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Case Report

Recurrent fibrous solitary tumor of the parotid gland with satellite location: imaging, clinical and histological findings of rare entity. ☆☆☆★

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ABSTRACT

Fibrous Solitary Tumors are infrequent neoplasms originating from mesenchymal tissues, most commonly arising from the visceral pleura and frequently exhibiting a benign behavior. Extra-pleural localization is unusual and the site of origin of these tumors from the parenchyma of the parotid gland is considered extremely rare. We report the case of a 66-years old woman with non-painful slow-growing left latero-cervical mass, who underwent a gadolinium-enhanced Magnetic Resonance Imaging showing a mass originating from the deep lobe of the parotid gland extending into the retro-pharyngeal space. After a total parotidectomy with tumor excision, a diagnosis of histologically proven fibrous solitary tumor of the parotid gland was made. Two years later, CT scan showed post-operative recurrence and further satellite localization in the neck, distant from the initial mass. We performed a literature review of the published similar cases, in order to clinicopathological and imaging features of this rare entity.

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Introduction

Solitary fibrous tumors (SFT) are ubiquitous rare spindle cell neoplasms, most commonly arising from the pleura. Pleural SFT were for the first time described in 1931 by Klemperer and Rabin [1], which assumed the mesothelial origin of the neoplastic cells. Subsequent immunohistochemical and ultrastructural studies have suggested that SFTs are most likely derived from adult mesenchymal stem cells rather than mesothelium [2]. Extra-pleural localization of SFT in the Head and Neck region represent only the 6% of all reported cases [3]. SFT arising from the parotid gland parenchyma are even rarer, with only 39 cases (including the current one) reported in literature since 1995, year in which parotid gland localization was reported for the first time by Hanau and Miettinen [4].

Most of the reported parotid gland SFTs (PG-SFTs) show a benign behavior, with local recurrence reported only in two cases [5,6] and distant pulmonary metastasis in only one case [7].

Due to the non-specificity of symptoms and imaging features, the diagnosis of SFT arising from the parotid gland is very difficult and requires histological demonstration.

In the following sections we report the clinical, histological and imaging findings of a case of recurrent PG-SFT with a further satellite localization in the neck, distant from the initial epicenter of the mass. Furthermore, we provide a literature review of the reported cases with the aim of describing the critical characteristics of this rare entity.

Case report

A 66 years-old woman was referred to our institution because of a slow-growing painless left latero-cervical mass. On physical examination in the left parotid region an elastic, mobile, well demarcated and tender mass with no signs of inflammation of the overlying skin was noticed. Facial nerve deficiency and systemic symptoms were absent.

The patient performed a contrast enhanced Magnetic Resonance Imaging (MRI) which revealed the presence in the left parotid space of a voluminous mass with polylobulated sharp margins arising from the deep lobe of the parotid gland and extending medially in the parapharyngeal space and anteriorly in the masticatory space, spreading the fibers of the ipsilateral pterygoid muscles. The mass presented an iso-intense signal in T1-weighted images and a slight hyperintense signal in T2w and in STIR sequences, with some focal areas of less hyperintense signal (Fig. 1). After the administration of gadolinium contrast medium, the lesion presented early, strong and almost homogeneous enhancement in T1 fat saturated sequences (Fig. 2).

A parotid benign lesion was suspected and the patient underwent surgical procedure.

A total parotidectomy with excision of the parapharyngeal extension of the tumor was performed. Post-surgical recovery was uneventfully and subsequent post-operative ultrasound evaluations were normal.

Gross examination of the resected specimen revealed the presence of a multinodular whitish mass, measuring $5 \times 3 \times 2$ cm originating from the deep lobe of the parotid gland.

The histological analysis reported a neoplastic proliferation with different cellular density areas on a fibrous collagen rich background, ranging from areas with high cellularity and less stroma to areas with a denser fibrous stroma in which numerous vascular channels with thin walls, tortuous appearance and open lumens were identified.

Neoplastic elements appeared monomorphic, of medium size, with only slight atypia, with a single round-oval vesicular nucleus and a small nucleolus often not evident, undefined cytoplasm edges and sometimes perivascular disposition, in a hemangiopericytoma-like fashion (Fig. 3A-B).

