

Rare Solitary Fibrous Tumor of the Thyroid

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Background

- Solitary fibrous tumors were initially thought to arise exclusively within lung tissue, and hence were often referred to as a form of mesothelioma.¹
- It is now known that SFT is not restricted in its distribution, and characteristically presents in tissue such as the peritoneal cavity, pleural cavity, and rarely in regions of the head and neck.¹
- These tumors are most classically identified by their histological findings²:
 - Concurrent and opposing hypercellular and hypocellular regions
 - Seemingly random pattern of growth
 - Hyalinization reminiscent of keloid formation
 - Spindle-like cell morphology
- Surgical excision remains the standard of care. Chemotherapy and radiation are not considered mainstays of care and their use remains non-standardized in treatment of SFT.³

Case Presentation

A 41-year-old woman presented with a 2-year history of a slow growing left anterior neck mass that demonstrated rapid growth over the previous 2 months. Presenting symptoms included dysphagia to solids, voice changes, intermittent shortness of breath, and positional “pressure-like” chest pain. The patient denied trauma, fever, chills, weight loss, or neck pain. Additionally, she had no significant medical history and denied a family history of cancer. The patient denied alcohol and tobacco use.

On physical exam, a firm, non-tender, non-mobile left anterior neck mass was present, causing tracheal deviation, without associated palpable cervical lymphadenopathy. Her TSH was elevated at 5.51, but her free T4 was normal (1.22). CT neck with contrast showed a large, 11cm, heterogenous mass arising from the left thyroid gland with internal necrotic and hemorrhagic components and no calcifications (Figure 1). Mass effect was present on aerodigestive tract structures. CT chest was unremarkable. A core biopsy was interpreted as a spindle-cell neoplasm.

A total thyroidectomy was performed a month after diagnosis. The patient tolerated surgery well with no complications. Pathology confirmed a completely encapsulated SFT of the left thyroid lobe with negative margins. The immunohistochemistry panel was positive for Vimentin, CD34, BCL-2, SMA, CD99, Ki-67, PAX8, and STAT6. Diffuse nuclear STAT6 positivity in the lesion supported the final diagnosis. The right thyroid lobe was free of tumor.

