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Case Report
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Treatment of Neck Solitary Fibrous Tumor: A Case Report

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Abstract

We present a 50-year-old male with a diagnostically arising solitary fibrous tumor in the soft tissue of the neck. The tumor was well circumscribed and deeply localized in the neck adherent to the inferior oblique muscle of the head and the posterior big rectus muscle.

Background: Solitary Fibrous Tumor (SFT) is a rare mesenchymal tumor, making up less than 2% of all soft tissue masses. While initially identified in the pleura, SFTs can develop anywhere in the body, including the head and neck region. Head and neck locations include the oral cavity, sinonasal region, neck soft tissues, thyroid, parotid gland, scalp, and larynx. Due to the low incidence of SFT, diagnostic strategies and treatment planning are challenging, as most data come from small retrospective series and case reports. SFTs have been linked to a NAB2-STAT6 gene fusion product, which is highly specific and sensitive for diagnosis. Pretreatment biopsy, ideally an open incisional biopsy or central needle biopsy, performed by an experienced professional, is crucial for diagnosis and classification. Tumor size varies significantly with location, with a median size between 7 cm - 10 cm. Grossly, these tumors are typically well-circumscribed with smooth surfaces, often lobular in shape. Distant metastasis is rare in reported SFT cases. This case report describes a solitary fibrous tumor located in the soft tissue of the neck in a 50-year-old male.

Methods: A 50-year-old male presented with a painless mass on the right side of his neck. A Computed Tomography (CT) scan showed a well-defined, ovoid mass measuring 5.63×5.06 cm. The occipital artery and nerve were clearly separated and preserved during surgery. Magnetic resonance imaging (MRI) revealed a mass intimately related to the right inferior oblique muscle of the head, appearing T2 hyperintense and T1 isointense to the muscle, with no post-contrast enhancement. Numerous blood vessels were observed within the mass. Fine-needle aspiration cytology-biopsy confirmed a solitary fibrous tumor. The patient underwent surgery at Cona hospital (FE) by a multidisciplinary team of otolaryngologists, orthopaedists, and neurosurgeons. The surgery lasted two and a half hours, involving incisions of the skin, subcutaneous tissues, trapezius muscle, splenius capitis muscle of the neck, semifinal muscle of the head, and small rectus muscle of the head. Blunt dissection localized the neoformation, which was adherent to the inferior oblique muscle of the head and the posterior big rectus muscle. The occipital nerve and occipital artery were preserved. The neoformation was completely removed down to the posterior arch of the atlas and sent for pathological examination. The patient was re-evaluated at 3 and 5 weeks post-surgery.

Results: The resected mass was a well-circumscribed, ovoid specimen measuring 7×6×5 cm, surrounded by fibro-adipose and skeletal muscle tissue. Macroscopically, the lesion was solid, whitish, and measured 4.4×3.8 cm. No necrosis or soft tissue invasion was identified. Microscopic examination revealed a well-circumscribed tumor composed of spindled to ovoidal cells with pale eosinophilic, scant cytoplasm, and indistinct cell borders. These cells were surrounded by abundant myxoid stroma mixed with branching and hyalinized staghorn-shaped blood vessels. Necrotic areas were absent, and mitotic figures were rare (equal to 1 per high power field). Based on these parameters, the patient's age (50 years, i.e, <55 years), and the lesion size (i.e, <5 cm), the neoplasm was classified as low risk for recurrence and distant metastases according to the WHO 5th edition. Immunohistochemical analysis showed strong and widespread cytoplasmic positivity for CD34 and nuclear positivity for STAT6, supporting the SFT diagnosis. The growth fraction of neoplastic cells, evaluated with Ki67, was 2%. Post-surgical follow-up at 3 weeks showed a good surgical wound condition, with the patient reporting paraesthesia in the cervical region and scalp. Physio kinesis therapy was recommended. At 5 weeks post-surgery, the surgical wound remained good, but dysesthesia persisted in the cervical and scalp regions. The patient also reported a limitation in head torsion to the right, while other movements (flexion-extension and rotation) were preserved. The anatomy-pathological examination confirmed negative surgical margins. Given the low-risk classification per WHO guidelines, postoperative adjuvant radiotherapy was not implemented.

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Conclusion: We report the diagnosis, treatment, and 3- and 5-week post-radical surgery follow-up of a solitary fibrous tumor in the soft tissue of the neck in a 50-year-old male patient. The median age for SFT presentation is 51 years, and the median tumor size in the neck is 5 cm. While recurrence (distal or local) occurs in 10-30% of SFTs, with 10-40% of recurrences reported after 5 years, positive surgical resection margins are identified as a significant risk factor for local recurrence. In this case, complete surgical resection with negative margins was achieved, and no necrosis or soft tissue invasion was identified. The low mitotic rate (1 per 10 high power fields) further supported the low-risk classification. The multidisciplinary oncology group opted against postoperative adjuvant radiotherapy due to the lesion's low-risk classification based on WHO guidelines. The literature generally suggests a favourable prognosis for SFTs. However, further studies on myxoid SFT are needed to fully understand its biological behaviour due to its rarity. A nuclear magnetic resonance imaging scan is recommended 3 months after the last follow-up.