No areas of necrosis were evident. Mitoses were noted, with 2 per 10 at high power fields.

At an immunohistochemical evaluation, the pathological cells showed diffuse cytoplasmic reactivity with CD34 (Fig. 3B) and vimentin, focal positivity with EMA and Bcl-2, tenuous positivity with CD99, but were negative with cytokeratin, S100, HMB45, desmin, actin, CD117 and CD31; A Ki67 proliferation index of 20-30% was found. Since not routinely tested, nuclear STAT6 expression levels were not evaluated.

Based on the histologic appearance and immunohistochemistry profile, a diagnosis of PG-SFT was made.

After an asymptomatic period of 2-years, the patient returned to our institution for the reappearance of facial swelling in the parotid region and in the neck, on the same previously affected side.

A CT scan (Optima 660, GE Medical Systems, volumetric acquisition, thickness 0,625, 120 Kvp) performed after iodinated contrast medium administration documented the surgical results of a total parotidectomy with the presence of a contrast enhancing polylobulated mass with sharp margins in the left retropharyngeal and masticatory spaces adjacent to the ipsilateral collapsed internal jugular vein and producing a partial compression on the internal carotid artery (Fig. 4).

Moreover, a distant oval mass of 2,5 cm of diameter located above the fibers of the ipsilateral sternocleidomastoid muscle, presenting the same density and enhancing characteristics of the parapharyngeal mass, was identified (Fig. 5). The excision of this recently appeared, more superficial latero-cervical nodular localization and subsequent histological evaluation revealed a morphologic and immunohistochemical result compatible with a new distant satellite localization of SFT.

At the time this report was written, the patient was advised to undergo an oncological consultation in order to decide whether to have an adjuvant chemotherapy before the surgical retreatment.

Discussion

PG-SFTs are rare fibrous neoplasms thought to arise from the mesenchymal cells of the parenchymal parotid gland, with only one reported case of tumor originating from the Stensen's duct [8].

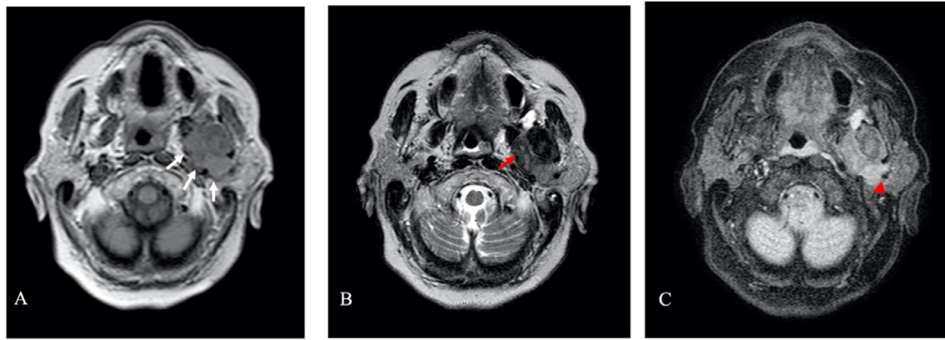


Figure 1 – Axial TSE T1-weighted image (A) showing an iso-intense mass (white arrows) with sharp margins originating from the deep lobe of the left parotid gland and extending in the ipsilateral parapharyngeal and masticatory spaces. (B) Axial TSE T2-weighted image showing an area of relative hypointensity in the mass (red arrow) probably due to the presence of fibrous tissue. Axial STIR sequence (C) which documents the continuity (arrowhead) between the deep lobe of the gland and the tumor. (Color version of figure is available online)

