CASE STUDY

Primary malignant solitary fibrous tumor/hemangiopericytoma of the parotid gland

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Abstract  Solitary fibrous tumor (SFT) was first described in the pleura by Lietaud in 1767; later in 1870, Wagner described the localized nature of this type of tumor and Klemperer and Rabin classified pleural tumors into two types: diffuse mesotheliomas and localized mesotheliomas. Recent years have seen the redefinition of this neoplasm, due to better technology; it is now proven that this neoplasm may have multiple different extrapleural origins including the head and neck regions. This diversity of locations is related to the particular mesenchymal histogenesis of SFT which allows its development from very unusual sites such as the salivary glands (SGs). In this particular site, this neoplasm is very infrequent and most of reported cases refer to benign disease, with just one case informed so far of primary malignant SFT.

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PALABRAS CLAVE

Tumor fibroso solitario maligno primario/hemangiopericitoma de la glándula parótida

Resumen  El tumor fibroso solitario (TFS) fue primeramente descrito en la pleura por Lietaud en 1767; posteriormente en 1870, Wagner describió la naturaleza localizada de este tipo de tumor y Klemperer y Rabin, en 1931, clasificaron los tumores pleurales en dos tipos: mesoteliomas difusos y mesoteliomas localizados. En los últimos años se ha redefinido esta neoplasia, debido a la mejora de las tecnologías; ahora se ha probado que puede tener múltiples y diferentes orígenes extra-pleurales, incluyendo la región de cabeza y cuello. Esta diversidad de...
Primary malignant solitary fibrous tumor/hemangiopericytoma of the parotid gland

Clinical case

A 78-year-old female, with a 1-year history of a progressively growing, painful, fixed and hard mass in the right parotid region. A head and neck CT scan showed a 39 × 29 mm² parotid mass, extending to the deep lobe without associated adenopathies. A radical parotidectomy was performed and during the surgery an irregular, poorly circumscribed mass compromising the entire gland was observed. The histopathological study identified a hypercellular mesenchymal tumor with a fusiform pattern (Figure 1); it presented 6 atypical mitoses per 10 high-power fields, pleomorphic areas and no areas of necrosis. Immunohistochemistry yielded positive for CD34 and negative for AE1/AE3, EMA, h-caldesmon, desmin, p63, ALK, CD99 and bcl-2; Ki67 was 35% (Figure 2). Diagnosis of malignant solitary fibrous tumor (SFT) was made. On further workup, a chest CT scan revealed multiple pulmonary nodules ranging from 3 to 44 mm, compatible with metastatic disease; these are biopsied, showing similar morphology and immunophenotype to the parotid lesion. After a follow-up of 7 months, the patient is in good condition. Literature search results are depicted in Table 1, and the immunohistochemical panel is described in Table 2.

Discussion

As in other extrapleural locations, SFT of the major SG is very rare. To date only 16 reported cases exist, one malignant. SFT has been reported in diverse sites such as serous membranes like the peritoneum, either parietal or visceral, retroperitoneum, mesentery, omentum, abdominal wall and hernia sacs, and pericardium. Also, other sites of origin are the meninges, spinal cord, oral cavity, mammary gland, gastrointestinal tract, skin, kidney, female genital tract and other 32 locations such as the orbit, nasal cavity, pelvic space, liver, mediastinum, prostate, upper airway, pancreas, heart, conjunctiva, thyroid, bladder, tonsils, carotid sheath and testicle among others.

All of these lesions, in spite of their site of origin, have the same morphological, cytogenetic and immunohistochemistry characteristics as those seen in cases of SFT of the pleura; therefore, they have been established to be the same neoplasm. This fact, and the pathogenic basis of this disease, has just been cleared in recent years by ways of cytogenetic and genetic polymorphism studies. It is now known that SFT is associated with multiple chromosome abnormalities and the presence of break points in chromo-

Figure 1  View of a hypercellular mesenchymal tumor with a fusiform pattern (A); it presented 6 atypical mitoses per 10 high-power fields, pleomorphic areas (sarcomatoid zones) (B, C). The metastasis shows similar morphology and immunophenotype to the parotid lesion (D).
Tumor cells were positive for CD34 (A) and negative for AE1/AE3 (B) and p63 (C); Ki67 was 35% (D).

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Thanks to these techniques it has been determined that numerous neoplasms previously classified as hemangiopericytomas are in fact SFTs.1,3,16,17 In the particular case of SFT of the major SG, it is very useful to have imaging studies, such as CT scans, before surgery, since they will help define the glandular compromise, partial or total, and the invasion or destruction of adjacent structures, which suggest a malignant behavior.18,19

Definitive diagnosis is made through histopathology, which allows the observation of a neoplasm composed of cells similar to fibroblasts, with little or no mitotic activity, distributing in a fasciculate and estoriform pattern along a stroma rich in collagen that resembles a cheloid.1,19 This tumor usually shows positivity for CD34, CD99, vimentin, laminin and type IV collagen; it yields negative results with cytokeratin, EMA, smooth muscle actin and desmin.21,22 However, some cases have shown marker overlapping by expressing cytokeratins.1 One reported to date,6 according to our literature search. This kind of tumor shows very similar cytogenetic features to its benign counterpart. However, some alterations have been observed like the mutation of the platelet-derived growth factor β receptor, which may be involved in its malignant transformation.23 Microscopically, it shows increased cellularity, marked atypia, prominent areas of necrosis and high mitotic activity, therefore, allowing its distinction.6,26 Also, in immunohistochemical analysis it mimics the benign tumor, but as distinctive features it shows greater expression of fibroblast growth factor, P53 and Ki67.1 For its differentiation, several diagnostic criteria have been used, like those enunciated first by England26 in 1989 and posteriorly modified by Chan27 in 1997, which were satisfied in our case. The vast majority of extrapleural SFTs are benign, but some cases have been reported as malignant, like the one informed by Ogawa et al.6 in the SG, which showed neither recurrence nor metastasis and unfortunately demonstrated in the present case. This fact may be attributed to variable presentation of recurrences and metastasis; in recent occasions there exist reports on latency periods up to 30 years after initial diagnosis.1

Differential diagnosis for SFT of the major SG includes myoepithelioma and its malignant variant, myoepithelial carcinoma, which was considered in our case.4,6,28 Other differential diagnoses include myoepithelioma-like pleomorphic adenoma, myoepithelial epithelial carcinoma, peripheral nerve sheath tumors, fibrous histiocytoma, synovial sarcoma, nodular fasciitis, leiomyoma and leiomyosarcoma.

The treatment of choice for SFT from any location is surgical resection of the encapsulated tumor; this is accomplished in SG by removing the tumor along with the entire gland.5,6 In cases with malignant features, follow-up is warranted, similar to cases of malignant SFT of the pleura; it consists of yearly assessment of local recurrence by imaging, and additional exhaustive imaging to assess the presence of metastatic disease.1,6
Table 2  Immunohistochemistry panel used for the study of the present case

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References