Keywords: Solitary Fibrous Tumor (SFT); 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 tumor has been identified in multiple anatomic locations and can arise anywhere in the body like extremities, abdomen, superficial trunk, as well as the 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 tumor most data have been derived from small retrospective series and case reports making it difficult to develop specific diagnostic strategies and treatment planning.

An extensive list of differential diagnosis must be considered to make a correct diagnosis and molecular genetic analysis has proven vital to further differentiate soft tissue masses. Solitary fibrous tumor has recently been associated with a NAB2-STAT6 gene fusion product that has been shown to be highly specific and sensitive [14].

A pretreatment biopsy to diagnose and classify the mass is ideal. This biopsy should be performed by a person experienced in soft tissue mass biopsy techniques. An open incisional biopsy by an experienced surgeon or, alternatively, a central needle biopsy is preferred. Repeat biopsy may be considered if the initial biopsy is inadequate or fails to make the diagnosis. Depending on location, radiologically guided biopsy may prove useful [10]. Tumor size at presentation is highly variable and is typically associated with location. The median size among several studies is known to be between 7 cm - 10 cm with a range from 1 to 40 cm. [5-8].

Grossly, these tumors are typically well circumscribed with smooth surfaces and many times a lobular shape [5,6].

In the soft tissues of the neck, we reported only one other case with a well-defined ovoid mass measuring 4.8 x 2.6 cm x 1.7 cm [9].

None of the few reported cases of solitary fibrous tumor showed distant metastasis [11-13].

Here we report a case of a soft tissue solitary fibrous tumor of the neck, specifically the mass was deeply localized between the small rectus muscle of the head and the inferior oblique muscle of the head.

Methods

The patient is a 50-year-old man who presented with a painless mass on the right side of his neck. Computed Tomography (TC) scan revealed a well-defined, ovoid mass measuring 5.63 cm x 5,06 cm (Figure 1a, Figure 1b). The occipital artery and the occipital nerve were clearly separated from the mass and will be preserved during surgical treatment. Magnetic Resonance Imaging (MRI) revealed a mass which was intimately related to the anterior aspect of the right inferior oblique muscle of the head and 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. Fine needle aspiration cytology-biopsy revealed a solitary fibrous tumor.

The patient was candidate for surgery and was operated on at Cona hospital (FE) by a multidisciplinary team consisting of otolaryngologists, orthopaedists, and neurosurgeons. The surgical act lasted two and a half hours and involved incision of the skin and

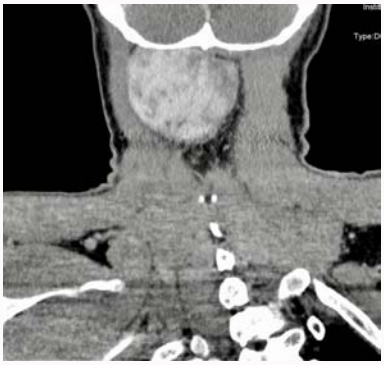

Figure: 1a Contrast-enhanced CT scan in coronal plane.

Figure: 1b Contrast-enhanced CT scan in sagittal plane.

Figure: 2a: Pre-operative surgical planning.

Figure: 2b: Identification of the lesion that appears adherent to the inferior oblique muscle of the head and the posterior big rectus muscle.

Figure: 2c: Complete removal of the lesion and identification of the posterior arch of the atlas.

subcutaneous tissues (Figure 2a), 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 with localization of the neoformation that was adherent to the inferior oblique muscle of the head and the posterior big rectus muscle (Figure 2b).

The Occipital nerve and occipital artery were preserved. Dissection continued to the posterior arch of the atlas to totally remove the neoformation (Figure 2c) that was sent to surgical pathologists (Figure 2d).

The patient was re-evaluated 3 weeks after surgery at our otolaryngology department (Figure 3a, Figure 3b). The condition of

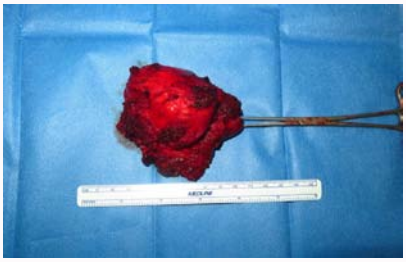

Figure: 2d: Macroscopic anatomy of the lesion.

Figure: 3a: 3-week follow-up.

Figure: 3b: 3-week follow-up.

the surgical wound is good. The patient reports paraesthesia in the cervical region and at scalp level. Physio kinesis therapy has been recommended.

We also report a subsequent follow-up at 5 weeks after surgery.

Good surgical wound condition, but dysesthesia remains in cervical and scalp region. The patient also reports a limitation in the torsion of the head to the right, while the remaining movements of flexion-extension and rotation remain preserved (Figure 4a, Figure 4b and Figure 4c).

