International Journal of Otolaryngology and Head & Neck Surgery, 2023, 12, 287-292
https://www.scirp.org/journal/ijohns
ISSN Online: 2168-5460
ISSN Print: 2168-5452

Kimura Disease: A Rare Case of Parotid Localization and Review of the Literature (Case Report)

Drissia Benfadil, Lachakar Azzedine, Fahd El Idrissi El Ayoubi

Department of Otorhinolaryngology and Head & Neck Surgery, Oto-Neuro-Ophthalmology Research Laboratory (LRONO), Medicine Faculty, University Mohammed I, Oujda, Morocco
Email: benfadildrissia2@hotmail.fr, drlachkar997@gmail.com, fahd.el@hotmail.fr

How to cite this paper: Benfadil, D., Azzedine, L. and El Idrissi El Ayoubi, F. (2023) Kimura Disease: A Rare Case of Parotid Localization and Review of the Literature (Case Report). *International Journal of Otolaryngology and Head & Neck Surgery*, **12**, 287-292.
https://doi.org/10.4236/ijohns.2023.124029

Received: March 30, 2023
Accepted: July 28, 2023
Published: July 31, 2023

Abstract

Kimura's disease is a rare chronic affection of unknown etiology, endemic in the far east with a net male predominance. We present the case of a 37-year-old female patient who developed a tumor process in the left parotid region over a period of 3 years. A total left parotidectomy was performed with simple postoperative course. The anatomopathological study of the surgical specimen was in favor of Kimura's disease. Through this case and a review of the literature, we present the main clinical, anatomopathological, therapeutic and evolutionary characteristics of this rare disease.

Keywords

Kimura Disease, Parotid, Rare

1. Introduction

Kimura's disease is a rare inflammatory disease, described in China in 1937 and was first published by the Japanese Kimura *et al.* in 1948 [1]. Its etiology remains undetermined, endemic in the Far East with a clear male predominance [2]. It usually manifests as a unilateral painless soft tissue mass or subcutaneous nodule in the head and/or neck, which can easily invade the salivary glands and lymph nodes.

In this article, we report a case of Kimura's disease in the left parotid region. We will review the main clinical, paraclinical, therapeutic and evolutionary characteristics of this disease.

This study will help us in the future to discuss this disease when a parotid tumor is associated with cervical lymph nodes or subcutaneous nodules, especially

when this clinical symptomatology is associated with a tumor of the other salivary glands and/or a kidney involvement.

2. Clinical Observation

A 37-year female patient, with no particular medical history, has recently presented to the Mohammed VI University Hospital in Oujda with a painful cervical mass in the left parotid region, progressively evolving for 3 years.

The clinical examination revealed a preauricular mass over the left parotid region measuring two centimeters, solid, movable, non-pulsatile and without skin changes (Figure 1). We also find a left jugulo-carotid lymph nodes of group IIA. The motor function of the facial nerve is preserved. The rest of the cervicofacial examination did not reveal any lesions of the skin or scalp. The right parotid region and other salivary glands show no abnormalities with quantitative and qualitative conservation of saliva.

The Parotid ultrasound revealed two hypoechoic nodular formations of the left parotid gland with a hypervascularized appearance on color Doppler.

As part of the paraclinical examination, the parotid MRI identified a superficial and a deep polar lesion of the left parotid gland with malignant signs (Figure 2).

Her laboratory assessment showed a normal kidney function and no hypereosinophilia. Blood sugar, urea, creatinine, and bilirubin levels were within normal range. No abnormality was found in the routine urine test. Serum total immunoglobulin E (IgE) level was normal. Allergen patch test was negative. Rheumatism, rheumatoid factor, anti-nuclear antibody, complete parasite set, routine stool and urine, thyroid function, immunoglobulin G4, complement C3/C4, and D-dimer levels were normal.

