Tracheal glomus tumor: An aggressive rare neoplasm

Sir,

Herein, we present a rare case of primary malignant glomus tumor (GT) of the trachea with lung metastasis. Primary tracheal tumor consists of fewer than 0.1% of all malignancies.[1] Tracheal GT is an even extremely rare subtype, which commonly arises from glomus bodies of the subcutis/dermis, surrounding the arteriovenous malformations and accounts for <2% of all soft-tissue sarcoma.[2] The World Health Organization classified GT into benign, malignant, and glomus tumor of uncertain malignant potential.[3] The diagnosis of GT is rendered with histopathology and immunohistochemistry pattern. The index case is 35-year-old male, nonsmoker, nonalcoholic, with no medical comorbidities, no relevant family history and no significant past medical or surgical treatments, presented 3 years ago elsewhere with complaints of cough with bleeding and respiratory distress. The high-resolution computed tomography scan of the chest revealed polypoidal mass in mid trachea size 3.2 cm × 2.1 cm with the loss of fat plane to esophagus. Patient underwent the right anterolateral thoracotomy with excisional biopsy of tracheal growth with tracheoplasty. Surgical specimen histopathology was suggestive of malignant GT with close resection margins. Tumor cells were immunopositive for smooth muscle actin (SMA) and vimentin and immunonegative for CK7, CK20, epithelial membrane antigen, and synaptophysin. Post 2 years disease-free interval, the follow-up positron-emission technology scan suggested an ^{18}Flurodeoxyglucose (FDG)-avid (standard uptake value - 21.6) 2.6 cm × 1.4 cm × 2.3 cm lesion in tracheoesophageal region. Multiple bilateral FDG avid lung nodules were noted, largest size 1.4 cm ×1.1 cm in the lingular lobe. Overall features revealed locoregional recurrence with bilateral lung metastasis. The patient self-referred to our center concerning further treatment. The paraffin block review of previous surgery confirmed the diagnosis as tracheal glomus tumor [Figure 1]. The multidisciplinary tumor board advised for palliative chemotherapy with single-agent doxorubicin. The patient received six cycles of chemotherapy with no Grade — toxicities. The radiological disease progression [Figure 2] occurred on chemotherapy with an initial temporary partial clinical response. Only one second-line chemotherapy pazopanib was administered because of uncontrolled hypertension and hand-foot syndrome. Prognostication of the disease explained along with the administration of etoposide-based metronomic palliative chemotherapy. The patient succumbed to respiratory failure after approximately 1 year of the above-mentioned multiple lines of chemotherapy. The diagnosis of malignant GT is critical for clinicians because of resemblance to carcinoids in gross morphological endoscopic and microscopic appearances in the frozen section. They are immunopositive for SMA and vimentin, but immunonegative for carcinoid-specific markers such as CK7, CK20, synaptophysin, and chromogranin. Salt and pepper appearance (characteristic of carcinoid tumor) of nuclear chromatin is generally absent in glomus tumor. Complete tumor resection with a negative margin is the standard of care for malignant tracheal GT because of high locoregional failure and distant metastasis in a patient with incomplete tumor resection.[4] Metastasis is the main cause of death in GT. Furthermore, data for adjuvant treatment are limited in literature.[5] Bronchoscopic resection does not suffice in the bulky aggressive tracheal tumor. However, initial tumor debulking/coring is pivotal for effective airway restoration and providing diagnostic information before definitive surgery.[2]

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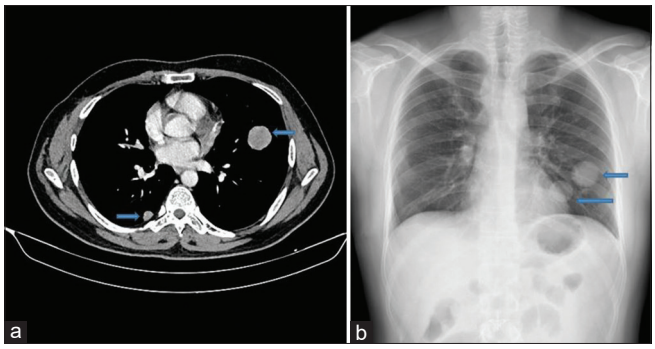

Figure 1: (a) Contrast-enhanced computed tomography scan showing bilateral multiple metastasis (blue arrows). (b) Chest X-ray image showing multiple lung metastasis (blue arrows)

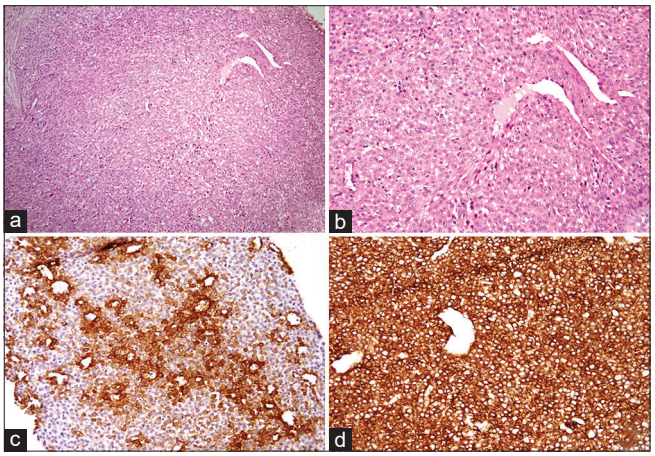

Figure 2: (a) Low-power photomicrograph of case of glomus tumor showing relatively monomorphic population of cells with intervening slit-like vascular channels. (H and E, ×100). (b) High-power photomicrograph showing cells with round nucleus, few showing prominent nucleoli, and moderate amount of eosinophilic cytoplasm. (H and E, ×200). (c) Immunostain for smooth muscle actin showing variable cytoplasmic positivity. (d) Immunostain for collagen IV showing cytoplasmic positivity

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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