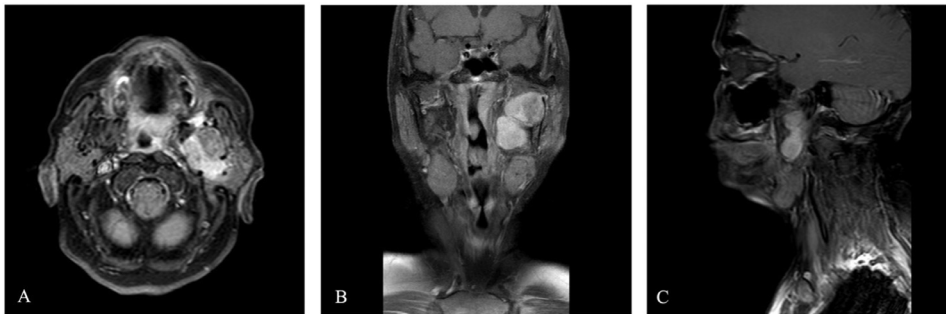


Figure 2 – Axial (A), coronal (B) and sagittal (C) T1-weighted fat saturated sequences post administration of gadolinium showing an almost homogeneous enhancement of the mass, which presents sharp, polylobulated margins and determines a mild compression on the oropharyngeal lumen.

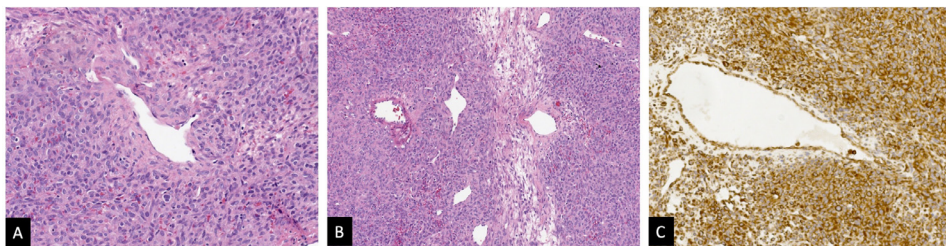


Figure 3 – (A) Fibrous solitary tumor of the parotid gland: hematoxylin-eosin stain on 20x magnification showing a neoplastic proliferation with an “hemangiocytopoma-like” pattern. (B) Hematoxylin eosin stain on a 10x magnification showing an irregular vascular pattern of the tumor. (C) CD34 immunohistochemical staining showing the strong immunoreactivity of the neoplastic cells.

Symptoms and radiological findings in patients with PG-SFTs are non-specific and consequently diagnosis is challenging.

Most of the patients report a slow-growing painless latero-cervical mass with a wide interval of symptoms duration, ranging from few months to several years.

A case with dysphagia, symptoms of obstructive sleep apnea and altered speech, consequent to compression exerted

from the mass on the pharyngeal structures, has been reported [9].

Despite the intrinsic anatomical relationship between the facial nerve and the parotid gland, functional nerve deficits have never been documented.

No systemic symptoms or lymph nodal involvement have ever been described, even in PG-SFT cases with an aggressive behavior or metastatic disease.

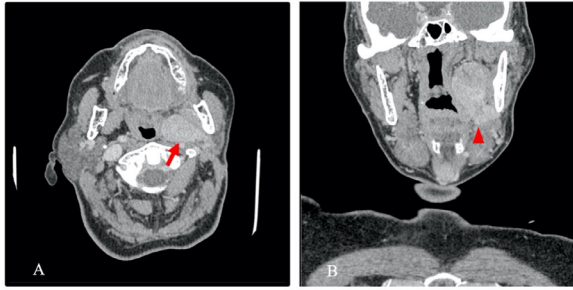


Figure 4 – Axial section (A) and coronal reconstruction (B) of a CT scan acquisition after iodinated contrast medium administration performed 2 years later the diagnosis of PG-SFT revealing the re-appearance in the left para-pharyngeal space (red arrow) of an oval-shaped mass presenting slightly heterogeneous enhancement and extending caudally in the ipsilateral submandibular space (red arrowhead). (Color version of figure ia available online)



Figure 5 – Different coronal reconstruction section of the enhanced CT scan showing the presence of an oval shaped nodule (red arrow) above the fibers of the left sternocleidomastoid muscle, presenting the same density and enhancing characteristics of the parapharyngeal mass, compatible with a 2-years later satellite location of recurrent Solitary Fibrous Tumor of the parotid gland. (Color version of figure ia available online)

PG-SFT has no gender predilection and the mean age of reported patients is 50,6 years (ranging from 11 to 78 years), with only one reported pediatric case in a patient with concomitant neurofibromatosis type I [10].