A total left parotidectomy was performed with a group IIA lymphadenectomy (Figure 3). Low-dose radiotherapy and glucocorticoid treatment were not

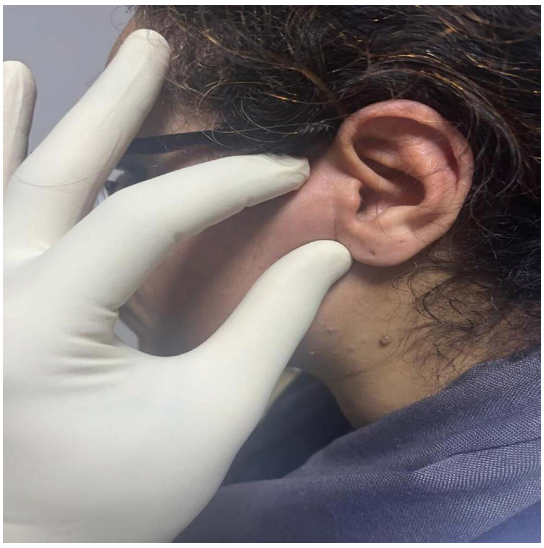

Figure 1. Clinical appearance of a left preauricular parotid tumefaction.

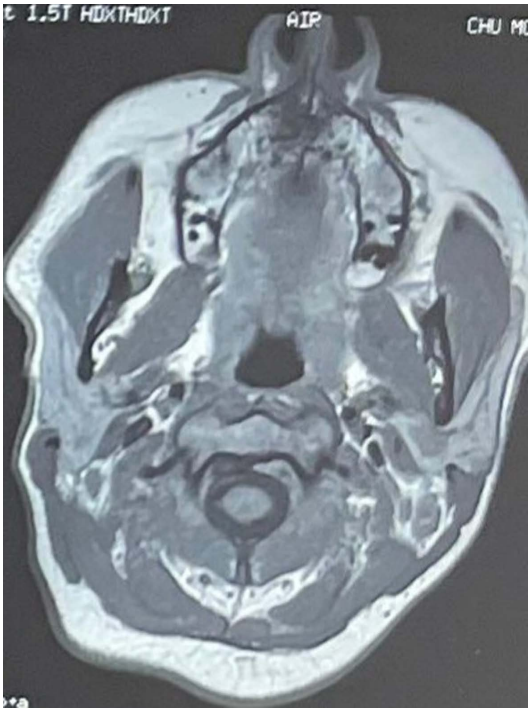

Figure 2. MRI section showing two nodules off the two parotid lobes.

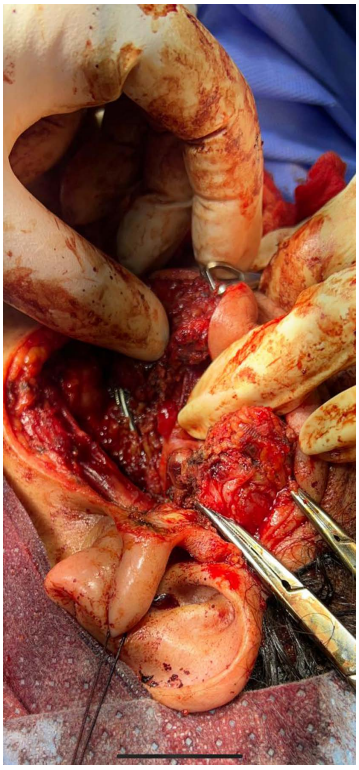

Figure 3. Intraoperative aspect of the deep parotid nodule.

necessary as the kidney function was normal and there was no hyperoesonophilia.

The histopathological examination revealed a Kimura's disease (**Figure 4**). and the postoperative course was simple.

