resection of pulmonary IMT, with a 5-year survival rate of 91.3%. However, a 60% recurrence rate in those receiving incomplete resection has been reported in the same study. Lee and colleagues [7] mentioned a case of IMT originating from the pleura and stated that a tumor measuring 27 × 22 mm was successfully removed from the adjacent rib tissue without performance of rib resection, and the patient’s 6.5-year follow-up visit was uneventful. Smaller tumors without chest wall invasion may allow total excision of the tumor without rib resection. In cases of chest wall tumors, wide excision is very important because local recurrence in incomplete resection is high, leading to poor survival [8]. Therefore, we performed a wide excision of the chest wall to allow complete mass resection. Nonsurgical treatment modalities including radiotherapy, chemotherapy, and steroids may have a place in the setting of incomplete surgical resection, multifocal disease, and tumor recurrence, but the efficacy of these alternative modalities remains controversial.

Nuchal fibroma is another rare, benign, solitary soft tissue tumor characterized by hypocellular dense collagens in the dermis and subcutaneous fat layer. It usually develops in the posterior neck and upper back. Although some cases have been associated with trauma and diabetes, the exact cause is still unknown [2]. Clinically and radiologically, it may resemble more common conditions. We have no information about an association between IMT and NF. In this respect, IMT and NF may be considered as unrelated pathologic conditions in our case.

In conclusion, pleural IMT and NF are rare clinical entities, and only limited data are available in the literature. To the best of our knowledge, this is the first report suggesting an association between IMT and NF. We also herein presented the largest intrathoracic IMT in reported cases so far, and emphasized another possible association between IMT and CA-125. In this respect, our case adds some new information to the literature.

References
a 5.8-cm × 3.7-cm poorly defined isodense soft tissue mass with marked enhancement in the middle and lower segments of the esophagus. The patient underwent Ivor-Lewis esophagectomy and reconstruction with mediastinal and abdominal lymph node dissection. The tumor had not infiltrated the bronchial tree. It was an R0 resection with a microscopically margin-negative resection, in which no gross or microscopic tumor remained in the primary tumor bed.

On macroscopic examination, the mass appeared to be as a poorly demarcated, circumscribed, solid tumor (5.8 cm × 4.5 cm × 4.0 cm), with a grayish white surface. The tumor invaded the adventitia of the esophagus. Histologically, the tumor was encapsulated and was composed of sheets and nests of small, uniform, rounded cells with centrally placed, round nuclei; amphophilic to lightly eosinophilic cytoplasm; and prominent nucleoli (glomus cells) surrounding capillary-sized vessels; and local necrosis seen in infiltrative sheetlike lesions. Nuclear atypia was present, with increased high mitotic activity (up to 10 mitoses per 50 high-power fields). Moreover, small tumor nodules were clearly intravascular and showed neural invasion (Fig 2). Immunohistochemically, the tumor cells were positive for smooth muscle actin and vimentin and negative for desmin, CD34, CD117, S-100 protein, and creatine kinase. Other markers such as Syn, CD56, neuron specific enolase, and L-chicoric acid were also positive. The proliferating marker Ki67 was 10%. Remarkably, two of three lymph nodes retrieved from the subcarinal area revealed metastasis, and another 17 lymph nodes from the mediastium and abdomen were free of metastatic tumor. The final pathologic diagnosis was malignant glomus tumor of the esophagus with mediastinal lymph node metastases. The patient recovered uneventfully and was discharged 8 days after the operation. He was doing well with no evidence of recurrence or metastasis at his most recent follow-up visit 11 months after treatment.

Comment

Glomus tumors, first described by Mason in 1924 [2], are neoplasms originating from glomus bodies in the dermis or the subcutis of the extremities. Extracutaneous presentations occur but are rare, especially in the visceral organs, where glomus bodies are sparse or even absent [1]. Rare sites of this disease have been reported, including mediastinum, respiratory tract (lung, trachea),
gastrointestinal tract (stomach, colon), and kidney [3–8]. Although glomus tumors are predominantly benign, they rarely demonstrate an aggressive and malignant clinical course and histologic characteristics. However, malignant glomus tumor of the esophagus is extremely rare. We believe that this is the first reported case of malignant glomus tumor arising from the esophagus.

Because of its rarity and nonspecific clinical presentation, this entity is diagnostically challenging. The results of imaging studies, including computed tomography, magnetic resonance imaging, endoscopic ultrasonography, and endoscopy, are not specific. Preoperative cytology may yield inconclusive or misleading results. Immunohistochemistry is an important part of the workup in precisely diagnosing malignant glomus tumor, particularly if the tumor is in a rare location.

Folpe and colleagues [9] and Gombos and colleagues [3] proposed a classification scheme of glomus tumor with atypical features. According to this classification, a malignant glomus tumor should fulfill at least one of the following criteria: (1) a deep location and size of more than 2 cm, (2) the presence of atypical mitotic figures, or (3) a combination of moderate to high nuclear grade and mitotic figures (5 mitoses/50 high-power field).

Histologically, the glomus tumor is a well-circumscribed lesion consisting of convoluted capillary-sized vessels surrounded by glomus cells in a hyalinized or myxoid stroma. The tumor may have a highly vascular pattern reminiscent of a hemangiopericytoma. The cells are monomorphic with a rounded, regular shape and are somewhat cohesive, giving them an epithelioid appearance.

Malignant glomus tumors have to be differentiated from other lesions, such as carcinoid tumor, hemangiopericytoma, paraganglioma, smooth muscle neoplasms, and metastatic tumors. Those tumors have distinctive histologic and immunohistochemical features and were effortlessly differentiated from glomus tumors.

The invasive and metastatic potential of glomangiosarcomas remains enigmatic because of the scarcity of reported cases. After a careful review of the literature, we summarize the five reported cases of malignant glomus tumor with metastases. Khoury and colleagues [8] in 2005 reported one case of aggressive malignant glomus tumors of the hand in a 48-year-old woman. A wide local excision with negative margins was performed, but multiple lung metastases were evident at the 8-month follow-up visit. Another report of a malignant glomus tumor with metastases is that of Masson-Lecomte and colleagues [4], who reported a case of a hypervascular penile tumor responsible for high-flow priapism as the first clinical symptom of a metastatic glomus tumor. The patient received three lines of chemotherapy, and the penile tumor was surgically removed because of persistent erectile dysfunction and perineal pain. In addition, three other reported cases of glomangiosarcomas involved widespread metastases leading to death. Shim and colleagues [7] reported a primary lesion in the urinary bladder of a 57-year-old woman with metastatic pulmonary nodules who died 2 months later. Lamba and colleagues [5] reported a case of widespread metastases to the spine, pelvic bones, and other organ systems that ultimately proved fatal. Song and colleagues [6] reported a case of a 65-year-old woman with a malignant glomus tumor of the stomach, with metastases to the kidney and brain, who died 7 months after the original diagnosis.

Since 2001, fewer than 13 cases of glomangiosarcoma have been reported. Most cases appear to have arisen from a preexisting glomus tumor without metastasis. Only in five cases have metastases been documented. Three of the five resulted in fatality, and two were being treated with chemotherapy. Clinical, histologic, and immunohistochemical examinations are essential in establishing the diagnosis of glomangiosarcoma. Aggressive surgical management remains the treatment of choice. Appropriate clinical follow-up is recommended.

References


Valve-Sparing Aortic Root Replacement With Translocation of Anomalous Left Coronary Artery

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An anomalous left main coronary artery arising from the right coronary with a single coronary ostium is an exceptionally rare anatomic variant. Here, we report a patient...