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Pathology Congress 2017: Primary pulmonary myxoid sarcoma: Rare entity - Shroque Zaher Consultant Histopathologist – UAE.

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This case represents a rare primary myxoid pulmonary sarcoma, of which, to our knowledge, only 10 other cases have been reported in the literature. They are defined by distinctive histo-morphological characteristics and characterized by a recurrent fusion gene. All the tumors involved a pulmonary parenchyma with a predilection for the endobronchial component. They seem to have a predilection for women, 7 of the 10 reported cases occurring in women. Microscopically, they are lobulated tumors comprising cords of polygonal cells, fusiform and star-shaped with a myxoid stroma, morphologically reminiscent of extra-skeletal myxoid chondrosarcoma. The tumors were immunoreactivity only for vimentin and weakly focal for EMA, although our specific case was negative for these markers. In 7 of 10 tumors, a specific EWSR1-CREB1 fusion gene was detected by reverse transcription polymerase chain reaction. This gene fusion has been designated formerly in 2 histologically and behaviorally different sarcomas: tumors of the gastrointestinal type and angiomatoid fibrous histiocytomas resembling sarcomas; however, this is a new finding in tumors with the morphology described and occurring in the pulmonary region.

Primary pulmonary sarcomas remain infrequent diseases unlike lung carcinomas. The occurrence of these sarcomas is between 0.013 and 0.40% of all malignant lung tumors. There are malignant mesenchymal tumors. They flow from the soft tissue of the lung. Pulmonary sarcomas are a heterogeneous group with various biological behaviors. Their morphological structure does not stand out from soft tissue sarcomas. Primary pulmonary sarcomas happen frequently in childhood and in young people, different lung carcinomas. Radiation and certain toxic substances are noted risk factors. Certain gene mutations, infectious pathogens and contraception have a possible impact on the origin of certain types of sarcomas. The current assumption is that most sarcomas, if not all sarcomas, originate from primitive multipotent mesenchymal cells by malignant transformation into one or more lines. The diagnostic standard is the biopsy of a tumor with histological and immune histochemical examination of a sample. The elementary investigative problem is the prohibiting of a secondary origin of sarcomatic cells in the lung, since pulmonary metastases from extra-pulmonary sarcomas are more often than primary pulmonary involvement. The optimal treatment is resection of the tumor. The other therapeutic modalities are radiotherapy and chemotherapy, but the results of these modalities are not satisfactory.

Primary pulmonary sarcomas can have aggressive progression. In our case, as in metastatic or advanced sarcomas, chemotherapy with doxorubicin and ifosfamid is the only active treatment, but the response is less than 20%. High-dose chemotherapy is feasible and provides attractive response rates in patients with soft tissue sarcomas, but cannot yet be considered standard practice. In localized disease, surgical removal is the treatment of choice for all histological types, followed by radiation therapy if the removal is incomplete. A thorough clinical examination, followed by a complete CT scan, was performed to exclude primary synovial sarcoma located at the periphery and distant metastases. Therefore, surgical excision was planned after three cycles of chemotherapy, but the patient died.

We describe a patient with primary fusiform sarcoma of the lung whose tumor could not be observed and who died after two cycles of chemotherapy. Because lung sarcoma is rare, data regarding its natural history and published cases are limited. Further investigation and data collection are necessary to optimize the treatment of this group of rare and aggressive tumors.

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The most important differential diagnosis of primary pulmonary sarcoma is the metastatic spread of additional pulmonary sarcoma. It is therefore necessary to obtain a detailed clinical history and to initiate appropriate investigations to treat this possibility. Our patient had no signs of present or past soft tissue neoplasms. Our patient had no history of radiation exposure, which was recognized as a predisposing factor for the development of sarcomas. Other differential diagnoses include bronchogenic carcinoma and malignant melanoma. Immunohistochemistry has an important role in classification. A panel of antibodies is required to properly classify soft tissue sarcomas. Immunohistochemistry is also essential for eliminating the much more common sarcomatoid lung carcinomas. In our case, the antibodies directed against high and low molecular weight cytokeratin, the epithelium specific antigen and the epithelial membrane antigen excluded sarcomatoid carcinoma, synovial sarcoma and diffuse malignant mesothelioma, a diagnosis with consider when the tumor touches the pleura or mediastinum.

Primary pulmonary sarcomas are rare. Most malignant tumors of the spindle-shaped lung cells have been shown to be sarcomatoid carcinomas with immunochemical staining for cytokeratin. In addition, metastatic sarcomas involving the lung are much more common than primary pulmonary sarcomas. Usually, recognizing sarcoma as a malignant spindle cell lesion in a cytology preparation is simple. The specific classification of the neoplasm can be problematic. Virtually all types of sarcomas can be primary in the lungs and cytological samples can have different appearances, depending on the cell of origin. Most sarcomas produce cellular samples of discohesive malignant spindle cells with irregular hyperchromatic nuclei. A fascicular or leaf arrangement of the cells is possible. Sometimes an osteoid, chondroid or other matrix can be identified to aid in the specific diagnosis. A block of cytological cells should be attempted in case of possible sarcoma to facilitate immune histochemical staining. Careful examination of a patient's clinical history is warranted to rule out the possibility of metastatic lung sarcoma. Above all, a malignant melanoma should also be considered whenever a malignant neoplasm of spindle cells of the lung is encountered.

Primary pulmonary sarcomas are rare, and a variety of them occur. The immune phenotype of neoplastic cells that form such neoplasms is essentially the same as that of sarcomas that occur in soft tissue and other organs. Etienne-Mastroianni and associates92 carried out a clinical pathological study of 12 cases of primary pulmonary sarcomas. The histological diagnosis of these 12 neoplasms was confirmed by detailed immunohistochemistry. Nearby remained 7 leiomyosarcomas, 2 monophasic synovial sarcomas, 1 peripheral nerve sheath tumor, epithelioid sarcoma and 1 malignant fibrous histiocytoma. Nine of the 12 patients were operated on; pneumonectomy "" and 6 lobectomies with additional resection in 2 cases. Four patients received chemotherapy and 2 patients received radiotherapy. Follow-up was available on the 12 patients. Survival ranged from 3 to 144 months, with an average of 42 months. Long-term endurance up to 3 years was pragmatic in 5 patients. The overall 5-year survival rate was 38%. The authors concluded that primary pulmonary sarcomas were rare and aggressive tumors, and that the treatment and prognosis did not differ from those used for other soft tissue sarcomas.

Biography: Shroque Zaher is a consultant Histopathologist specializing in hematopathology, pulmonary pathology and medical training. She completed her pathology training at the deaneries of London and the east of England and obtained her CCT in 2016. She obtained her FRCP from the Royal College of Pathologists, United Kingdom in 2015.

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