Case Report

TREATMENT OF NECK SOLITARY FIBROUS TUMOR: A CASE REPORT

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ABSTRACT

Solitary fibrous tumor is a rare tumor of mesenchymal origin that accounts for less than 2% of all soft tissue masses. Head and neck sites include the oral cavity, sinonasal region, soft tissues of the neck, thyroid, parotid gland, scalp, and larynx. Because of its rarity, developing specific diagnostic strategies and treatment planning was difficult. We report a case of a soft tissue solitary fibrous tumor of the neck in a 50-year-old male patient, localized between the small rectus muscle and the inferior oblique muscle of the head. We describe the diagnosis, pathological pattern, surgical treatment, and follow-up at 3 and 5 weeks after radical surgery.

KEYWORDS: *solitary fibrous tumor, sarcoma, neck, soft tissue neoplasms*

INTRODUCTION

Solitary fibrous tumor is a rare tumor of mesenchymal origin that accounts for less than 2% of all soft tissue masses (1). Initially identified in the pleura, solitary fibrous tumors has been identified in multiple anatomic locations and can arise anywhere in the body, like extremities, abdomen, superficial trunk, head, and neck (2-4).
Head and neck sites reported in the literature include the oral cavity, sinonasal region, soft tissues of the neck, thyroid, parotid gland, scalp, and larynx (4).

Because of the low incidence of solitary fibrous tumors, several data have been derived from small retrospective series and cases, making it difficult to develop specific diagnostic strategies and treatment planning. An extensive list of differential diagnoses and molecular genetic analyses must be considered to make a correct diagnosis. Solitary fibrous tumors have recently been associated with a NAB2-STAT6 gene fusion with high specificity and sensitivity (5).

Open incisional biopsy by an experienced surgeon or a central needle biopsy is mandatory. Depending on location, radiologically guided biopsy can be useful (6). Tumor size at presentation is highly variable and is typically related to location. The median size is between 7-10 cm, ranging from 1 to 40 cm (7-10). We report a case of a soft tissue solitary fibrous tumor of the neck, deeply localized between the small rectus muscle and the inferior oblique muscle of the head.

MATERIALS AND METHODS

Received: 15 October 2022
Accepted: 22 November 2022

A 50-year-old man presented a painless mass on the right side of his neck. Computed tomography (TC) scan revealed a well-defined, ovoid mass measuring 5.63 x 5,06 cm (Fig. 1a, b). The occipital artery and the occipital nerve were separated from the mass. Magnetic resonance imaging (MRI) revealed a mass related to the anterior border of the right inferior oblique muscle of the head. It was T2 hyperintense and T1 isointense to the muscle, with no post-contrast enhancement. Many blood vessels were seen within the mass. Based on these imaging findings, a borderline vascular lesion arising from the right inferior oblique muscle of the head was suggested. Instead, fine-needle aspiration cytology-biopsy revealed a solitary fibrous tumor.