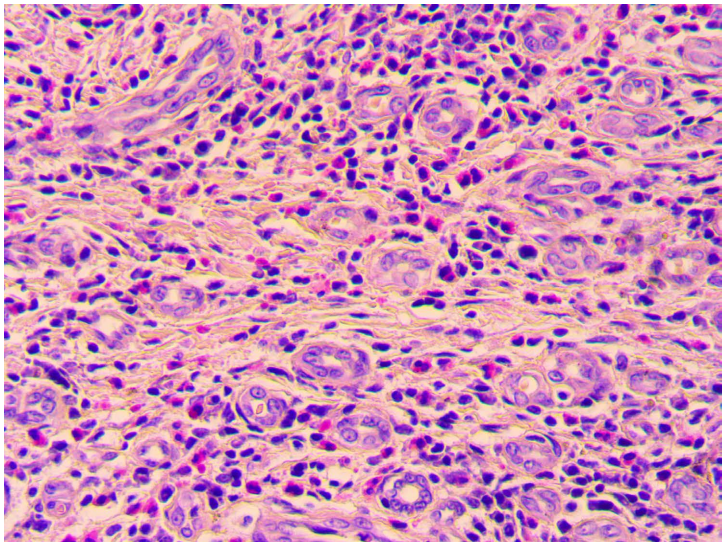

Figure 4. Image showing the anatomopathological appearance of Kimura disease of our patient.

3. Discussion

Kimura's disease was first published by the Japanese Kimura in 1948, also called unusual granuloma associated with hyperplastic changes in lymphoid tissue [1]. This disease affects almost exclusively patients from the Far East, and more exceptionally Caucasian subjects with a male predominance and a sex ratio varying according to studies from 3 to 7 [2]. All age groups are affected, with a peak around the third and fourth decade [3]. In our context it is a very rare case, and it was the first case of Kimura's disease operated on in our department. In our context, it is indeed an extremely rare disease. This is the first case of Kimura disease operated in our department.

It is an inflammatory disease of unknown etiology, but it is supposed to be an allergic or autoimmune process involving blood vessels, lymphocytes and eosinophils. This process is thus responsible of the development of subcutaneous nodules or plaques of varying size [1].

Clinically, the disease is characterized by the appearance of subcutaneous nodules that progressively increase in volume with clear boundaries. These nodules are usually painless and fixed in the superficial plane. There is also, solid, painless, or itchy single or multiple lesions on the head and/or neck, especially around the parotid gland and submandibular region, often associated with lymph node in the lateral cervical region and the supra-clavicular fossa [4]. The orbits, the ears, the scalp and the forearm are rarely described. Exceptionally, lesions may occur in the mucous membranes. Other regions such as axilla, groin, trunk, abdomen, peripheral limbs, chest wall, and median nerve have also been involved, their prevalence is extremely rarely [5]. Subcutaneous tissue and muscles may be affected. Locoregional adenopathy is frequently found as in our patient.

During its development, this disease affects the salivary glands with a predilection for the parotid gland. A nephrotic syndrome, related to extra-membranous glome-

rulonephritis, is present in 50% of the biological checkup often reveals blood hypereosinophilia with a hyperimmunoglobulinemia E.

Albuminuria is systematically requested in order to look for proteinuria, which would indicate a nephrotic syndrome [1] [6]. For the case in this study, the patient's kidney function was normal and no proteinuria was observed in her urine.

Cervical ultrasound, CT scan and MRI may be useful to determine the spread of the disease. Salivary gland and cervical lymph node involvement with contrast is the main findings and vascular involvement is suggested by the authors [7].

Elevated eosinophil count and total IgE levels in peripheral blood are the most prominent features in the laboratory findings of KD, but specific IgE level was not clinically significant in the diagnosis of KD. Blood eosinophil count and total IgE levels, in particular, can help monitor the prognosis of KD, which is very important in its diagnosis and treatment.

Pathological examination of the lesions after surgical resection shows hyperplasia of the lymphoid tissue with florid germinal centers. The main differential diagnosis is angiofollicular hyperplasia or Hale's disease. This is a western pathology that affects middle-aged women. The same anatomopathological aspect as Kimura's disease is noted, but with the addition of vascular proliferation that characterizes it [8].

In our case, there were not enough specific signs to suggest the disease before the surgery and, the anatomopathological study of the specimen.

