Inflammatory myofibroblastic tumor: difficult to manage

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Introduction

Inflammatory myofibroblastic tumor (IMT) is considered a neoplastic reaction to an inflammatory insult (1). As a rare condition but masquerading as a number of common presentations, this inflammatory tumor deserves particular attention. IMT also causes dilemmas in diagnosis and management. Based on mainly small case series and scattered case reports, establishing a guide to diagnosis and management is difficult. In view of variability of its nature and complexity of information from the literature, a general consideration and approach to IMTs is discussed in this article.

Difficulty: diagnosis

As a rare tumor with incidence not more than 0.1% (2), and more common in the young population especially the paediatric patients (3), the presence of a lung tumor in a young patient seldom raises a suspicion of either lung malignancy or this ‘pseudotumor’. Other differential diagnoses, e.g., hamartoma, haemangioma or chondroma may be considered well-above an IMT. Clinically the presentation may be similar to a respiratory tract infection (e.g., cough, fever, pleuritic chest pain, dyspnea etc.) or asthma (4-6), or appear like malignancy-associated constitutional symptoms (e.g., weight loss, fatigue) (7). Association with a number of conditions (such as bacterial or viral infections, Sjogren syndrome, lymphoma, IgG4 syndrome, post-stem cell or solid organ transplantation etc.) seldom help raise the suspicion of IMTs, and the causal links are far from established (8-10). Radiologically when it presents as a solitary pulmonary nodule, it shows a solitary, sharply circumscribed and lobulated mass. Pleural effusion may occasionally be an accompanying feature (3). When developed endobronchially, atelectasis or obstructive pneumonia ensue. These findings add no distinguishing values to it from other types of lung tumors. The appearance of IMTs on computer tomography (CT) scans are variable and non-specific, and the addition of positron-emission tomography makes the picture even more perplexing, as IMTs demonstrate variably intense 18-FDG uptake depending on its pathological properties (e.g., cellularity, proliferative index, and the amount of plasma cell infiltrates) (11). Therefore, based on radiological features, surgeons can neither propose nor oppose the possibility of malignancy. There is no evidence to suggest that bronchoscopic or percutaneous needle biopsy can confidently diagnose or exclude malignancy (12). The appearance of spindle cells in the tiny specimen usually does not point towards IMTs unless a representative sampling and analysis including CD68 and vimentin can be performed (13). Therefore, confirmation of diagnosis mostly required excision of the tumor en bloc with the lobe in which it is located.

Difficulty: treatment

IMT has the potential to undergo malignant transformation or metastasize (13-15). Surgical resection with a view to both diagnosis and treatment should be the mainstay of management. However, major lung resection (which may be to the extent of lobectomy or even pneumonectomy) for a potentially benign ‘spindle cell’ tumor in young patients must be justified in times of uncertain preoperative diagnosis. Firstly, CT scan of the thorax and abdomen
must be performed to exclude synchronous inflammatory tumors in other organs (16). Secondly, complete resection should be achieved to maximize post-operative survival (17), which could be more than 91% (13). Recurrence may be up to 8% if resection is partial without adjuvant therapy (14). Pre-operative embolization of the feeding artery to the IMT may reduce hypervascularity and facilitate complete resection (18). Still, contralateral recurrence after complete resection by pneumonectomy had been reported (19). Endobronchial IMTs can be removed under rigid bronchoscopy (20). Close surveillance is necessary for any persistence of the lesion (21).

There is no medical therapy indicated as primary therapy for IMTs. Scattered reports on the effectiveness of Celecoxib, a non-steroidal anti-inflammatory drug (NSAID), were available (22,23). Celecoxib inhibits cyclooxygenase-2 enzyme and vascular endothelial growth factors which are essential for angiogenesis. Regression of an IMT after a ‘neoadjuvant’ NSAIDs therapy may spare a pneumonectomy (22). Combination of Celecoxib with chemotherapeutic agent was reported to induce durable remission (24). It is known that IMTs also show response to corticosteroid therapy (25,26). However, this response may be attributed to the response of IgG4-related diseases to steroids, while not all IMTs are related to IgG4 disease. On the other hand, steroids had also been reported to aggravate disease progression of IMTs (27). IMTs may exhibit anaplastic lymphoma kinase (ALK) mutation in 40–50% cases (28,29). The use of ALK receptor tyrosine kinase inhibitors (e.g., Crizotinib, Ceritinib, and Alectinib) can be effective (30-33), and could be considered for inoperable cases.

Conclusions
A general summary of the difficulties in management of inflammatory fibroblastic tumor is discussed. Limited evidence from the literatures signifies the need of gathering and reporting more experience to the pool. Adequate suspicion of this condition and thorough discussion with patients regarding subsequent treatment are necessary. Optimal survival requires careful work-up and planning, and complete resection for surgical candidates.

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Footnote
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