


Sarcomatoid Carcinoma From a Thyroglossal Duct Cyst in a 20-Year-Old Woman: A Case Report

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Thyroglossal duct cysts (TDCs) are common; however, malignancies originating from these cysts are less frequent.¹ Approximately 1% of all TDCs develop some form of malignancy. The most common type is papillary carcinoma and typically presents later in life with a mean age of 39.5 years.^{1–3} Less frequent and with a worse prognosis is nonpapillary carcinoma originating from a TDC. In this article, a 20-year-old patient is presented with a carcinoma originating from a TDC wall, diagnosed as a sarcomatoid carcinoma. According to Dutch law, there is no need for Institutional Review Board approval for a case report.

Case Report

A 20-year-old woman was referred to our center with a slowly progressive tumor of the neck on the left side. On physical examination, a nonpainful relatively mobile mass (5×5 cm) was present just along the left side of the median line at lymph node level 2. No fluctuations or skin abnormalities were observed. General examination, otolaryngologic status, and routine blood test findings were normal. Ultrasound and computed tomography scans of the neck, thorax, and abdomen were performed, as well as a magnetic resonance imaging (MRI) scan of the neck. On ultrasound, a solid mass was seen with a diameter of 45×26 mm, suspected for a pathologic lymph node on which fine-needle aspiration cytology was performed. After the computed tomography scan, an MRI scan was advised for further specification of the abnormality. On MRI, a partly cystic and partly solid mass was seen left from the midline in the neck (**Figure 1**). Based on these results, the diagnosis could be a TDC, an epidermoid cyst, a pathologic lymph node, or a carcinoma originating from a TDC. The cytology showed a malignant cell population. Additional typing yielded no evidence of a primary origin. Therefore, histologic examination was required. A Sistrunk

procedure of the thyroglossal cyst was planned and performed, which went uncomplicated.

Histology revealed a cystic structure that was partly lined by multilayered flattened nonatypical epithelial cells. Next to the cyst, ectopic thyroid tissue was present as well as part of the hyoid bone: a frequent finding in TDC resections (**Figure 2**). However, most of the resection consisted of a proliferation originating from the cyst wall. These lesional cells partly had an epithelioid appearance and partly a distinct spindle cell morphology. Immunohistochemistry showed that the spindle cell component was reactive to BCL2, CD99, and vimentin (**Figure 3**). The epithelioid component reacted only to pan keratin. This morphology and immunohistochemically profile fits multiple entities, such as synoviosarcoma, (epithelioid) malignant peripheral nerve sheath tumor, and sarcomatoid carcinoma. Translocation analysis was performed (Archer FusionPlex), but no translocation could be found. Therefore, sarcomatoid carcinoma was the diagnosis per exclusionem, with a note that malignant peripheral nerve sheath tumor could not be ruled out. The patient was referred to the radiation oncologist for postoperative radiotherapy consisting of 54 Gy in 30 fractions.

Discussion

A recently published review on TDC carcinoma underlined the rarity of these cases.¹ This study extracted 164 cases from 98 articles. Pathology showed papillary (92.1%), squamous cell (4.3%), follicular (1.2%), combined papillary and squamous cell (1.2%), mucoepidermoid (0.6%), and adenosquamous cell (0.6%) carcinoma. Studies have shown that the most common diagnosis of carcinoma within TDCs is papillary thyroid cancer, followed by squamous cell carcinoma.^{1,4}

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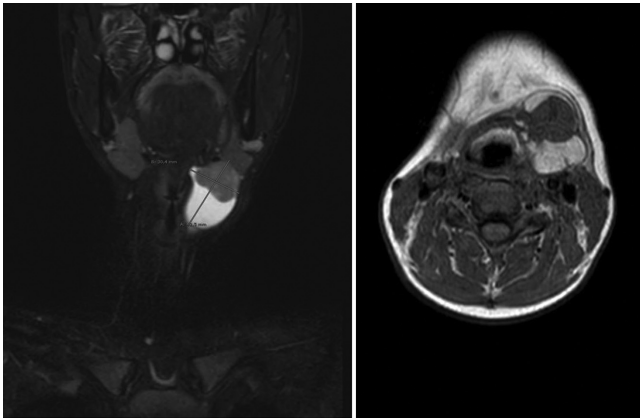


Figure 1. Magnetic resonance images show a partly cystic and partly solid mass left from the midline in the neck.

