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A rare case of solitary fibrous tumor of the temporal region: 7-year-follow-up clinical-radiographic evaluation and literature review

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ABSTRACT

Solitary fibrous tumor is a rare spindle-cell neoplasm of mesenchymal origin. In head and neck region, the tumors present slow-growing masses, often with local compressive symptoms. Although it is generally benign, malignant variants have been identified. The radiological diagnosis of solitary fibrous tumor is usually based on computer tomography and/or magnetic resonance imaging. Microscopically, a solitary fibrous tumor is characteristically a circumscribed neoplasm composed of variably cellular and patternless distributions of bland spindle and ovoid cells within variably collagenous stroma that frequently shows areas of dense hyalinisation, as well as interspersed large branching or "staghorn"- shaped thin-walled vessels. Immunohistochemical staining is very effective to distinguish solitary fibrous tumors from other fibroblastic tumors. Recently, NAB2–STAT6 gene fusion derived from inv12 (q13q13) has been reported as the genetic hallmark of solitary fibrous tumor. Complete local surgical excision appears to be the treatment of choice for solitary fibrous tumor of the head and neck region. Recurrence was reported in 5% of cases. The median recurrence-free interval was 36.5 months. We report the case of a solitary fibrous tumor of the temporal region, surgically excised and with no clinical and/or radiological signs of recurrence after 7 years of follow-up.

1. Introduction

Solitary fibrous tumor (SFT) is a rare spindle-cell neoplasm of mesenchymal origin. It was first described by Klemperer and Rabin in 1931 [1]. Before the widespread acceptance of the term "solitary fibrous tumor," pleural SFT had been referred to by a number of other terms, including fibrous mesothelioma, benign mesothelioma, localized mesothelioma, subpleural fibroma, and localized fibrous tumor of the pleura, to distinguish it from the clinically more aggressive mesothelioma. Another synonymous term is hemangiopericytoma (HPC), described first in 1942 and originally thought to be a neoplasm arising from perivascular smooth muscle cells [2]. For many years, HPC and SFT were considered as different entities. With the development of immunohistochemistry techniques and sophisticated cytogenetic analyses, it was shown that SFT have features and an immunohistochemical pattern that makes it almost indistinguishable from HPC. It has led to their unification as SFT in contemporary WHO classification. Initially, SFTs were thought to arise only in the pleural cavity, but other primary sites have been reported. In 1991, Witkin and Rosai reported the first

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recognized case within the head and neck region [3]. Currently, it is estimated that 6–18% of all SFT arise within the head and neck region, representing approximately one quarter of all extrathoracic SFT [4].

2. Case presentation

A 35-year-old man was referred to our Department in 2014 for the evaluation of a painless nodule in the left temporal region. On local examination, a relatively firm, well circumscribed, dome shaped mass with normal color was found in correspondence of the left temporal muscle.

Ultrasonography showed the presence of a solid, hypoechoic, oval nodule, with clear contours, 1,8cm in diameter, with moderate vascularization and located within a muscular structure. Arterial doppler analysis of the main vascular peduncle showed a regular systolic-diastolic modulation compatible with the presence of an anarchic vascularity (Fig. 1).

MRI revealed the presence of a solid nodular formation localized in the context of the superficial bundles of the left temporal muscle, with a homogeneous signal, not modifying in fat-sat acquisitions and without liquid component inside it. The margins of the nodule appeared well delimited and after the administration of the contrast medium there was a lively, rapid and complete enhancement (Fig. 2).

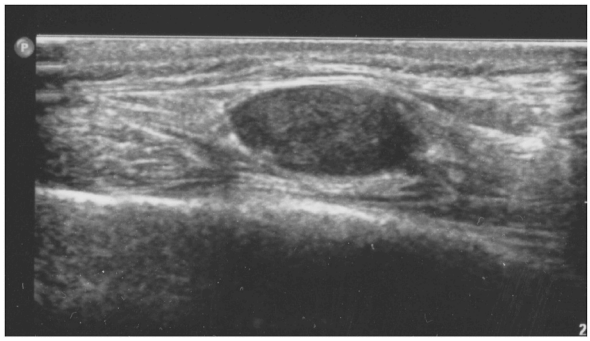

Fig. 1. Ultrasonography showed the presence of a solid, hypoechoic, oval nodule, with clear contours, 1,8cm in diameter, with moderate vascularization and located within a muscular structure.