Case Presentation Imaging and Pathology

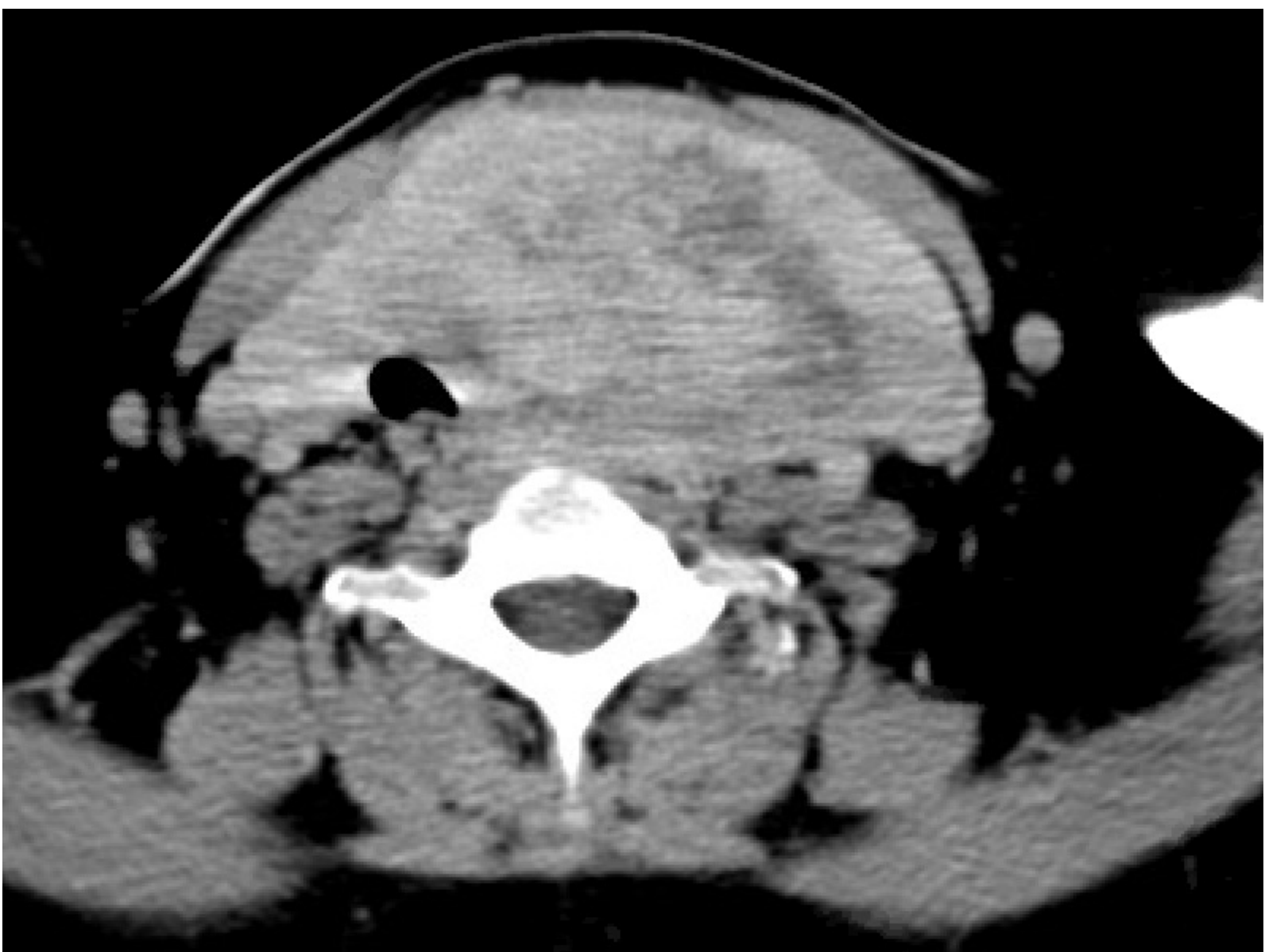


Figure 1: Computed tomographic image with contrast showing the 11cm left neck mass and resulting tracheal deviation

