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Case Report

Parotid swelling revealing an unusual pathological state: HIV and ewing sarcoma

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ABSTRACT

Swelling of the parotid region is a frequent reason for consultation and the first clinical sign of both benign and malignant parotid tumors.

These parotid tumors represent 80 % of tumors of the main salivary glands. They present a highly variable clinical and radiological picture and are known to have a wide histological diversity. Some histological types are rare entities found in the parotid gland, such as Ewing's sarcoma and lymphoepithelial cyst, the latter of which is often associated with HIV co-morbidity, and therefore requires careful and adequate therapeutic attitudes and strategies.

We report two cases of parotid swelling revealing unusual pathologies: HIV and Ewing's sarcoma.

1. Introduction

Parotid swellings are a frequent reason for consultation and the primary means of discovery of parotid tumors [1]. Parotid tumors account for 80 % of tumors of the main salivary glands. These tumors are characterised by great histological diversity, with benign forms predominating, led by the pleomorphic adenoma [2,3]. Although the diagnostic approach to parotid tumors has benefited greatly from magnetic resonance imaging (MRI) and fine-needle aspiration, it is the pathological examination of the surgical specimen that provides a definitive diagnosis [4]. We report two cases of parotid swellings revealing unusual pathologies: HIV and Ewing's Sarcoma.

2. Case reports

This case has been reported in line with the SCARE criteria [5].

2.1. Case 1

A 43-year-old female patient, with a history of right mastectomy for breast cancer in 2015 completed by radiotherapy + chemotherapy sessions stopped in August 2016, and followed by hormonal treatment (Tamoxifen 1cp/d), who presented to our maxillofacial surgery department in November 2020 for a left parotid swelling evolving for 3 months. Clinical examination revealed a parotid mass of approximately 4 cm in greatest axis, with normal skin appearance, firm to palpation, painless, mobile superficially and deeply, without facial paralysis, with moderate trismus. There was also a homolateral, centimetric, mobile cervical jugulocarotid adenopathy with no inflammatory signs opposite. An examination of Stenon's duct orifice showed no inflammation or bloody discharge, and otoscopic examination revealed no evidence of invasion of the external auditory canal.

Facial MRI revealed a mildly limited tissue process in the left parotid gland with poly-lobed contours in the T1 isosignal and T2 isosignal, which was heterogeneously enhanced after injection of contrast material [Fig. 1], with thickening and infiltration of the opposing soft tissues in contact with the medial pterygoid muscle and the external jugular vein, with no damage to the mandibular bone on further scanning.

After obtaining the patient's consent, she underwent a total parotidectomy with preservation of the facial nerve.

Pathological study of the surgical specimen concluded to Ewing sarcoma parotid with positivity of CD99, cytokeratin AE1AE3 and synaptophysin. The patient was subsequently referred to oncology for adjuvant radiotherapy and then followed up after discharge on an

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outpatient basis at a rate of twice a month for the first two months and then once a month for one year.

2.2. Case 2

A 12-year-old girl was admitted to our maxillofacial surgery department for bilateral parotid swelling that had been progressively evolving since the age of 3 years (Fig. 2). She was the last of two children from a non-consanguineous marriage, with a well-monitored pregnancy carried to term and a normal vaginal delivery. Her immunisation status was up to date according to the national immunisation programme, with good psychomotor development. Her family history included a mother who died of a hepatic hydatid cyst. Clinical examination revealed swellings of both parotid regions, without trismus, nor any notion of recurrent otitis with preservation of general condition. Palpation of the parotid region revealed on the right a bilobed subcutaneous mass of soft consistency, painless, mobile with the superficial and deep planes, about 5 cm long, with healthy skin opposite, and on the left a moderately tender mass, of soft consistency, slightly mobile, with healthy skin opposite. Saliva was normal in appearance. Examination of the lymph nodes revealed multiple painful, mobile cervical adenopathies in the superficial and deep planes, the largest of which measured 1.5 cm in length. The radiological work-up, including ultrasound, showed bilateral parotidomgaly with multiple intraparotid cysts with fine echogenic content and vascularized septa, associated with bilateral subangulomandibular and jugulocarotid lymph nodes. Magnetic resonance imaging (MRI) of the parotid glands showed an enlarged parotid gland with lobular contours and multi-localised cystic formations involving superficial and deep lobes, with a pure liquid signal, the partitions of which are of intermediate signal on T1 and T2 sequences, enhanced after gadolinium injection. After parental consent, a superficial parotidectomy was performed under general anaesthesia. Intraoperatively, the lesions were cystic, measuring approximately 40 mm and 10 mm in diameter. Histopathological analysis of these cysts described a salivary parenchyma, a cystic formation with a regular squamous mucosa without atypia, on a chorion rich in lymphocytes which are organised in lymphoid nodules, without tumor proliferation and concluding to benign lymphoepithelial cysts. HIV serology was performed on the child who was HIV positive. The parents' and his brother's serology was negative.