The treatment of Kimura's disease is not yet codified. Surgical treatment is always indicated in the first instance, as in the case of our patient, and consists of a wide and deep excision in order to prevent recurrences, which are frequent. Corticosteroid therapy is indicated in profuse forms, when surgery is not possible and systematically in case of kidney damage. A dose of 0.5 to 1 mg/kg/day can be admitted with a progressive regression over 6 months. Radiation therapy (25 to 30 Gy) is indicated in corticosteroid resistant forms or when surgery is impossible [4]. Currently, other therapies are used but have not proven to be effective, such as cetirizine and interferon alpha or chemotherapy with 5-fluorouracil and azathioprine [4]. In our case, corticosteroid therapy was not necessary due to a complete succesful surgery. However, long term monitoring is necessary to detect further disorders.

Kimura's disease often has a chronic, indolent and benign course, without alteration of the general condition. Thus, tumor eruptions may occur with periods of complete remission, and recurrences after treatment of 15% - 40% [9].

Although the prognosis of the disease is benign, there is always a possibility of recurrence of tumors or renal disease. This requires long-term monitoring, consideration of a second surgery if needed or the possible use of various treatment protocols.

4. Conclusion

Kimura's disease is a rare entity of unknown etiology, common in the Far East

with a clear male predominance. Its diagnosis is based on the definitive anatomopathological result. The treatment is not yet codified but essentially based on surgical removal and corticosteroid therapy protocols.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Chen, H., Thompson, L.D.R., Aguilera, N.S.I. and Abbondanzo, S.L. (2004) Kimura Disease: A Clinicopathologic Study of 21 Cases. *The American Journal of Surgical Pathology*, **28**, 505-513. https://doi.org/10.1097/00000478-200404000-00010

[2] Docteur Claire Laroch, and Professeur Olivier Bletry O. (2005) La maladie de kimura. Encéclopédie orphanet. fevrier. https://www.orpha.net/data/patho/FR/fr-kimura.pdf

[3] Marcoux, C., Bourlond, A. and Decroix, J. (1991) Hyperplasie angiolymphoïde avec éosinophilie; Rémission sous acitrétine. *Annales de Dermatologie*, **118**, 217-221.

[4] Dubertret, L., Aractingi, S., Bodemer, C. and Cribier, B. (2001) Thérapeutique dermatologique, Médecine-Sciences. Flamarion; Kimura et hyperplasie angiolymphoïde avec éosinophilie.

[5] Ray, V., Boisseau-Garsaud, A.M. and Hillion, G. (2003) Maladie de Kimura à localisation palatine chez un Antillais. *La Revue de Médecine Interne*, **24**, 253-256. https://doi.org/10.1016/S0248-8663(03)00076-6

[6] Shetty, A.K., Beaty, M.W., McGuirt, W.F., Woods Jr., C.R. and Givner, L.B. (2002) Kimura's Disease: A Diagnostic Challenge. *Pediatrics*, **110**, e39. https://doi.org/10.1542/peds.110.3.e39

[7] Som, P. and Biller, H. (1992) Kimura's Disease Involving Parotid Gland and Cervical Lymph Nodes: CT and MR Findings. *Journal of Computer Assisted Tomography*, **16**, 320-322. https://doi.org/10.1097/00004728-199203000-00028

[8] Dib, N., Benhammou, A., Nazih, N., Essakali, L. And Kzadri, M. (2008) Maladie de Kimura: À Propos D'UN cas. *Journal Tunisien d'ORL et de Chirurgie Cervico-Faciale*, **20**, Article ID: 57933. https://www.ajol.info/index.php/jtdorl/article/view/57933 https://doi.org/10.4314/jtdorl.v20i1.57933

[9] Kawada, A. (1976) Morbus Kimura: Darstellung der Erkrankung und ihre Differential diagnostic. *Hautarzt Reutlingen*, **27**, 309-317.

KD: Kimura's disease