PG-SFTs frequently exhibit benign behavior and tend not to recur if the tumor is completely excised, anyway aggressive forms with recurrence or pulmonary metastasis have been reported [5–7,11]. In our case we observed the recurrence of the mass in the parapharyngeal space and the appearance of a new nodular latero-cervical location, separate from the initial

site of the mass, which we thought to be a new satellite location.

These tumors are frequently misdiagnosed as pleomorphic adenoma, schwannomas, paragangliomas, hemangiomas or others more common benign lesion occurring in that region [13].

Currently there are no specific imaging biomarkers that allow to reach the correct diagnosis using only radiological investigations. MRI studies can be used to document the presence of the mass, which is generally oval or polylobulated with sharp margins, exerting compression on adjacent structures without clear sign of infiltration.

MRI signal is often iso-hypointense in T1 and mildly hyperintense in T2 with evidence of some areas of relative hypointensity probably related to the presence of focal areas of more fibrous tissue deposition, which has generally a low signal in T2 weighted images. Rarely fibrous T2 hypointense intratumoral septa have been reported [13].

When contrast agent is administered in CT and MR studies, the tumor frequently presents an intense early enhancement, reflecting the high vascular component reported in the histologic studies.

CT scan can be useful to identify involvement of the surrounding bony structures, which has been reported in some aggressive cases, probably due to remodeling and compressive rarefaction rather than real bone destruction [5].

Since clinical and radiological findings are non-specific, histologic evaluation has a fundamental role.

SFT is characterized by fibrous hypocellular areas that alternate with hypercellular areas composed of round-to-spindle cells with a fascicular, storiform arrangement. A distinguishing characteristic of fibrous SFT is the presence of numerous, medium-sized, ramified vessels with thickened and hyalinized walls [17].

The differential diagnosis of SFT involving parotid gland includes a variety of spindle cell neoplasm, and many show similar histological characteristics [18].

However, the immunohistochemical diagnosis has been simplified by the recent detection of STAT6, a very sensitive and specific marker for SFT, which identifies the NAB2-STAT6 fusion product. It is a consequence of an intrachromosomal inversion, *inv12(q13q13)*, resulting in NAB2-STAT6 gene fusion, which exhibits variable breakpoints and drives STAT6 nuclear expression. Nuclear expression of STAT6 has been observed in more than 95% of SFT, whereas low-level of expression is typically seen in other mesenchymal tumors [19].

Moreover, even if the exclusive use of histological features does not reliably predict malignant clinical behavior, some aggressive feature as high mitotic rate ($\geq 4/10$ mitoses at high-power fields), cytological atypia, tumor necrosis, and/or infiltrative margins have been used in addition to clinical and morphological data as prognostic factors to create a risk stratification model in order to predict the malignant course [20].

Only one case of malignant tumor presenting a central area of dedifferentiation has been reported [11].

PG-SFTs treatment is surgical and the excision of a benign formation with negative margins often results in definitive cure. Anyway, since tumor recurrence has been reported even several years after the onset of a SFT, serial

clinical and imaging evaluations need to be performed as follow-up.

In two cases, one of which was a giant high vascular PG-SFT, pre-operative angiography was performed to obtain tumor embolization. [12,16]

Some cases with recurrence or aggressive behavior have been treated with adjuvant radiotherapy [11,14,15]; anyway, since there are only few cases of reported malignant tumors and there are no guidelines regarding this issue, the treatment strategy in these cases is still unclear.

Conclusions

PG-SFT is a rare entity. As the clinical and radiologic features are non-specific, the diagnosis is based on morphological and immunohistochemical analyses. More studies are needed to identify biomarkers allowing to differentiate SFT from other salivary gland and mesenchymal tumors using imaging.

Patients with SFT should undergo serial screening given the possibility of recurrence, although infrequent.

Effective treatment options in case of recurrence or presence of local aggressive signs should be studied in order to create a standardized treatment.

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