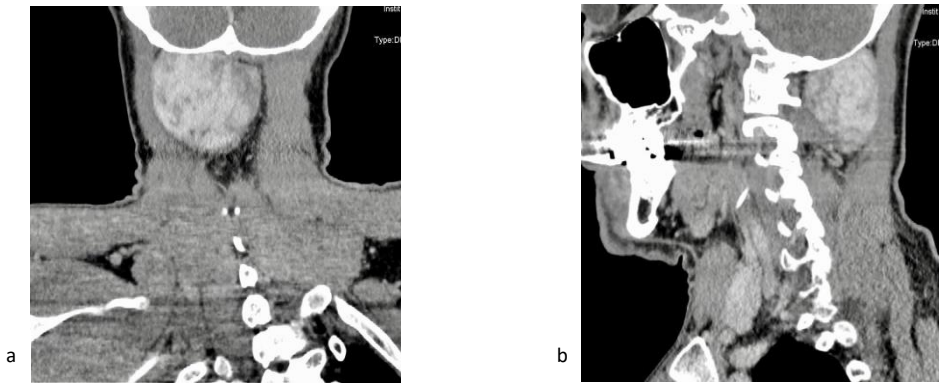
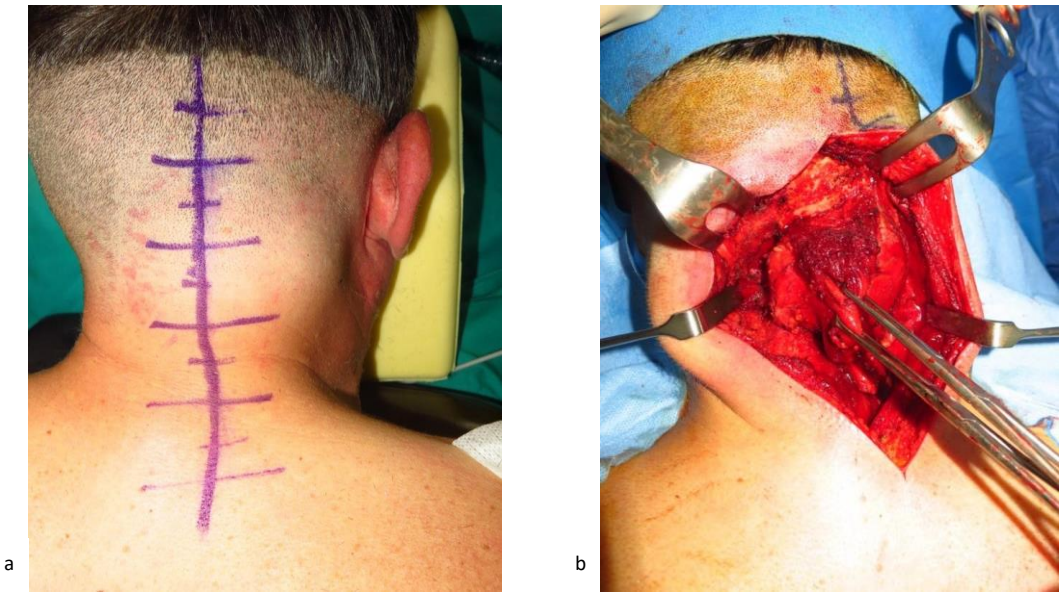


Fig. 1. *Computed tomography (TC) scan revealed a well-defined, ovoid mass; **a**): frontal view **b**): lateral view.*

Surgery was performed at Careggi Hospital by a multidisciplinary team consisting of maxillofacial surgeons, orthopedists, and neurosurgeons. The first surgical step was the incision of the skin and subcutaneous tissues (Fig. 2a), the incision of the trapezius muscle, the *Splenius capitis* muscle of the neck, the semifinal muscle of the head, and the small rectus muscle of the head. Subsequently, a blunt dissection was performed to find the mass that resulted adherent to the inferior oblique muscle of the head and the posterior big rectus muscle (Fig. 2b). The Occipital nerve and occipital artery were preserved. Dissection continued to the posterior arch of the atlas for complete mass removal (Fig. 2c). It was sent to a pathologist as a definitive exam (Fig. 2d).



