Shroque Zaher, J Clin Exp Pathol 2017, 7:5 (Suppl)
DOI: 10.4172/2161-0681-C1-040

Primary pulmonary myxoid sarcoma: Rare entity

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This case represents a rare entity-primary pulmonary myxoid sarcoma, of which to the best of our knowledge only 10 other cases have been reported in the literature. They are defined by distinctive histo-morphological features and characterized by a recurrent fusion gene. All tumors involved pulmonary parenchyma with a predilection for the endobronchial component. They appear to have a predilection for females, with 7 of the 10 reported cases, occurring in women. Microscopically, they are lobulated tumors comprising cords of polygonal, spindle, stellate cells with myxoid stroma, morphologically reminiscent of extra-skeletal myxoid chondro-sarcoma. Tumors were immune-reactive for only vimentin and weakly focal for EMA, although our specific case was negative for these markers. In 7 of the 10 tumors, a specific *EWSR1-CREB1* fusion gene was demonstrated by reverse transcription-polymerase chain reaction. This gene fusion has been described previously in 2 histologically and behaviorally different sarcomas: Clear cell like sarcoma-like tumors of the gastrointestinal tract and angiomatoid fibrous histiocytoma; however, this is a novel finding in tumors with the morphology described and occurring in the pulmonary region.

Biography

Shroque Zaher is a Consultant Histopathologist with a specialist interest in hematopathology, pulmonary pathology and medical education. She has completed her Pathology training at the London and East of England Deaneries and gained her CCT in 2016. She has obtained her FRCP from the Royal College of Pathologists, United Kingdom in 2015.

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