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. On macroscopic examination the lesion appeared solid, whitish and

Figure: 4a: 5-week follow-up.

Figure: 4b: 5-week follow-up.

Figure: 4c: 5-week follow-up.

measures 4.4 x 3.8 cm (Figure 2d), No necrosis or soft tissue invasion was identified.

Histopathology

Microscopic examination (Figure 5a, Figure 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) blood vessel. 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 edition) [23].

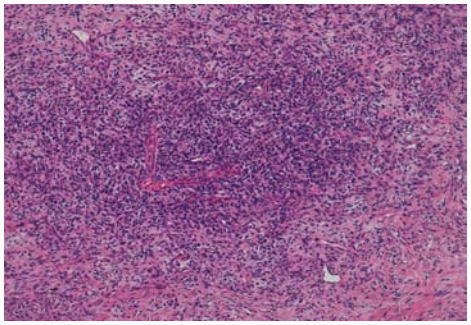

Figure 5a: Hematoxylin and Eosin stain, x100 magnification.

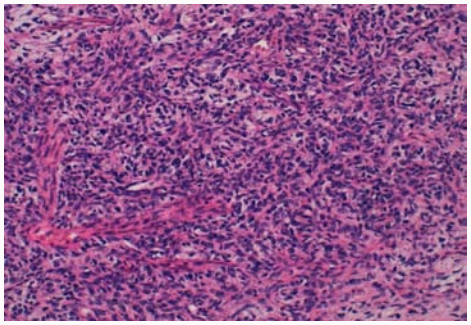

Figure 5b: Hematoxylin and Eosin stain, x200 magnification.

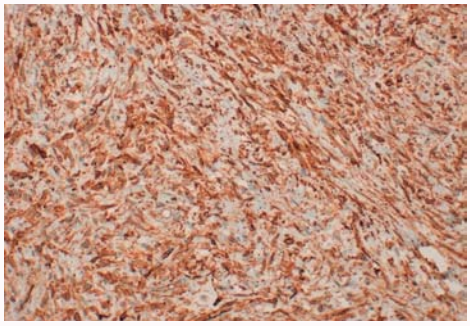

Figure 6a: The tumor cells diffusely expressed CD34 protein. Immunohistochemistry, CD34 antibody, x200 magnification.

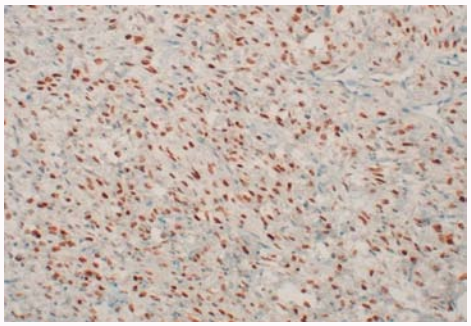

Figure 6b: The tumor cells diffusely expressed STAT6 protein. Immunohistochemistry, STAT6 antibody, x200 magnification.

Immunohistochemistry

Immunohistochemical (IHC) investigations showed a strong and widespread cytoplasmic positivity for CD34 and a nuclear positivity for STAT6 in agreement with the diagnosis of SFT (Figure 6a, Figure 6b).

The growth fraction of neoplastic cells evaluated with Ki67 is equal to 2%.

Discussion

Solitary fibrous tumors is a fibroblastic tumor characterized by a prominent, branching, thin-walled, dilated (staghorn) vasculature and NAB2-STS6 gene rearrangement [14,15].

The head and neck region remains uncommon, 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 butt well-recognized entities. Davanzo et al. reported that SFT of the head and neck may originate from the sinonasal tract, oral cavity or orbit. [1]. In a recent meta-analysis, the median age at presentation of SFT was 51 years and the median tumor size in the neck was 5 cm [4]. Recurrence (distal or local) occurs in 10% - 30% of SFTs, with 10-40% of recurrences reported after 5 years [15].

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/mm2 or ≥ 4 mitoses/10 HPF), patient age (≥ 55 years), and tumour size stratified by 5 cm tiers to classify tumours into low, intermediate, and high risk group. A subsequent refinement includes necrosis as a fourth variable [15-18].

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 [11].

Taking these analyses into account, 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 1 per 10 high power fields. Our multidisciplinary oncology group decided not to implement postoperative adjuvant radiotherapy because the lesion is classified as low risk according to WHO guidelines [19-23].

A nuclear magnetic resonance imaging scan is recommended 3 months after the last follow-up described here (5 weeks after radical surgery).

The literature suggests that SFTs are associated with a favorable prognosis [11,12]. However, this is limited by the rarity of the entity so more studies on myxoid SFT are needed before its biological behavior can be fully elucidated.

In conclusion, we describe the diagnosis, treatment, and follow-up at 3 and 5 weeks after radical surgery of solitary fibrous tumor arising the soft tissue of the neck in a 50-year-old male patient.

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