Turkish Archives of Otolaryngology
Türk Otolarengoloji Arşivi

Turk Arch Otolaryngol 2014; 52: 151-4

Solitary Fibrous Tumor of the Supraclavicular Region

Case Report

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Abstract

Solitary fibrous tumors of the head and neck are extremely rare, slow-growing, and generally asymptomatic benign neoplasms. To the best of our knowledge, only 153 cases of solitary fibrous tumors in the head and neck have been previously reported in the English literature until 2010. However, none has been in the supraclavicular region. Herein, we report a case of solitary fibrous tumor arising from the supraclavicular region. Characteristics, differential diagnosis, and treatment of the disease were also summarized.

Keywords: Solitary fibrous tumor, mesenchymal tumor, head and neck, treatment

Introduction

Solitary fibrous tumor (SFT) is a rarely seen mesenchymal tumor that generally develops on serosal surfaces (1, 2). More than 50% of these tumors occur in the thoracic cavity, but SFT cases were also reported to develop in extra-thoracic regions such as the liver, adrenal gland, and skin (3). SFT is rarely observed in the head and neck region (4). In this region, SFT is well-demarcated, grows slowly, and frequently occurs in the oral cavity (4-6). The symptoms are non-specific and depend on the affected areas where the tumor has developed (6). The immunohistochemical identification of CD34 positivity is the most important diagnostic finding. The most appropriate treatment method for SFTs in the head and neck region is complete surgical excision (6, 7). To the best of our knowledge, 153 SFT cases in the head and neck were published in the English literature until 2010 (6). Nevertheless, no case was reported in the supraclavicular region.

Case Report

A 53-year-old male patient was admitted to our clinic due to a slow-growing painless mass in the right supraclavicular region. The patient specified that he had firstly noticed the mass 5 years ago and he did not feel any discomfort except for esthetic concerns. His physical examination revealed a solid mass (diameter=5 cm) in the right supraclavicular region (Figure 1a). The mass was mobile, painless, and smooth-surfaced and the skin on it appeared natural. The results of other ear-nose-throat and system examinations were normal. On precontrast T1-weighted magnetic resonance imaging (MRI), a 55×50×45 mm-sized, well-demarcated, lobulated, and solid mass was found in the right supraclavicular region, posterior to the sternocleidomastoid muscle, and deep cervical plane (Figure 1b). While the solid mass displayed heterogeneous involvement in the postcontrast T1-weighted MRI, central hyperintensity was detected in the T2-weighted MRI (Figure 1c, d). No specific diagnosis was established via fine-needle aspiration biopsy (FNAB). Complete surgical excision was planned for a final diagnosis and treatment. The patient was informed about the surgical procedure in detail and then his written informed consent was obtained. The supraclavicular mass was completely excised with trans-cervical approach under general anesthesia. On macroscopic examination, it was observed that the solid mass was 65×50×30 mm in size and encapsulated and it included heterogeneous areas, the color of which changed from white to brown in the incision site (Figure 2a). Microscopic examination revealed a tumor comprising fusiform cells bordered by hyalinizing collagen bundles (Figure 2b). The tumor cells included elastic nucleus and narrow cytoplasm. Small myxoid focuses were available in the collagenous posterior plane. Features reminding malignancy potential such as margin infiltration, hypercellularity, nuclear atypia, and high mitotic activity were not observed. Immunohistochemically, the tumor cells were positive for CD34, vimentin (Figure 2c, 2d), and factor VIII, but negative for S100. Ki-67 proliferative index was low. These findings were consistent with the diagnosis of SFT. No problem was encountered during the postoperative period. Any finding associated with the disease was not found in the final control examination performed in 6 months postoperatively.

Discussion

SFT is a rare lesion. It was first defined in the pleura by Klemperer and Rabin in 1931. These tumors

This study was presented as a poster at the 35th National Otolaryngology Congress, 2-6 November 2013 Antalya, Turkey.

