CASE STUDY

Solitary Fibrous Tumour of the Submandibular Gland

Tumor fibroso solitario de la glándula submandibular

Vicenç Martínez Vecina, a,∗ Iñaki Zarraonandia, a Montserrat Monzón, b Dolores Ferrer c

a Unitat d’ORL, Hospital Nostra Senyora de Meritxell, Escaldes-Engordany, Andorra
b Servei de Diagnòstic per Imatge, Hospital Nostra Senyora de Meritxell, Escaldes-Engordany, Andorra
c Servei d’Anatomia Patològica, Hospital Nostra Senyora de Meritxell, Escaldes-Engordany, Andorra

Received 26 February 2014; accepted 9 March 2014

Clinical Case

A 43 year-old patient presenting with a right-sided submandibular tumour of 2 months’ evolution. It was hard to the touch and slightly painful. The patient did not report any medical history of interest, known allergies to medicines or toxic habits of note.

ENT examination revealed a right-sided submandibular tumour of about 3 cm, very hard, adhering to the deep layers and slightly painful on pressure and cervical movement. No swollen lymph nodes were palpated and no significant findings during the rest of the ENT examination.

The patient brought an ultrasound reporting "undetermined tumour in right submandibular gland".

The CT scan performed (Fig. 1) revealed a right-sided intraglandular submandibular lesion, with a high contrast uptake, fairly homogeneous and well-defined in appearance, with dimensions of 22 mm × 28 mm × 20 mm and with no adenopathies suspicious of malignancy. The initial imaging diagnosis was of a pleomorphic adenoma, although a more aggressive process could not be ruled out.

Figure 1 Right-sided intraglandular submandibular lesion with a high contrast uptake, fairly homogeneous and well-defined in appearance, with dimensions of 22 mm × 28 mm × 20 mm and with no adenopathies suspicious of malignancy.

Direct surgical excision was planned without undertaking any other diagnostic tests (ultrasound, FNAP), given the patient’s young age, the rapid growth of the tumour, and because the results of further tests would not alter the diagnostic-therapeutic approach of submaxillectomy.

The right submandibular gland was excised using the Hayes-Martin technique, without incident, as the lesion was purely intraglandular, with no involvement of the capsule or adherences which would impede surgery.
Solitary Fibrous Tumour of the Submandibular Gland

Figure 2  (A) The normal salivary gland can be seen on the left, on the right a fusocellular tumour that is well-delimited but not encapsulated, homogeneous, densely cellular with fusiform cells with clear cytoplasm and cigarette-shaped nuclei, mixed with dense collagen fibres (haematoxylin–eosin 40×). (B) The normal salivary gland can be seen on the left. On the right is an SFT that is clearly Bcl2 marker positive (immunohistochemistry, Bcl2).

The definitive anatomo-pathological study showed a fusocellular tumour that was well-delimited but not encapsulated, measuring 26 mm×24 mm×20 mm, growing from the salivary gland stroma, with a normal base. It was homogeneous, densely cellular with fusiform cells with clear cytoplasm and cigarette-shaped nuclei, mixed with dense collagen fibres (Fig. 2A), and with a mitotic index of 6–8/10 high-power fields. No necrosis. Tumour-free surgical margins.

The immunohistochemistry study showed that the tumour cells expressed CD34 and Bcl-2 (Fig. 2B), and were negative for other markers of histological cell lines: protein S-100, actin and desmin, CD99, cytokeratines and C-kit (CD117). The Ki-67 proliferation factor was 5.10%.

The definitive diagnosis was "Solitary fibrous tumour of the right submandibular gland, likely to behave aggressively (Ki-67 5%–10%)".

The patient was asymptomatic and disease-free after a follow-up period of 12 months.

Discussion

Solitary fibrous tumours (SFT) are rare, described in 1931 by Klemperer and Rabin as a variant of benign pleural mesothelioma.

The histogenesis of the tumours has been the subject of debate for years. Traditionally considered as mesothelial in origin (Stout and Murray; Briselli et al.), their mesenchymal origin was confirmed as late as 1987, thanks to the development of immunohistochemical and ultrastructural techniques (El Naggar et al.); supported by tumours with identical anatomical and pathological characteristics being found in other locations where there is no mesothelial tissue.

Over the last 25 years, cases of SFT in almost all locations have been published. In the head and neck they have been described at sinonasal level, in the oral cavity, the major salivary glands (especially the parotid), the thyroid, pharynx and larynx.

SFT that are located extrapleurally have traditionally been considered benign tumours. Subsequent reviews have demonstrated that around 10% of these tumours can behave malignantly.

The main factor determining the prognosis of mesenchymal tumours is their origin. Histologically, for differential diagnoses of SFT, there are numerous soft tissue tumours, both benign and malignant (fibrous histiocytoma, neurofibroma, neurilemmoma, myofibroma, leiomyoma, fibroma, etc.), that should be considered.

Differential diagnosis can be made by means of immunohistochemical study. SFT are defined, wherever their anatomical location, by being positive to CD34, Bcl2, CD99 and vimentin, and negative to S-100, actin, desmin, factor VIII, specific muscular actin, cytokeratins, epithelial membrane antigen (EMA) and CD68. Haemangiopericytoma can pose difficulties in differential diagnosis as it also expresses CD34.

There is no unanimity in histological criteria to predict whether an SFT will behave aggressively. The most accepted criteria are: hypercellularity, nuclear atypia, more than 5 mitoses per 10 high-power fields and tumour necrosis.

Of particular note, at the level of the major salivary glands, is the metaanalysis of Bauer et al., in which they study the 22 cases of SFT located in the parotid gland published between 1960 and 2011. Of these 22 parotid gland cases, 3 were considered histologically malignant. The mean age of presentation was 51.2 for the men and 49.5 for the women, with no gender preference (11 male cases and 11 female cases); these figures are very similar to SFTs in all other locations. They present clinically as tumours that are slow-growing, hard, well-differentiated, and mildly painful and of variable size. All of the cases were
treated with surgery, complemented with radiotherapy in 2 of the malignant cases; of these 2 cases only one is still alive and the disease persists.

In our search (Medline, keywords solitary fibrous tumour, salivary glands, submandibular gland, sublingual gland) over the same period we found 4 cases with submandibular locations\(^7\)\(^-\)\(^10\) and 2 sublingual.\(^1\)\(^1\)

All the cases with submandibular or sublingual locations were considered histologically benign, were treated with surgery and were disease-free (although with relatively short follow-up periods).

In the case presented it was decided not to give complementary treatment other than surgery because the resection was considered to be completely satisfactory, although the patient underwent exactly the same systematic monitoring as any other head and neck oncology patient.

Conclusion

SFT of the major salivary glands is a rare tumour, which is difficult to diagnose, but very characteristic histologically. Because there are cases which behave very aggressively locally at parotid level, it is advisable to ensure that patients diagnosed with this tumour located in any major salivary gland are regularly and frequently monitored.

Conflict of Interests

The authors have no conflict of interests to declare.

References