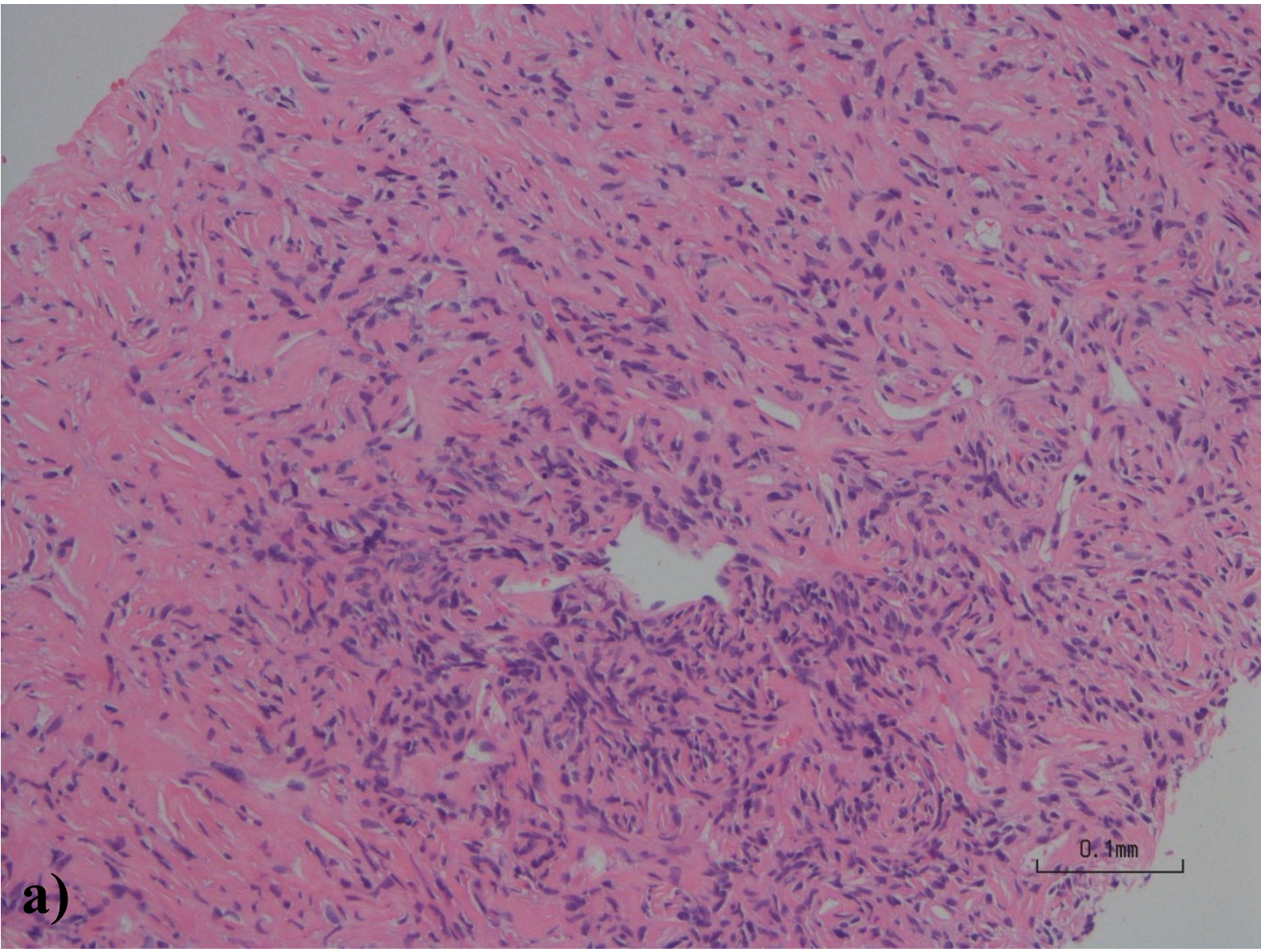


Figure 2: Core biopsy images demonstrating a) hypercellular and hypocellular regions b) random growth pattern