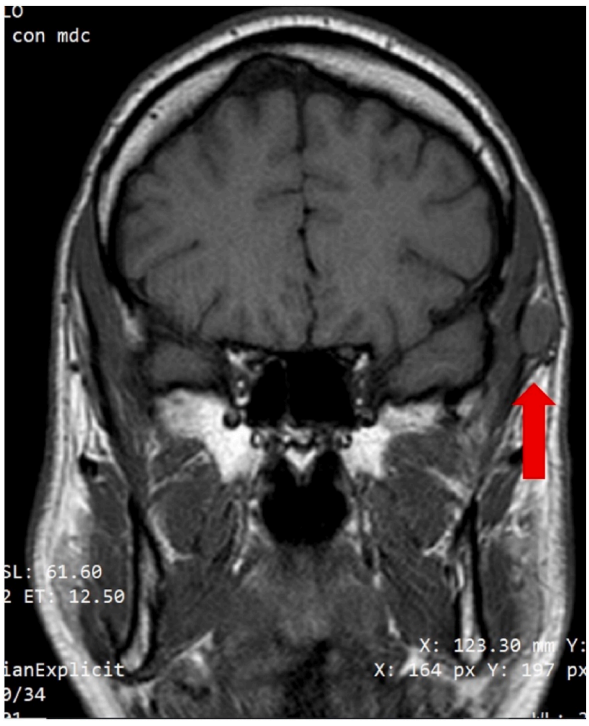

Fig. 2. Pre-operative MRI revealed the presence of a solid nodular formation localized in the context of the superficial bundles of the left temporal muscle.

Surgery was performed under general anesthesia. A pre-auricular skin incision was made extended to the left temporal region; a skin flap was prepared to highlight the site of neoformation. Then the incision of the temporal fascia and the removal of the rounded lesion that was included in the structure of the temporal muscle were performed (Figs. 3–4).

No complications were observed. A regular post-operative course was noted.

The histological examination showed, macroscopically, a grayish tesoelastic nodule with a compact and uniform cutting surface of 2x1.8 cm. Microscopically, was observed a benign spindle cell mesenchymal neoplasm with hypercellulated areas, deposits of collagen material and "staghorn" vessels with hemangiopericytoma-like pattern, in the absence of significant mitotic activity and/or necrosis.

7-year-follow-up evaluation showed no signs of recurrence of the lesion; the clinical examination was normal. MRI confirmed the absence of recurrence (Fig. 5).

3. Discussion

In head and neck region, the tumors present slow-growing masses, often with local compressive symptoms. According to the systematic review published by Stanisce et al. [4], evaluating 343 cases reported in the literature from 1991 to 2019, SFT in the head and neck demonstrate no predilection for gender. The median age of presentation was 51 years old (range 8 months–94 years). The oral cavity (31%) was the most commonly reported location, followed by sinonasal (16%) and neck (12%) regions. In our case, the lesion occurred in temporal region. According to the evidence of the literature, it represents an event rarely described. Regarding symptoms, nearly 62% of valid cases (240 cases) reported a chief complaint of a new mass or localized swelling. Other symptoms were linked to the anatomical location of the tumors: dysphonia, nasal obstruction and epistaxis. Pain was described in only 38 cases (11%).

Head and neck SFTs were described macroscopically as circumscribed, solid, indurated, fibrous lesions that were white, tan, or gray in color. Microscopically, an SFT is characteristically a circumscribed neoplasm composed of variably cellular and patternless distributions of bland spindle and ovoid cells within variably collagenous stroma that frequently shows areas of dense hyalinisation, as well as interspersed large branching or "staghorn"- shaped thin-walled vessels [5]. The proportion of cellular versus stromal components, the presence of adipocytic (fat-forming variant) and multinucleated cells, i.e. a giant-cell angiofibroma-like appearance, and variations in nuclear atypia and mitotic activity result in a broad spectrum of SFTs in histomorphology and tumor behavior [6].

