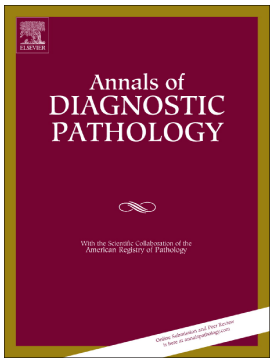

Extrapleural solitary fibrous tumor: A distinct entity from pleural solitary fibrous tumor. An update on clinical, molecular and diagnostic features

Andrea Ronchi, Immacolata Cozzolino, Federica Zito Marino, Marina Accardo, Marco Montella, Iacopo Panarese, Giuseppe Roccuzzo, Giorgio Toni, Renato Franco, Annarosaria De Chiara

PII: S1092-9134(17)30309-X
DOI: doi:10.1016/j.anndiagpath.2018.01.004
Reference: YADPA 51226

To appear in:

Please cite this article as: Andrea Ronchi, Immacolata Cozzolino, Federica Zito Marino, Marina Accardo, Marco Montella, Iacopo Panarese, Giuseppe Roccuzzo, Giorgio Toni, Renato Franco, Annarosaria De Chiara , Extrapleural solitary fibrous tumor: A distinct entity from pleural solitary fibrous tumor. An update on clinical, molecular and diagnostic features. The address for the corresponding author was captured as affiliation for all authors. Please check if appropriate. Yadpa(2017), doi:10.1016/j.anndiagpath.2018.01.004

Title: Extrapleural solitary fibrous tumor: a distinct entity from pleural solitary fibrous tumor. An update on clinical, molecular and diagnostic features.

Authors: Andrea Ronchi [a], Immacolata Cozzolino [a], Federica Zito Marino [a], Marina Accardo [a], Marco Montella [a], Iacopo Panarese [a], Giuseppe Roccuzzo [a], Giorgio Toni[a], Renato Franco [a], Annarosaria De Chiara [b]

Affiliations: [a] Pathology Unit, Department of Mental and Physical Healt and Preventive Medicine, Università degli Studi della Campania "Luigi Vanvitelli", via Luciano Armanni 20, 80100, Naples, Italy. [b] Pathology Unit, Department in Support of Oncology Paths, Diagnostic Area, Istituto Nazionale dei Tumori I.R.C.C.S. Fondazione "Pascale", via Mariano Semmola 52, 80131, Naples, Italy.

Corresponding Author:
Renato Franco
Pathology Unit, Department of Mental and Physical Healt and Preventive Medicine, Università degli Studi della Campania "Luigi Vanvitelli"
Via Luciano Armanni 20, 80100 Napoli, Italy
mail: renato.franco@unicampania.it
phone: +390815664000

Abstract:

Solitary fibrous tumor is a mesenchymal neoplasm originally described in the pleura, but thereafter reported in several anatomic sites. Although the etiology of the neoplasm remains largely unknown, the pathogenesis seems related to NAB2-STAT6 fusion gene due to paracentric inversion on chromosome 12q13. The diagnosis of extrapleural solitary fibrous tumor may be challenging, due to its rarity, and requires an integrated approach including specific clinical, histological, immunohistochemical and even molecular findings. Histologically, extrapleural solitary fibrous tumor shares the same morphological features of the pleural cases, since it is characterized by a patternless distribution of both oval and spindle shaped cells in a variable collagen stroma. In addition, morphological variants of mixoid, fat-forming and giant cells rich tumors are described. A correct diagnosis is mandatory for a proper therapy and management of the extrapleural solitary fibrous tumor patients, which is usually more aggressive when compared to pleural forms, particularly cases occurring in mediastinum, retroperitoneum, pelvis and meninges. Although solitary fibrous tumor is usually considered as a clinically indolent neoplasm, the prognosis is substantially unpredictable and only partially related to morphological features. In this context cellularity, neoplastic borders, cellular atypias and mitotic activity can show a wide range of variability. We review extrapleural solitary fibrous tumor, discussing diagnostic clues, differential diagnosis, recent molecular findings and prognostic factors.