A rare case of primary pulmonary myxoid sarcoma

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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 histomorphological 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 within myxoid stroma, morphologically reminiscent of extraskeletal myxoid chondrosarcoma. Tumors were immunoreactive 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 sarcoma like tumors of the gastrointestinal tract and angiomatoid fibrous histiocytomas; however this is a novel finding in tumors with the morphology described and occurring in the pulmonary region.

Biography

Shroque Zaher has completed her Pathology training and has been awarded her CCT. She has gained her FRCPath from the Royal College of Pathologists, United Kingdom in 2015. Her main interests are lung and hematopathology as well as medical education.

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