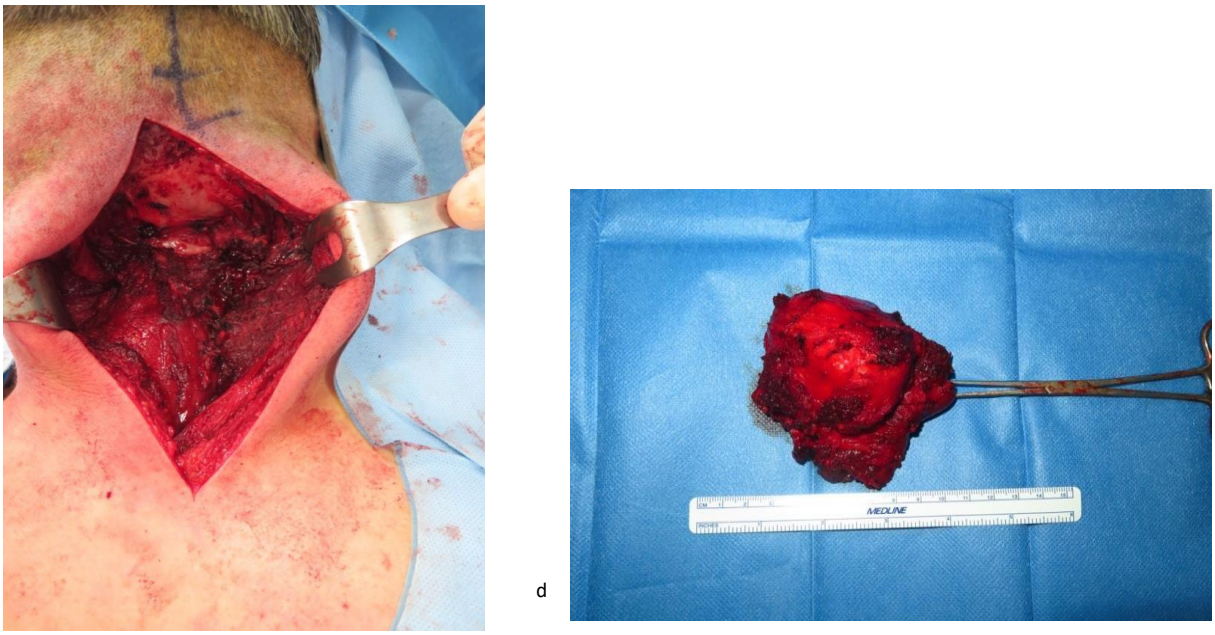


Fig. 2. ***a****): skin incision landmarks;* ***b****): blunt dissection of the mass;* ***c****): neck region after mass removal;* ***d****): excised mass.*

The patient was re-evaluated 3 weeks after surgery at our maxillofacial surgery department (Fig. 3a, b).The condition of the surgical wound was good. The patient reported paresthesia in the cervical and scalp region. Physiokinesis therapy was recommended.

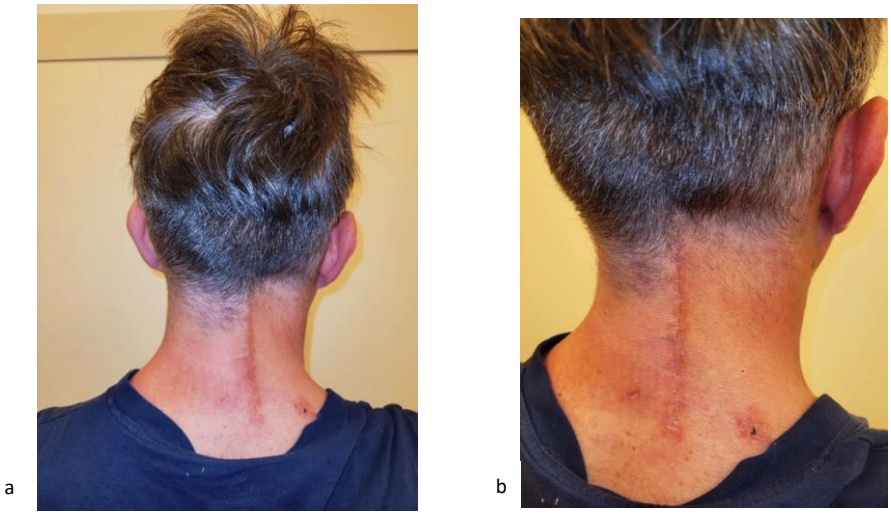


Fig. 3. *Check-up of surgical wound after 3 weeks.* ***a****): Posterior view;* ***b****): Lateral view.*

We also reported a subsequent follow-up 5 weeks after surgery: the patient showed a good surgical wound condition, but dysesthesia remained in the cervical and scalp region. The patient also reported a limitation in head torsion to the right, while the remaining movements of flexion-extension and rotation were preserved (Fig. 4a).

