Giant ancient schwannoma of the pleura: Commentary

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A case of giant ancient schwannoma of the pleura was reported in the current issue of the journal.[1] The pleura forms a continuous layer over thoracic structures and is divided into visceral and parietal pleurae. The former is devoid of somatic innervation though receives innervation from the vagus and sympathetic trunks while the latter is innervated by both somatic and sympathetic fibers, as well as parasympathetic fibers, via the intercostal nerves. In addition, the diaphragmatic pleura is supplied by the phrenic nerves. A benign peripheral nerve sheath tumor (BPNST) can be classified as schwannoma, neurofibroma, and perineurioma while hybrid BPNSTs, biphasic tumors with discrete areas of more than one histologic type have also been reported.[2] A BPNST can arise as a primary pleural tumor originating from the above-mentioned nerves, and intrathoracic BPNSTs are commonly located in the posterior mediastinum though rarely in the pleura.[3-5] Since nerves form peribronchial and periarterial plexuses in the lungs, in extremely rare cases, BPNSTs have been reported to develop within the pulmonary parenchyma and without pleural attachment.[5] Schwannomas originate in the Schwann's cells in the nerve sheath and are truly encapsulated.[6]

Microscopic examination of a schwannoma reveals two different patterns, designated Antoni Types A and B. Type A exhibits areas of spindle cell hypercellularity often arranged in a palisading fashion or an organoid arrangement (Verocay bodies) while Type B shows the areas of hypocellularity with abundant edematous fluid. Schwannomas have a high frequency of regressive changes, such as fatty degeneration, hemorrhage, perivascular hyalinization, and cystic formation. Intrathoracic schwannomas exhibiting prominent cystic and other regressive changes have been described as "ancient," under the assumption that they are slow-growing and have been present for a very long time. Ancient schwannomas can be very cellular, somewhat pleomorphic, and mitotically active, and thus easily confused with malignancy.[7] Although the authors emphasized its rarerity, the term "ancient" is of no particular clinical significance.

To a certain extent, computed tomographic (CT) scanning of the chest seems useful for preoperative evaluation of intrathoracic schwannomas as those findings reveal their well-defined margin, an ovoid or round shape, and either iso- or hypo-attenuated density as compared to the muscles of the chest walls, along with minimal or heterogeneous enhancement on contrast-enhanced images.[3] Such findings will vary according to the extent of regressive change within the tumor. CT is also able to reveal the position of the tumor and its extent, and may also show small tumors that have originated from the pleura. On the other hand, the origin of a huge tumor is more difficult to identify as shown in the present case;[1] thus, careful examination during surgery is essential to determine its source. Magnetic resonance imaging may be helpful for evaluating a pleural BPNST. Schwannomas typically show hyper- or iso-intense on T1-attenuated images and heterogeneous hyperintense on T2-attenuated images according to regressive changes. However, a definitive preoperative diagnosis of pleural schwannoma is usually challenging for the following reasons: (1) A schwannoma exhibits hypocellularity in Antoni Type B areas in sections obtained with an ultrasound-guided biopsy procedure, (2) tumor origin is difficult to identify, especially with a huge tumor, and (3) a pleural schwannoma is extremely rare.

For the treatment of a pleural schwannoma, complete removal of the tumor is indispensable. Video-thoracoscopy is useful for determining the best approach together with information on its position and extent shown by CT scanning. However, in cases with a huge tumor, complete removal will require an open thoracotomy,[1,8,9] with the aspiration of fluid from the cystic part during the operation possibly useful to reduce its mass.[10]

A pleural schwannoma is extremely rare, and reports of this tumor are useful in the field of clinical medicine. It is important to keep this rare tumor in mind for differential diagnosis of an intrathoracic lesion.

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