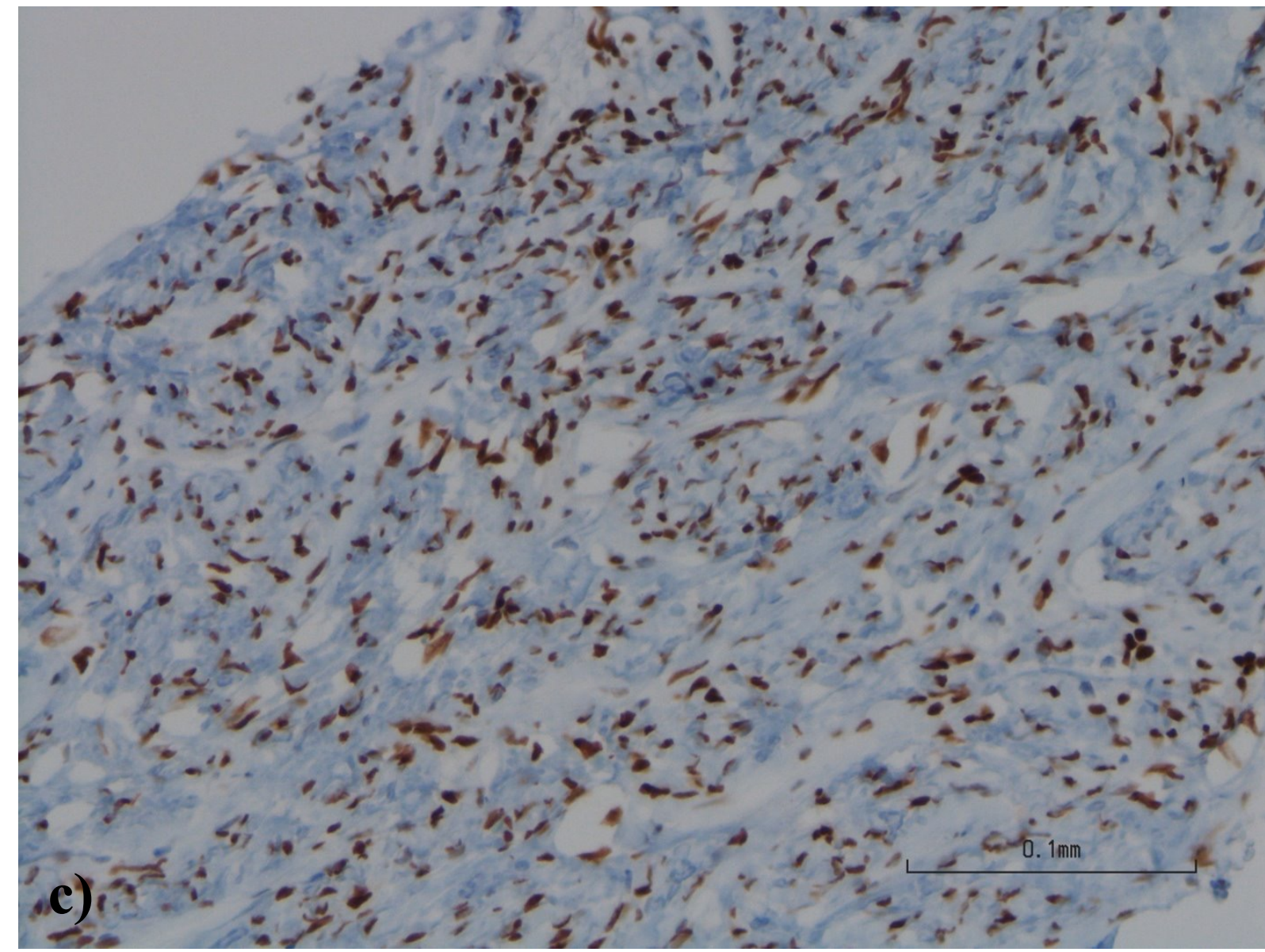
