistry can be helpful in diagnosis and differential diagnosis.

Disclosure of conflict of interest

None.

Address correspondence to: Limei Xie, Ultrasound Department, Shengjing Hospital of China Medical University, 36 Sanhao Street, Heping District, Shenyang 110004, Liaoning Province, China. Tel: +86 18940251927; E-mail: xielm72@163.com

References