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Received Date/Geliş Tarihi: 29.12.2013
Accepted Date/Kabul Tarihi: 15.04.2014
Available Online Date/Çevrimiçi Yayın Tarihi: 08.07.2014

DOI:10.5152/tao.2014.303

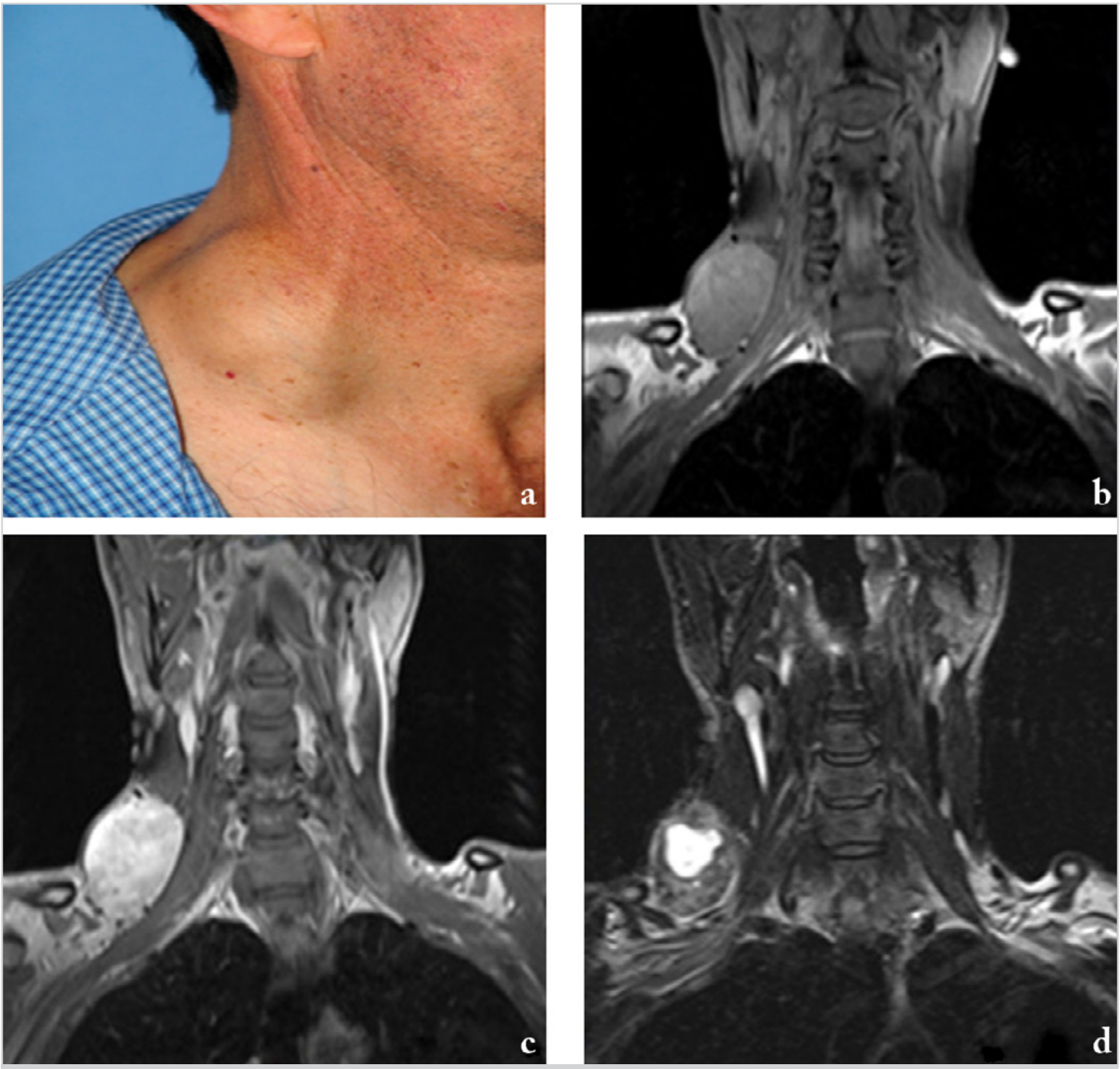

Figure 1. a-d. A soft tissue mass in the right supraclavicular region (a), a supraclavicular solid mass in the precontrast T1-weighted coronal MRI image (b), heterogenesis involvement of supraclavicular solid mass in postcontrast T1-weighted coronal MRI image (c), central hyperintensity in the supraclavicular solid mass in T2-weighted coronal MRI image (d)

are generally localized in the serosal surfaces (1, 2). In addition, their presence has been reported in non-serosal regions such as renal pelvis, periosteum, spinal cord, lung, liver, breast, skin, and head–neck (3, 5). The diagnosis of this rarely observed mesenchymal tumor was established at the rate of 2.8 per 100.000 patients in the records of Mayo Clinic (8). Primarily, it is observed in adults between the ages of 30 and 70 years and at equal rates in both genders (9). SFT is often benign. However, 5–20% of pleural SFT cases and 10% of extra-pleural SFT cases are malignant (7). Metastasis occurs firstly in the lungs and then in the liver and bones.