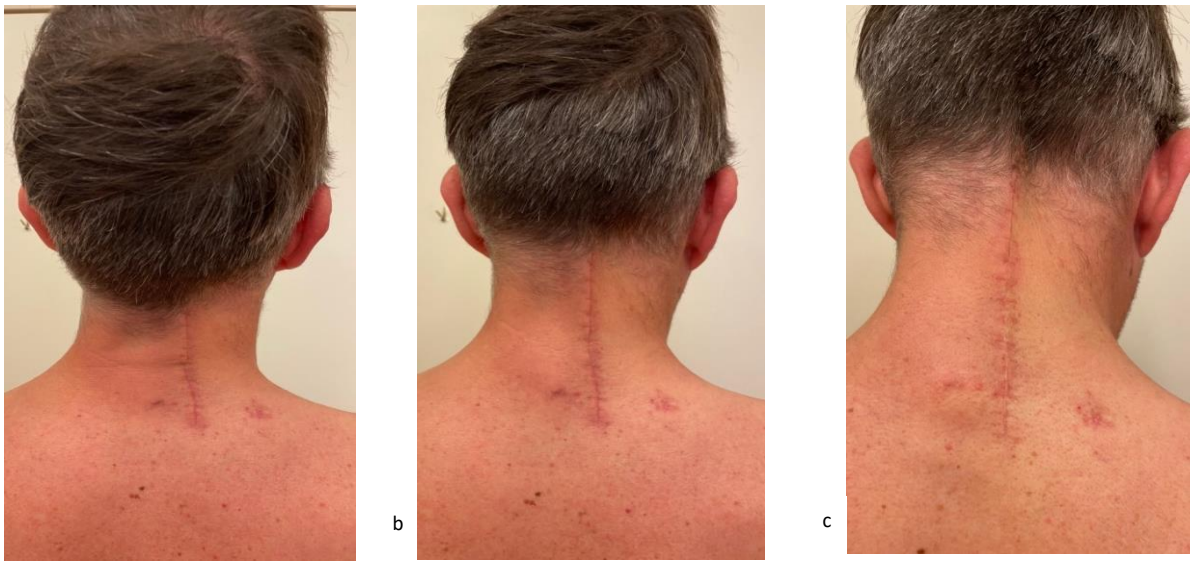


Fig. 4. *Check-up of neck movement at 5 weeks.* ***a****): Extension;* ***b****): Normal position* ***c****): Flexion.*

RESULTS

Gross specimen

The well-circumscribed ovoid mass measured 7 x 6 x 5 cm and was surrounded by fibro-adipose and skeletal muscle tissue. The lesion appeared solid and whitish on macroscopic examination and measured 4.4 x 3.8 cm (Fig. 2d). No necrosis or soft tissue invasion was identified.

Histopathology

Microscopic examination (Fig. 5a, Fig. 5b) revealed a well-circumscribed tumor composed of spindled to ovoidal cells with pale eosinophilic scant cytoplasm and indistinct cell borders surrounded by abundant myxoid stroma admixed with branching and hyalinized staghorn-shaped (haemangiopericytomatous) bloodvessel. No necrotic areas were found, and mitotic figures were rare (= 1/HPF). In view of these parameters,the age of the patient (<55 years), and the size of the lesion (<5 cm), the neoplasm has a low risk of recurrence and distant metastases (WHO 5th ed.).

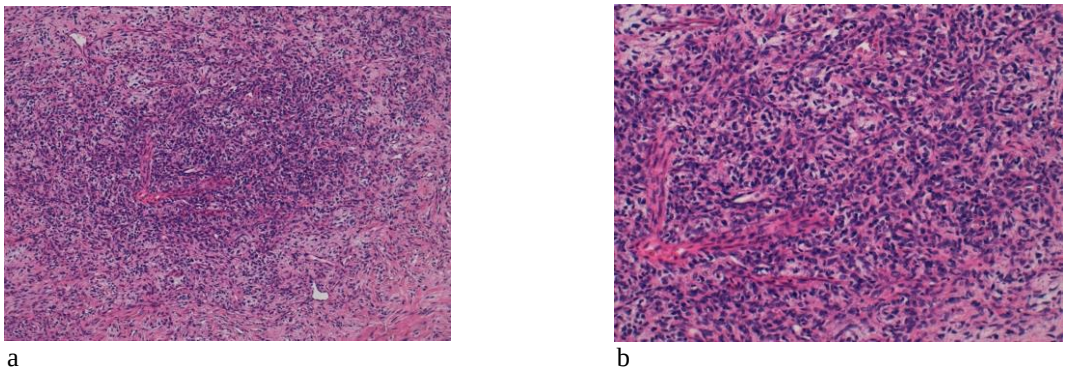
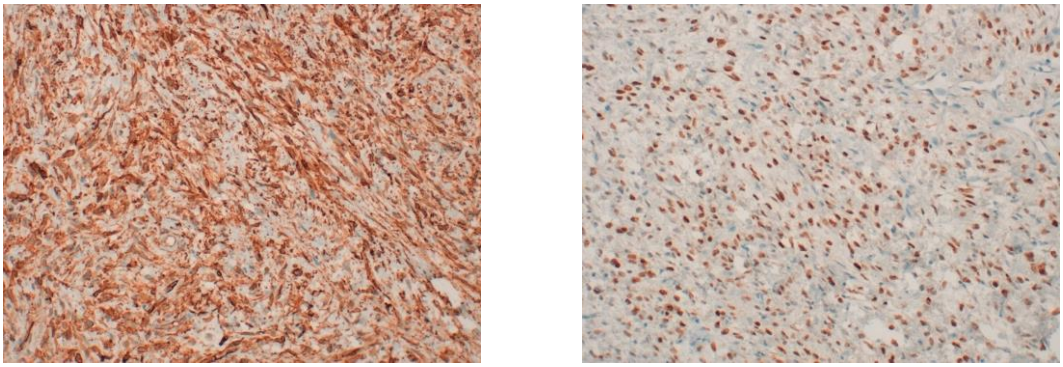