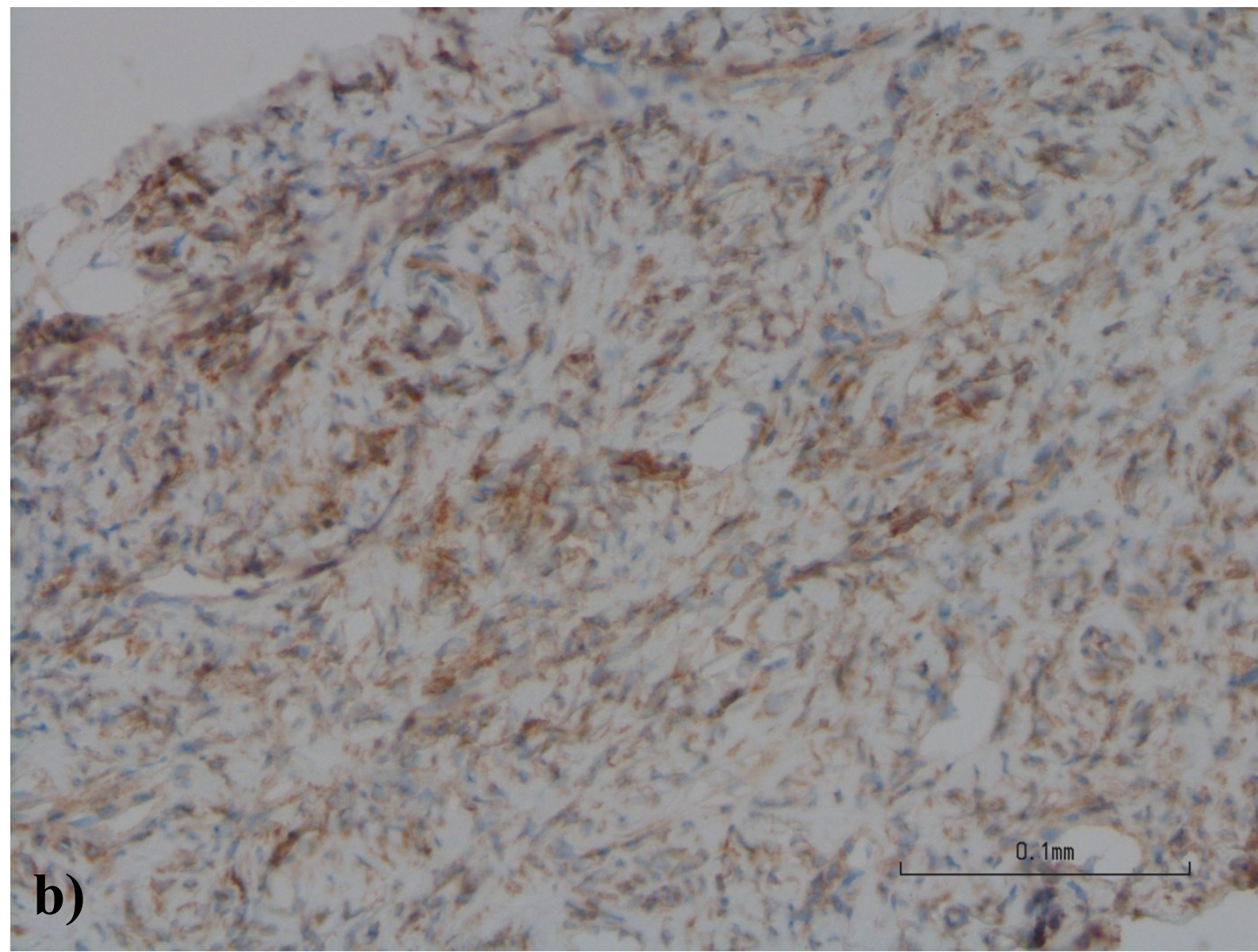
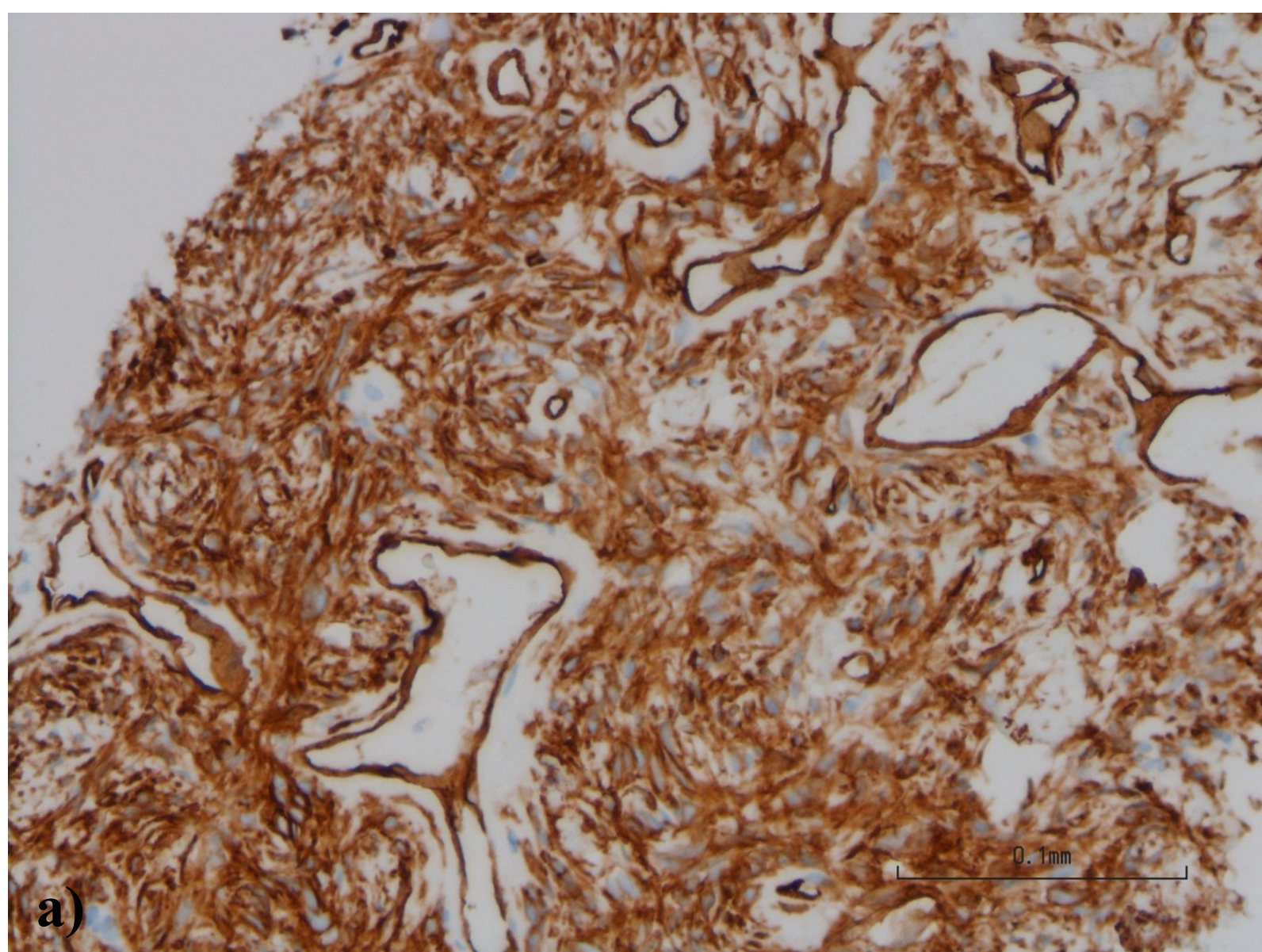