The first SFT case in the head–neck region was reported by Witkin and Rosai in 1991 (10). In this region, it is mostly observed in the oral cavity and sinonasal area and then in the orbita. Furthermore, SFTs have previously known to have developed in the nasopharynx, parapharyngeal region, larynx, hypopharynx, major salivary glands, thyroid, and meninx (5, 6, 11, 12). Of all cases, only 6% develop in the head–neck region (3, 13).

SFT is generally a slow-growing painless benign mass (7). The symptoms are non-specific and depend on the lesion's size and localization (6). However, it may not present with clinical signs

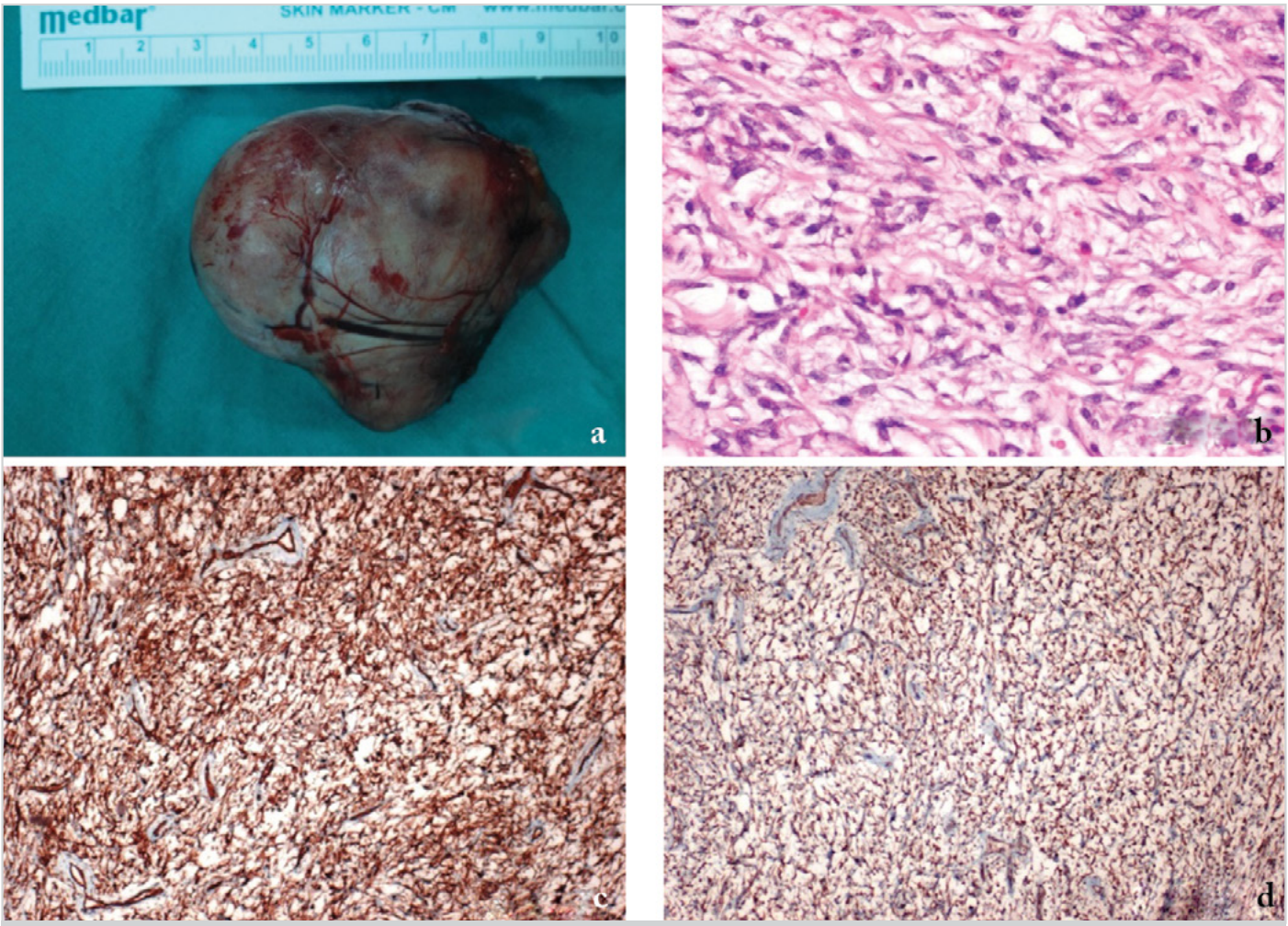

Figure 2. a-d. Well-demarcated, encapsulated solid mass after surgical resection (a), fusiform cells with round/oval nucleus (hematoxylin-eosin staining, 200×) (b), tumoral cells showing positive reaction with CD34 (100×) (c) tumoral cells showing positive reaction with Vimentin (100x) (d)

before reaching a notable size or putting pressure on vital structures (4).

The mean size of SFTs in the head–neck region is 2.6 cm (6). A larger tumor can rarely cause paraneoplastic syndrome. In about 5% of SFTs developing out of the head–neck region, hypoglycemia associated with the release of insulin-like growth factor coexists (14). Fever, chest pain, and osteoarthropathy were also reported to be among the symptoms of SFTs developing out of the head and neck region (5). In literature, hypoglycemia has not been reported as a complication in the head–neck SFTs (6).

Clinical diagnosis of head–neck SFTs is quite difficult. In the differential diagnosis, other soft tissue tumors such as synovial sarcoma, fibrosarcoma, metastatic malignant mesothelioma, solitary myofibroma, hemangiopericytoma, neurofibroma, benign, and malignant nerve sheath tumors should be considered (7).

The most remarkable feature of SFT is that it shows rich staining in computed tomography and MRI, but this finding does not have a specific diagnostic value for the disease. Ganly et al. (7) reported that SFTs displayed a well-demarcated isodense appearance in CT, while they appeared isointense in the precontrast T1-weighted images and greatly isointense in the T2-weighted images in MRI.

The MRI of the patient revealed a well-demarcated, lobulated, and solid mass in the right supraclavicular region, posterior of the sternocleidomastoid muscle, and deep cervical plane. While the supraclavicular solid mass displayed a heterogeneous involvement in the postcontrast T1-weighted MRI, central hyperintensity was found in the T2-weighted MRI.