Fig. 5. ***a****): hematoxylin and Eosin stain, x100 magnification;* ***b****): hematoxylin and Eosin stain, x200 magnification.*

Immunohistochemistry

Immunohistochemical (IHC) investigations showed strong and widespread cytoplasmic positivity for CD34and a nuclear positivity for STAT6 in agreement with the diagnosis of SFT (Fig. 6a, b). The growth fraction of neoplastic cells evaluated with Ki67 equals 2%.



a b

Fig. 6. ***a****): the tumor cells diffusely expressed CD34 protein. Immunohistochemistry, CD34 antibody, x200 magnification;* ***b****): the tumor cells diffusely expressed STAT6 protein. Immunohistochemistry, STAT6 antibody, x200 magnification.*

DISCUSSION

Solitary fibrous tumors are fibroblastic tumors characterized by a prominent, branching, thin-walled, dilated (staghorn) vasculature and NAB2-STS6 gene rearrangement (12). Head and neck regions remain uncommon (18-22), accounting for only 11% in a series of 110 SFT cases by Demicco et al. (2).

Stanisce et al. estimated that a quarter of all extra-thoracic SFTs are localized in this region (4). Solitary fibrous tumors arising in the head and neck region are uncommon but well-recognized entities.

Davanzo et al. reported that SFT of the head and neck may originate from the sinonasal tract, oral cavity ororbit (1). In a recent meta-analysis, the median age at presentation of SFT was 51 years, and the median tumor size inthe neck was 5 cm (4). Recurrence (distal or local) occurs in 10-30% of SFTs, with 10-40% of recurrences reported after 5 years (12).

According to new recent WHO Classification of Tumor 5th Edition “Soft Tissue and Bone Tumors”, the most widely used model for metastatic risk incorporates mitotic count (≥ 2 mitoses/mm^2 or ≥ 4 mitoses/10 HPF) patient age (≥ 55 years) and tumor size stratified by 5 cm tiers to classify tumors into a low, intermediate,and high-risk group. A subsequent refinement includes necrosis as a fourth variable (12). Stanisce et al. found positive surgical resection margins to be the only significant risk factor for local recurrence (4).

Lau et al. presented a myxoid SFT occurring in the soft tissue of the neck and was treated with surgical resection alone and had no evidence of disease after 84 months of follow-up (13).

Our case was treated with surgical resection with negative margins at anatomy-pathological examination. No necrosis or soft tissue invasion was identified. The mitotic rate was 1per 10 high-power fields. Our multidisciplinary oncology group avoided postoperative adjuvant radiotherapy because the lesion was classified as a low-risk mass according to WHO guidelines (15). A nuclear magnetic resonance imaging scan was recommended 3 months after the last follow-up describedhere (5 weeks after radical surgery).

CONCLUSIONS

Literature suggests SFTs are associated with a favorable prognosis (11, 12). In conclusion, we described the diagnosis, treatment, and follow-up at 3 and 5 weeks after radical surgery of a solitary fibrous tumor arising from the soft tissue of the neck in a 50-year-old male patient.

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