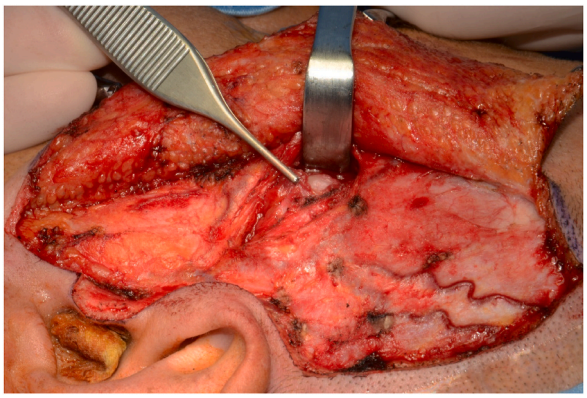

Fig. 3. Surgical excision: a skin flap was prepared to highlight the site of neoformation, then the incision of the temporal fascia and the removal of the rounded lesion that was included in the structure of the temporal muscle were performed.

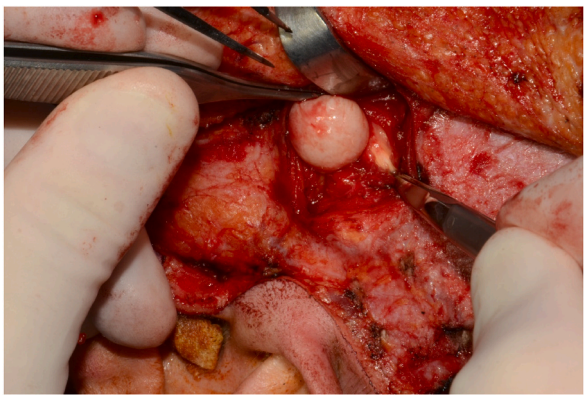

Fig. 4. Surgical excision: a skin flap was prepared to highlight the site of neoformation, then the incision of the temporal fascia and the removal of the rounded lesion that was included in the structure of the temporal muscle were performed.

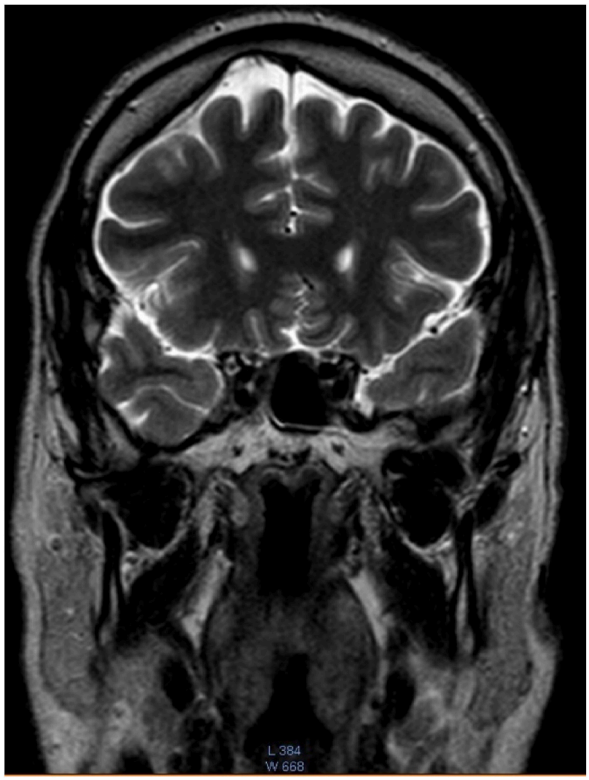

Fig. 5. 7-year-follow-up MRI revealed the absence of recurrence.

SFTs are characterized by a wide histological variability. Sometimes it can be difficult to distinguish from other benign and malignant tumors that have similar histological features. Immunohistochemical staining is very effective to distinguish SFTs from other fibroblastic tumors. SFTs show positive reactivity for CD34, CD99, Bcl-2 and EMA, while desmin, cytokeratin and S-100 protein are usually negative. On immunohistochemical staining, CD34 has been considered the most reliable marker for the diagnosis of SFT. However, CD34 expression is also common in other tumors such as soft tissue perineuroma, dermatofibrosarcoma protuberans and spindle cell lipoma, which are included in the differential diagnosis of SFT [5].