3. Discussion

Salivary gland tumors are rare, accounting for 0.2 % of all tumors. The parotid gland is the most affected gland in 80 % [2,3]. The incidence of parotid tumors has increased in the 5th and 6th decade [17]. However, the study of parotid tumors is of great interest due to the various paraclinical investigation possibilities and especially the great histological diversity [2,3]. Benign tumors remain dominant in all series of parotid tumors [6–8], and pleomorphic adenoma is the most frequent histological type [2,3]. Malignant pathology of the parotid gland is often underestimated, their incidence varying between 12 % and 39 % of parotid tumors [5].

Clinically, these parotid tumors are discovered by a parotid swelling, which remains the most frequent reason for consultation; the tumor may develop in the gland or in its anatomical extensions, which may lead to a misdiagnosis [1].

Clinical examination can sometimes reveal signs strongly suggestive of malignancy such as pain, paresis or facial paralysis, tumor fixity, rapid development or the association of satellite adenopathies [9,10]. These signs are inconstant and are only reported in 10–45 % of cases of malignant tumors [11,12]. Long-standing swelling and slow progression are not always synonymous with benignity. About two thirds of malignant parotid tumors have a benign clinical presentation [13].

This difficulty in clinical diagnosis justifies the use of additional examinations to provide a sound diagnostic approach for better therapeutic management.

Imaging, particularly MRI, provides excellent anatomical resolution and reliable information on the nature of the tumor, and enables tissue lesions to be distinguished from cystic lesions and contiguous lesions to be eliminated [13]; as in our study, where it revealed a tissue character in the first case and a cystic character in the second. However, it is the histological examination that gives the definitive diagnosis of parotid tumors [4].

In our study, the two cases consulted in our department for parotid swellings, with an acute evolution for the first case and a chronic evolution for the second case, there was no facial palsy in both cases. After paraclinical investigation, notably MRI, they both underwent parotidectomy, followed by anatomopathological examination of the surgical parts for diagnostic purposes.

In the first case, the histological diagnosis was in favor of Ewing's sarcoma and in the second case in favor of lymphoepithelial cysts.

Extra-skeletal Ewing's sarcoma is known to be a rare malignant tumor of mesenchymal cell origin with histological features similar to those of Ewing's sarcoma of bone. It is characterised by its locoregional aggressiveness and high metastatic potential. However, its cervico-facial localization is very rare and few cases have been published in the literature. The clinical manifestations of this sarcoma in the parotid region are nonspecific and include rapidly progressive swelling of the parotid region, trismus, locoregional pain, peripheral facial paralysis

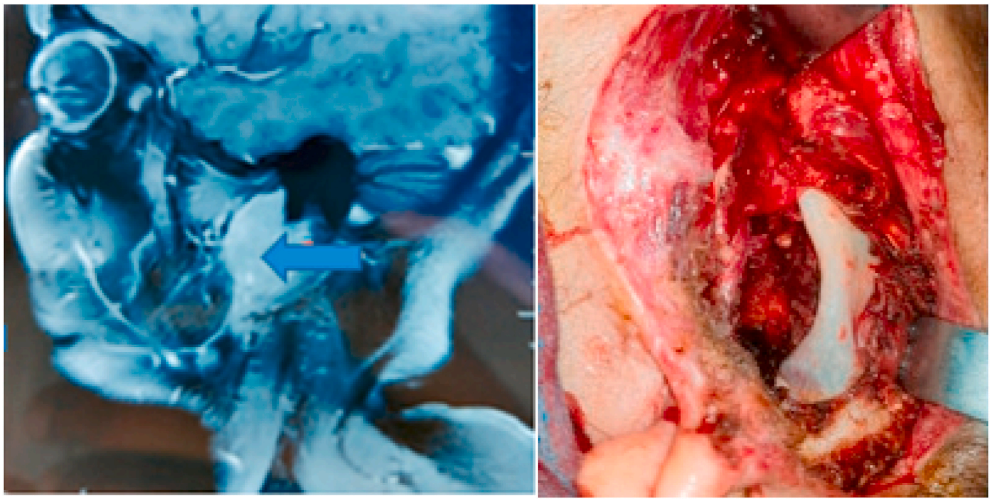

Fig. 1. Facial MRI AND Surgical field after left total parotidectomy [15].