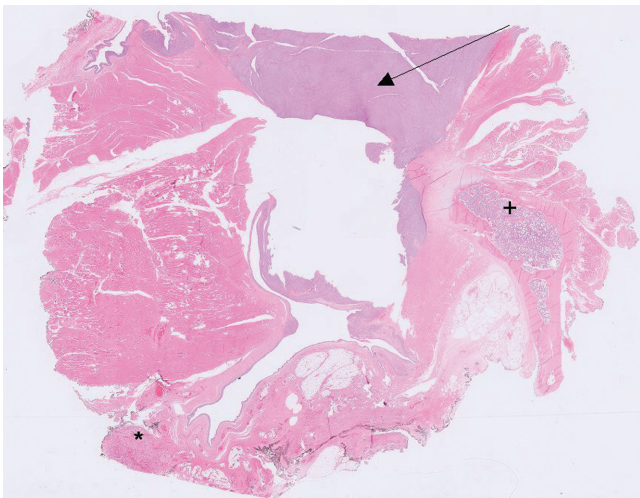


Figure 2. An overview of the resection specimen. *Left: thyroid tissue. + Right: part of the hyoid bone. Arrow: the proliferation origin from the cyst wall.

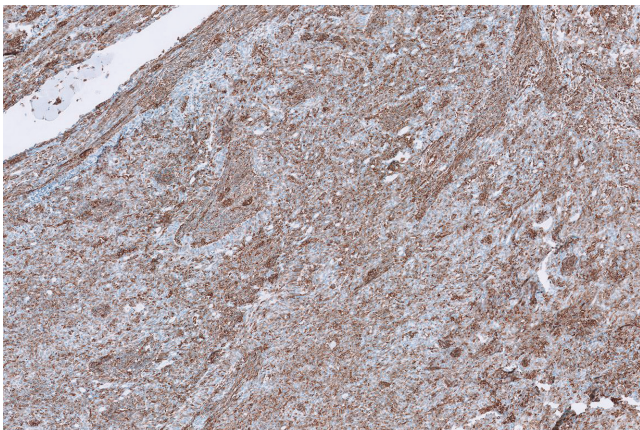


Figure 3. The vimentin-reactive cells in the lesion underline the sarcomatoid nature of the tumor.

Nonpapillary malignancies of the TDC are rare, thought to arise from the cyst walls, and are associated with a worse prognosis.⁵ A case report on a synovial sarcoma from a TDC therefore recommended a histologic diagnosis in all cases,

even typical clinical cases.⁶ A Sistrunk procedure is advisable in all malignancies in a TDC. Yet, due to the rarity of these entities, there is no unambiguous international postoperative treatment protocol. The criteria for postoperative radiotherapy generally include positive resection margins, lymph node involvement, and recurrent disease.⁵ However, in our institute, we decided to start with postoperative radiotherapy due to the difficult-to-classify tumor type. This shows that there are different approaches possible for the treatment of rare cases as these. It is therefore important to approach these cases from a multidisciplinary point of view to ensure an optimized treatment plan for the individual patient.

Conclusion

We present a 20-year-old woman with a sarcomatoid carcinoma originating from a TDC. This case report shows that because of the rarity of these entities, a multidisciplinary approach is necessary, and further diagnostics and treatment should always be performed when in doubt about the diagnosis.

Author Contributions

Wessel T. Stam, participated in the design of the study, collection and interpretation of the data, wrote and submitted the manuscript and gave final approval of the version to be published; **Sjors A. Koppes**, participated in the interpretation of the data, wrote parts of the manuscript and gave final approval of the version to be published; **Jeroen T. Kraak**, participated in the design of the study, interpretation of the data, revised the manuscript critically and gave final approval of the version to be published.


Disclosures

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