

中文題目：產後婦女在肺部出現罕見的肺發炎性肌纖維母細胞腫瘤 - 和懷孕相關嗎？

英文題目：A Rare Inflammatory Myofibroblastic Tumor in a Postpartum Woman : Is It Associated with Pregnancy ?

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Introdcution

Inflammatory myofibroblastic tumors (IMTs) are considered a subset of inflammatory pseudo -tumors. Their typically features are spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils. Here, we report a case of inflammatory myofibroblastic tumor of the lung with presentation of progressive dyspnea and bilateral edema in a young pregnant woman who just underwent normal spontaneous delivery 3 days ago.

Case presentation

The 26-year-old female with no known systemic disease and she had normal spontaneous delivery. There was no significant finding through her antenatal examination, delivery process, and there was no postpartum hemorrhage .

However, 3 days after delivery, she presented to our emergency department with progressive dyspnea, paroxysmal nocturnal dyspnea, orthopnea, chest tightness, progressive bilateral lower limbs edema, and facial swelling.

Chest radiography revealed a round 5.4 cm solid nodule over the left upper lung field. Serology revealed elevated C reactive protein , B-type natriuretic peptide, and D-dimer and decreased hemoglobin. In order to exclude pulmonary embolism related respiratory distress, chest computed tomography was arranged and disclosed a round ,well-defined and homogenous mass in the left upper lobe of the lung. She was admitted to the chest ward for treatment.

CT-guided biopsy showed an inflammatory myofibroblastic tumor (IMT) and thoracoscopic segmentectomy of lung was performed ,microscopic examination disclosed a neoplasm composed of spindled cells mixed with inflammatory cells, including plasma cells, lymphocytes, giant cells and histiocytes. These cells are also immunoreactive for Anaplastic lymphoma kinase (ALK). The finding is compatible with IMT. After completely resection of the tumor, she had an uneventful recovery and received a regular follow-up now.

Discussion

IMT used to be considered to be a subtype of Inflammatory pseudotumor (IPT), histiocytoma, fibrous histiocytoma, xanthoma, xanthofibroma, xantogranuloma and plasma cell granuloma.

In adults, IMT accounts for 1% of lung tumors in adults. The etiology and mechanism of pulmonary/ extrapulmonary IMT genesis is still unclear. Inflammation, trauma might be part

of the pathogenesis. Viral infections like Epstein-Barr virus and human herpesvirus-8 are also considered to cause, but strong evidence is required.

IMTs are usually benign or slow-growing tumors without local invasion or distant metastases, however, 5-10% of the patients present aggressive locally invasion or metastatic disease. Overall, IMTs have good prognosis with a 5-year survival rate of 74% to 91%. IMT is characterized by spindle-shaped tumor cells in a background of infiltration with lymphocytes and plasma cells. Lungs are the most common site of IMTs, although they can occur at any site of the body.

In addition, 50% of IMTs are ALK rearrangement-harboring tumor, and positive for ALK rearrangement is also considered for better prognosis. Surgery is still the best option of treatment for IMT. For some metastatic or aggressive IMT patients harboured ALK translocation, crizotinib, an ALK inhibitor, has been proved to treat successfully. In contrast, ALK negative IMT, the prognosis is poor.

The presented case was noted to have a huge IMT in postpartum. Generally speaking, pregnant woman always refused to take a chest x-ray or chest CT though some respiratory symptoms occurred. The phenomenon may delay the diagnosis of lung tumors, especially cancers.

In the report, all pregnancy-related IMTs all were strongly positive for progesterone receptor (PR) and variably positive for estrogen receptor (ER). Given that progesterone is a dominant regulatory hormone during pregnancy, it is not surprising that pregnancy associated IMTs express PRs, which are also expressed by and regulate the physiology and transformation of the decidua during pregnancy and parturition.

Conclusion

Pregnancy-associated IMTs are more common in the uterus, placenta, or urinary bladder and this is the third case report of pulmonary IMT associated with pregnancy in a pubmed search. Surgery is still the best option of treatment for IMT. For some metastatic or aggressive IMT patients harboured ALK translocation, crizotinib, an ALK inhibitor, has been proved to treat successfully.