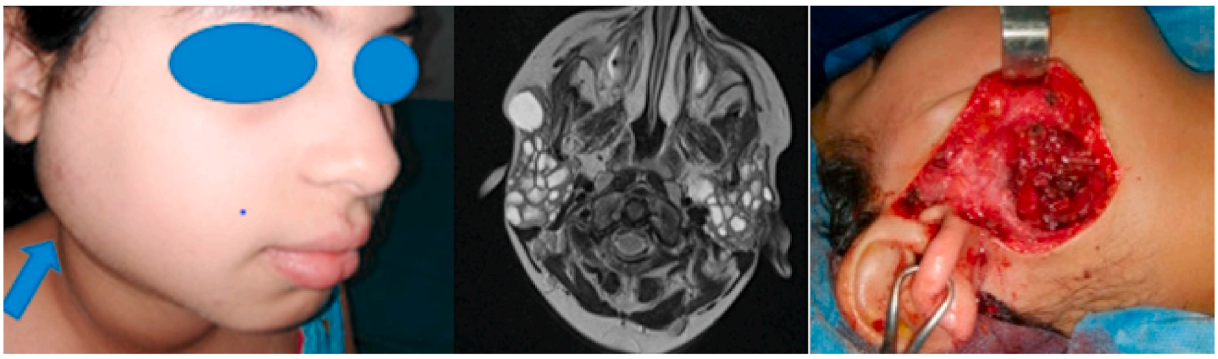

Fig. 2. Patient picture, axial MRI image and intraoperative image of the parotid area.

and homolateral cervical lymphadenopathy. Imaging, particularly high-resolution CT and MRI, is useful in the diagnosis and evaluation of extension, although the features are not specific [14]. However, the definitive diagnosis relies on histology to demonstrate a small round cell tumor with a positive marker CD99/MIC2 in immunohistochemistry [14], in our case the immunohistochemical study was positive for CD99, cytokeratin AE1AE3, and synaptophysin.

Management was completed after pathological results were obtained with adjuvant radiotherapy.

Lymphoepithelial cysts are rare and benign lesions consisting of one or more cysts of the salivary glands or neck regions. Their parotid location occurs in 3–6% of HIV-positive patients [15]. They may serve as the first clinical manifestation of the virus.

Clinically, patients present with chronic parotid swelling that is often bilateral, painless, and sometimes associated with xerostomia and polyadenopathy, indistinguishable from other parotid tumor pathologies. These cysts do not invade surrounding structures and have no malignant potential, but may cause significant local disfigurement. They rarely cause facial paralysis and xerostomia [16].

Non-invasive diagnostic modalities include ultrasound, CT and MRI, which can detect several thin-walled cysts with diffuse neck lymph nodes. And histopathologically, lymphoepithelial cysts usually consist of several benign epithelial cysts with dense lymphoid tissue [16].

The origin of the HIV infection in our case remained unknown, as the serology of the parents and siblings was negative. The hypotheses of rape or sexual abuse were evoked, but quickly dismissed in the face of an interrogation by a child psychiatrist and a normal gynaecological examination. The child, after evaluation of her illness, was put on antiretroviral combination treatment, with periodic follow-up every 3 months.

Parotid swelling is the first clinical sign of a parotid tumor, both benign and malignant. Given the close relationship between the presence of lymphoepithelial cysts and HIV infection, it is important to consider serological screening for HIV once the diagnosis is evoked when we are in the presence of multiple intraparotid cysts objectified on imaging, particularly MRI. This attitude will allow adequate surgical precautions to be taken, both for the surgeons and the entire hospital care team on the one hand, and for the parents, siblings and the patient's entourage on the other. But also in front of these parotid swellings, to think of very rare entities like Ewing's sarcoma, whose parotid localization is not often evoked, whereas they are known to have an aggressive locoregional character and a strong metastatic potential.

4. Conclusion

HIV and Ewing's sarcoma in the parotid region are diseases that are rarely considered in case of parotid swelling, sometimes even age can be a concealing element. However, these pathologies must be envisaged, in particular in the presence of a lymphoepithelial cyst, which is a means of discovering HIV, as found in our study.

Conflict of interest

Authors of this article have no conflict or competing interests. All of the authors approved the final version of the manuscript.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2021.102623.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

Belem Ousmane: Corresponding author writing the paper. Rachid Aloua: writing the paper. Ouassime kerdoud: writing the paper. Kaouani amine: writing the paper. Faiçal Slimani: Correction of the paper.

Guarantor

Belem Ousmane.

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