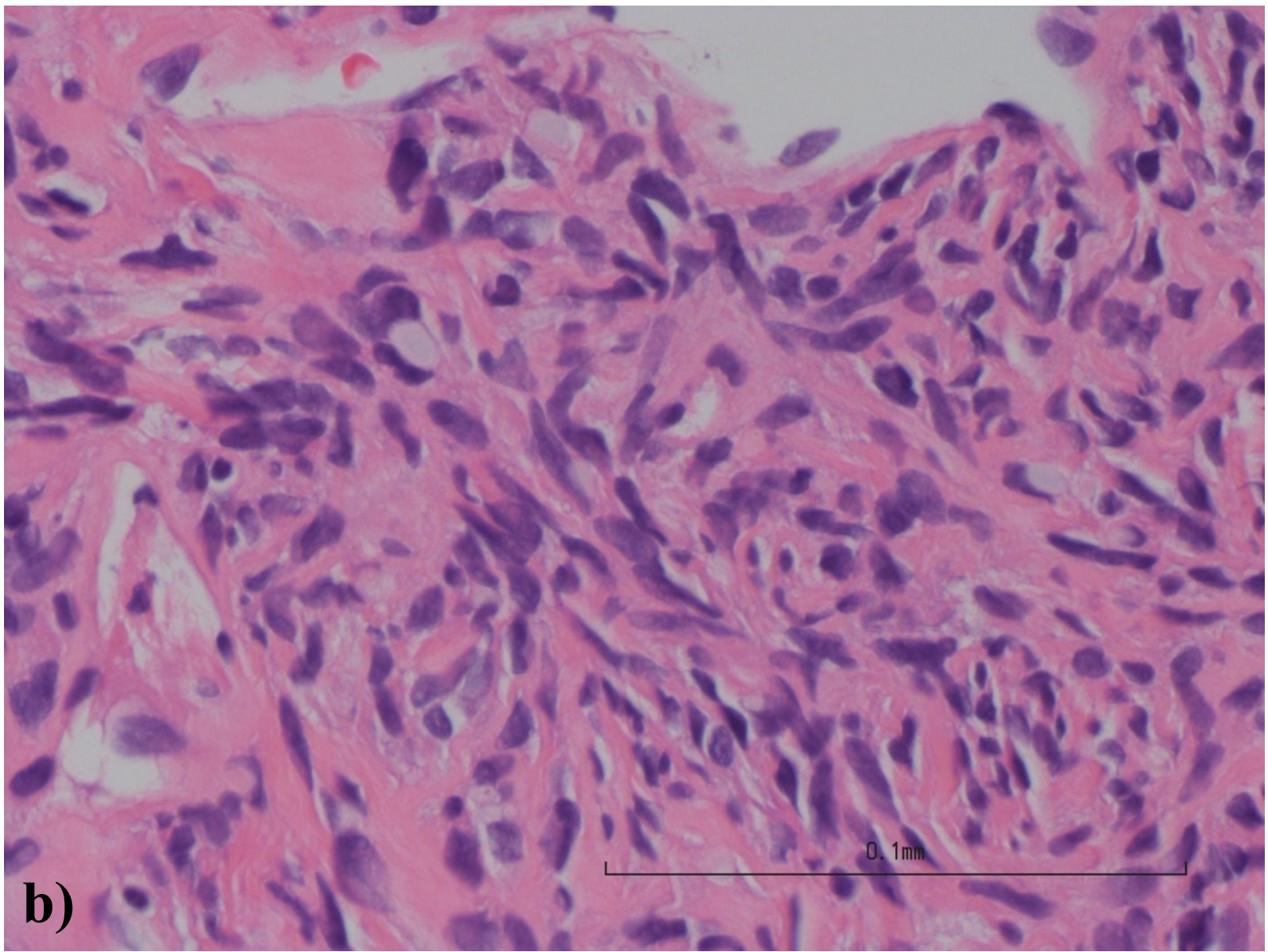