Pre-operative histopathologic diagnosis is important for deciding the surgical approach and FNAB can help establish the diagnosis. The presence of fusiform cells mixed with collagen in the bloody posterior plane in FNAB can be valuable for the diagnosis of SFT. However, these findings are not specific for SFT (7). In the case presented, no specific diagnosis was made with FNAB.

In most of cases, the diagnosis of SFT is established based on the postoperative immunohistochemical staining characteristics with CD34, factor VIII, vimentin, CD99, and Bcl2 (7,

11). SFT is generally negative for S100 protein, desmin, and cytokeratines (7).

Malignant transformation is rare but it requires wide local excision (12). Furthermore, in patients having undergone incomplete excision, postoperative radiotherapy and chemotherapy can also be appropriate (2, 4, 10). Long-term follow-up is necessary even for patients in whom complete excision was performed because recurrence and metastases can occur after many years (7).

Witkin and Rosai reported that the most important prognostic factor was the tumor being able to be excised completely. Prognosis also depends on the size of tumor, localization, and histologic characteristic (4).

In the case presented, the supraclavicular mass was excised completely with a trans-cervical approach. No tumor was observed in the surgical margins. The post-operative period passed without any problem. Immunohistochemically, the tumor cells were positive for CD34, vimentin, and factor VIII, but negative for S100. Ki-67 proliferative index was low. These findings were consistent with the diagnosis of SFT. Features reminding malignancy potential such as margin infiltration, hypercellularity, nuclear atypia, and high mitotic activity were not found. No finding of the disease was detected in the control examination performed in 6 months postoperatively. The patient was followed up with routine control examinations.

Conclusion

SFT is an asymptomatic benign lesion that develops quite rarely and grows slowly. For histopathological diagnosis, immunohistochemical CD34 positivity is the most valuable finding. The recurrence rates after complete surgical excision are low.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - C.H.U.; Design - C.H.U.; Supervision - C.H.U.; Funding - Y.K.V.; Data Collection and/or Processing - C.H.U., Y.K.V., O.Y.; Analysis and/or Interpretation - C.H.U., Y.K.V., O.Y.; Literature Review - C.H.U., Y.K.V.; Writing - C.H.U.; Critical Review - C.H.U.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

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