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PRIMARY THYROID SARCOMA: A CASE REPORT AND REVIEW OF THE LITERATURE

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Introduction. Primary thyroid sarcomas (as a rule, a majority of them presents as angiosarcoma, leiomyosarcoma and malignant peripheral nerve sheath tumour) are one of the rarest types of mesenchymal malignancies with reported incidence worldwide less than 1%. *Ewing's sarcoma* is the second most common malignant bone tumour in children & young adults, however, it is rarely found in the elderly population. And *Malignant Gastrointestinal Stromal Tumour (GIST)* is a rare sarcoma located in the stomach and intestines. Therefore, it's a challenge for oncologists and pathologists to identify and classify the aforementioned tumours and put it under a single category if it's observed in unusual areas such as the thyroid.

Aim of the study: To present a unique case of primary thyroid sarcoma of uncertain histogenesis with regional lymph node metastases.

Material and methods. Data from Municipal Clinical Centre of Oncology, Minsk, PubMed, Cochrane Library, Scopus databases. Routine and immunohistochemical examination of biological material obtained from the primary tumour and its metastases was carried out according to generally accepted protocols, considering the recommendations of the manufacturer, staining was performed with Leica ST5020 and Bond max (Leica), and consuming ready-to-use antibodies.

Results and their discussion. A 67-year-old female patient was referred with a mass found in the left part of her neck. Total thyroidectomy with lymph nodes dissections was accomplished. The histology indicated an epithelioid type of malignancy with non-specific immune-morphological features: tumour cells did not express markers of epithelial differentiation such as cytokeratin, clone AE1/AE3, p63, EMA, and it was not lymphoma because no stain with CD45 antibody was revealed. Also, no muscle, vascular or nerve sheath differentiation was sustained. However, tumour cells reacted with antibodies CD117 (GIST, neuroendocrine tumours, melanoma), SOX10 (melanocytes and schwannian differentiation), TLE1 (synovial sarcoma, neuronal differentiation) and CD99 (differentiation of primitive neuroectodermal cells). FISH (Fluorescence in situ hybridization) with a number of probes approved for treatment decisions protocols did not reveal any specific mutations associated with melanoma, Ewing's sarcoma, GIST or synovial sarcoma. Therefore, this neoplasm was classified as a *"Malignant Extra-gastrointestinal Neuroectodermal Tumour" of the thyroid gland, pT2N1M0.* As a result of post-surgical complications, the patient succumbed to the vocal cord paralysis and paresis of the left recurrent laryngeal nerve. Chest X-ray, ultrasonography and scintigraphy did not reveal any distant metastases or other primary tumours that could have been the source of the aforementioned thyroid mass and metastases.

Conclusion. The major challenge in such rarest of rare case is to identify the exact type of tumour and to tailor an individual mode of treatment. Molecular testing like next-generation sequencing (NGS) may help to find specific therapeutic targets, however, treatment modalities are very scarce. Till date, total thyroidectomy and compartmental lymph nodes dissections with adjuvant external beam irradiation can be the only hope for patients to achieve remission.