Figure 3: Immunohistochemistry staining patterns showing a) positive CD34 stain b) positive CD99 stain c) positive nuclear STAT6 stain

Discussion

- Solitary fibrous tumors tend to grow slowly and painlessly, thereby often going unnoticed until the tumor becomes large enough to cause compressive symptoms.
- Historically, SFT diagnosis was tedious, with no pathognomonic imaging findings, and thus relied on the previously mentioned non-specific histology features for diagnosis.⁴
- More recently, immunohistochemistry and genetic analysis has proven the NAB2-STAT6 gene fusion product to be consistently implicated in SFTs.⁴
- The STAT6 biomarker demonstrates good sensitivity (98%) and specificity (85%) for SFTs, especially those of nuclear origin. Therefore, this genetic marker serves as an excellent diagnostic tool for SFT, regardless of anatomical location.
- To date, there are no definitive pathologic criteria that differentiate benign from malignant SFTs. The SFT presented displayed characteristics of a malignant diagnosis based on its recent rapid growth, necrosis and hemorrhagic patterns, and large size (>10cm). Following current standard of care guidelines, surgical intervention without adjuvant chemotherapy or radiation was performed and the patient is currently disease free at 24 months post-op.

Conclusion

- Because SFTs of the thyroid are rare, it is important that we accurately identify SFTs in the thyroid in order to gather more information on the clinical and pathologic characteristics, treatment delivery and outcomes.
- The STAT6 marker continues to prove to be important in making the pathologic diagnosis and should be investigated as a potential therapeutic target in the future.

References

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