Recently, NAB2–STAT6 gene fusion derived from inv12 (q13q13) has been reported as the genetic hallmark of SFT. NAB2-STAT6 protein is a transcription factor. It serves as a driver for tumor growth by activating NAB2 target genes, which have an early growth response-binding domain fused to the activation domain of STAT6. Overexpression of NAB2-STAT6 induces proliferation in cultured cells and activates the expression of early growth response-regulated genes [7]. The breakpoints of NAB2 and STAT6 are highly variable and may yield diverse NAB2–STAT6 fusion types of complex exon compositions. Intriguingly, the variability in NAB2–STAT6 fusion variants has recently been shown to be associated with anatomical site, age, histology, and, possibly, the clinical behavior of SFTs originating in a broad range of primary sites [6].

The radiological diagnosis of SFT is usually based on computer tomography (CT) and/or magnetic resonance imaging (MRI). On CT, SFT is isodense to muscle with hypodense areas, which may represent myxoid structures. It shows strong enhancement due to its rich vascularity, occasional calcification and necrosis and remodeling of the adjacent bone. On MRI, SFTs show hypointense or isointense signal on T1WI and T2WI related to the rich collagenous component. High T2 signal intensity is also seen in hypercellular region of the tumor, or secondary to hemorrhage, or cystic or myxoid degeneration [8].

SFTs are known to have unpredictable clinical behavior, and the natural course can vary from a very indolent, localized tumor to a presentation with early metastasis and aggressive systemic spread.

Although no universal definition for defining a malignant SFT is accepted, several histopathological criteria have been proposed. A first risk stratification was published in 1989 by England et al. based on the following histopathological criteria: high cellularity and mitotic activity (>4 mitotic figures per 10 high-power microscopic fields (HPF)), cellular pleomorphism, presence of hemorrhage and necrosis [9]. Refining these criteria for extrathoracic SFT, Demicco et al. demonstrated that age $\geq$55 years old, tumors $\geq$10 cm, positive surgical margins, the presence of histological necrosis, and mitoses $\geq$4/10 HPF predicted for local recurrence and metastasis [10]. In the head and neck region, although it is generally benign, malignant variants have been identified. Stanisce et al. evaluated definite details of local invasion on pre-operative imaging or gross examination during surgical intervention. Local invasion into bone, vasculature and nerve was noted in 13% of cases. Bony erosion was noted in 10% of cases, vascular invasion was noted in 2% of cases and peri-neural involvement was reported in 1% of cases [4]. In our case, there were no evidence of mitotic activity or necrosis, tumor size was widely less than 10cm and pre-operative imaging and gross examination did not provide signs of local invasion.

Complete local surgical excision appears to be the treatment of choice for SFTs of the head and neck region. Adjuvant therapy, including post-operative radiation therapy and pre-operative embolization treatment, was utilized in 7% of cases. Recurrence was reported in 5% of cases. The median recurrence-free interval was 36.5 months. The presence of positive margins was associated with shorter recurrence-free survival.

In our case, SFT was completely excised without adjuvant therapy and we found no recurrence of the lesion after 7 years of follow-up.

Consent statement

Formal consent was not elaborated and obtained because the figures used are completely anonymized and without any marks that can make the patient identificable.

Funding

The authors did not receive support from any organization for the submitted work.

Proof of consent

Formal consent was not elaborated and obtained because the figures used are completely anonymized and without any marks that can make the patient identificable.

Authors' contributions

Prof. Ugo Consolo, Prof. Attilio Carlo Salgarelli and Prof. Pierantonio Bellini performed surgery. Dr Francesco Diamante performed clinical and radiographic follow-up. All authors reviewed the results and approved the final version of the manuscript. Ugo Consolo: conceptualization, investigation (performed surgery), supervision Francesco Diamante: data curation, writing – original draft. Attilio Carlo Salgarelli: investigation (performed surgery), Pierantonio Bellini: writing – review & editing, supervision.

Declaration of competing interest

The authors declare no conflict of interest.

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