Zebra tumor in uncommon location as a therapeutic challenge

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Introduction:

Thymic neoplasms, predominantly thymomas, account for almost one third of all tumors located in the anterior mediastinum. Among the histologic subgroups of neoplasms arising in this location, carcinoid thymic tumors represent a very rare subtype, that constitute only for 2-4% of all anterior mediastinal masses.

Case description: 34-years old male, Caucasian with a history of smoking, contacted his general practitioner due to radiating chest pain accompanied by tightness in the thorax. An X-ray followed by CT scans revealed a mass with solid and cystic components in the anterior mediastinum, sized: 9.3 x 5.8 cm and 7.5 cm (cc). Tumor was located between the ascending aorta, the aortic arch and both the manubrium and the body of the sternum. Within IX segment of the right lung spot sized 0.6 x 0.5 cm and 0.4 cm (cc) and other minor, infrapleural lesions in both lungs were revealed, comparing with available past CT scans no signs of progression were present. Neither lymph nodes enlargement nor other findings of metastatic nature were observed. Performed biopsy displayed mixed morphology of the tumor including atypical carcinoid and large-cell neuroendocrine carcinoma features with mitotic activity estimated as 13 mitotic figures /10 HPF (PHH-3), moderately and poorly-differentiated. Due to advanced local stage of the disease with major blood vessels infiltration and left brachiocephalic vein stricture the tumor was primary qualified as unresectable. Concerning ESMO/NCCN guidelines nine courses of cisplatin, six courses of etoposide and four courses of lanreotide in neoadjuvant fashion were administered. Partial response enabled further, possibly radical surgical approach with vascular prosthesis substitution, however the patient did not approve such extensive surgical plan, which resulted in performing R2 resection (ypT3N1Mx). Two months after the procedure CT scans revealed irregular, hypodense residual mass in left superior mediastinum alongside the aortic arch sized 4.8 x 2 cm and 4 cm (CC) and stable minor lesions in lungs. Subsequently urged SPECT scan did not reveal radioactive tracer uptake in residual mass, although abnormal concentration in pancreas was displayed without evident findings in low dose CT images. Patient received postoperative definitive radiotherapy which targeted residual tumor and tumor bed.

PTV 1 included tumor bed with total dose of 50Gy/25 fractions and boost to residual tumor with total dose of 65Gy/33 fractions. Subsequent PET scans showed abnormal metabolic activity in few minor lesions in lungs (SUVmax 2.7-3.2) of unknown nature without other relevant findings. Monthly administration of lanreotide in adjuvant fashion has been continued (six courses) after radiotherapy. Most recent CT scans, one year after initial surgery, displayed further discrete regression of residual mass sized 4 x 1.5 cm and 3.5 cm (CC) and stable lesions in lung.

Discussion: The clinical characteristics of our case are coherent with typical presentation resultant form literature (typical features were: adulthood, male-predominance, rare association with carcinoid syndrome, frequent presentation of nonspecific signs of an expanding mediastinal mass). Most authors go along with published guidelines and performed surgery in adequate cases. Such approach concerned our patient. Postoperative radiotherapy was administered due to risk factors and based on NCCN guidelines. Complexity of this rare case illustrates difficulties occurring in planning the treatment of uncommon malignancies.