

Received: February 6th, 2023
Accepted: April 10th, 2023
Published Online: April 25th, 2023

*** CORRESPONDING AUTHOR:**

Larhrabli Ibtissam
Residence jnane psteur, boulevard
abdelmoumen, casablanca, morocco
Code postal: 20470
212615912975
Bettylar92@gmail.com
conflicts of interest: All authors attest that
they meet the current ICMJE criteria for
Authorship.
sources of funding: No funding or grant
support
ethical approval: done
Consent: Consent to publish the case
report was not obtained. This report does
not contain any personal information that
could lead to the identification of the
patient.

5-.PATIENT CONSENT:

Consent to publish the case report was
obtained.

6-.FUNDING:

No funding or grant support.

7-.AUTHORSHIP:

All authors attest that they meet the
current ICMJE criteria for Authorship.
Bushra abdulhakeem:
bushraabdulhakeem99@gmail.com
Larhrabli ibtissam :
bettylar92@gmail.com
Oukessou Youssef:
oukessouyoussef89@gmail.com
Rouadi Sami :
sami.rouadi80@gmail.com
Abada Redalah larbi :
larbiabada77@gmail.com
Roubal Mohamed :
roubal.mohamed11@gmail.com
Mahtar Mohamed :
mohamedmahtar00@gmail.com

CITATION

Abdulhakeem B, Ibtissam L,
Oukessou Y, *et al.*, 2024,
Rhabdomyosarcoma of the parotid
region in a child. *Cancer Plus*,
5(1):2483.
DOI:

Copyright: ©2023

Abdulhakeem, *et al.* This is
an Open Access article
distributed under the terms
of the Creative Commons
Attribution-NonCommercial 4.0
International License
(<http://creativecommons.org/licenses/by-nc4.0/>), permitting
all non-commercial use,
distribution, and reproduction
in any medium, which provided
that the original work is
properly cited.

CASE REPORT

Rhabdomyosarcoma of the Parotid Region in a Child

Bushra Abdulhakeem¹, Larhrabli Ibtissam^{1*}, Youssef Oukessou¹, Sami Rouadi¹, Redallah Larbi Abada¹, Mohamed Roubal¹, Mohamed Mahtar¹

¹ ENT Head and Neck Surgery Department, Ibn Rochd University Hospital, Faculty of Medicine and Pharmacy, Hassan II University, Casablanca, Morocco.

Abstract: Idiopathic granulomatous mastitis is a chronic granulomatous disease of the breast with unknown etiology. Clinical features of idiopathic granulomatous mastitis overlap with features of carcinoma breast. Fine needle aspiration cytology may be misleading in few cases. However confirmatory diagnosis is made from the core needle biopsy or excision biopsy. Steroids and immunosuppressive medications are the primary modality of the treatment. Surgery is the better option for patient who is fails to respond with steroid or immunosuppressive treatment. Surgery will remove the disease pathology and some times it may also remove invasive carcinoma which may occur concurrently with IGM.

Key words: Idiopathic granulomatous mastitis; Carcinoma breast; Breast abscess; Mastectomy; Immunosuppression

1. Introduction

Malignant tumors of the parotid gland are reported to be rare during childhood. Sarcomas account for < 1.5% of malignant tumors of the parotid gland^[1-3].

Rhabdomyosarcomas is one of the more common sarcomas of the parotid region occurring during childhood and adolescence, but overall its incidence remains rare^[1-2].

The parotid localization remains the most frequent, followed by that of the submandibular gland and exceptionally accessory salivary glands^[4].

It is a high-grade malignancy tumor. It is characterized by its locoregional aggressiveness, its metastatic evolution and its unfavorable prognosis^[3-5].

As a result, information on the clinical presentation, management, and outcome of patients with RMS in the parotid region is based on small patient series or case reports^[1,2,4,5] covering adults with two histological types of parotid tumors, as well as children^[1-4] or report Consider patients with parotid tumors and patients with Rhabdomyosarcomas in other head and neck regions^[5-6].

Based on our case report, we propose to review the data in the literature.

Bachround: A rare case of rhabdomysarcoma of parotid in child, locally advanced, well treated, better outcome.

Case presetation: an 8 year old child, from a rural region near casablnc, with no medical history has developped a mass in the left parotid region growing fast, fixing

a facial paralysis within 3 months, diagnosed with ct scan and biopsy, the patient underwent surgery for mass reduction and chemotherapy ivado than iva protocol, a notable reduction of the swelling was noticed after one year.

2. Case report

We reported a case of A 8-year-old child, with no medical history, consulted for a swelling of 9 to 10 cm in the long axis of the left parotid region, associated with complete ipsilateral facial paralysis developed for 3 months (Figure 1).



Figure 1. A large swelling of the left parotid region associated with a ipsilateral facial palsy.

The Examination of the oral cavity revealed bulging of the soft palate and the inner side of the left cheek and the examination of the ipsilateral external auditory canal revealed an externalization of the mass through the canal which completely obstructs it

3 adenopathy's were found in the examination of lymph nodes submental, submandibular and high carotid jugular 1 to 2 cm in diameter.

Our patient benefited of a facial CT scan that showed parotid tumor with significant locoregional extension medially to parapharyngeal space, outside pushes back the left auricle, anteriorly and below fills the infratemporal fossa, further forward it comes into contact with the retromolar trigone, behind arrives at the pre vertebral space, with no bone lysis (Figure 2).

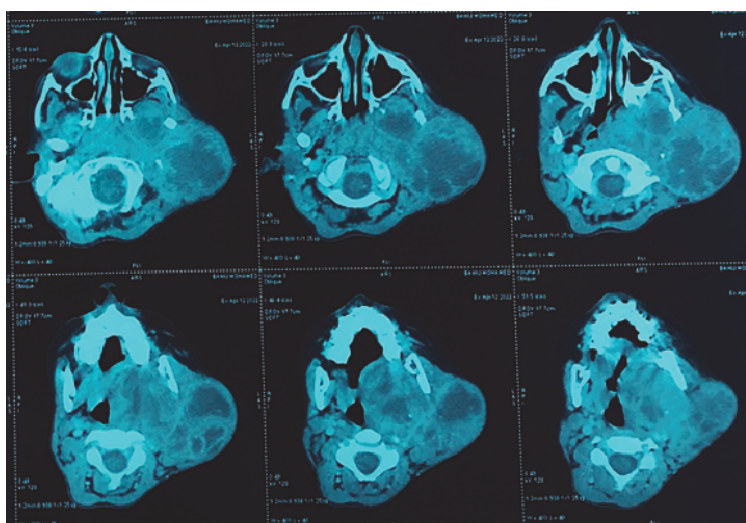


Figure 2. Axial section: Left parotid tumor with significant locoregional extension medially to parapharyngeal space, outside pushes back the left pavilion, anteriorly and below fills the infratemporal fossa, further forward it comes into contact with the retromolar trigone , behind arrives at the pre vertebral space , with no bone lysis.

The biopsy revealed an alveolar rhabdomyosarcoma whose macroscopic appearance was that of a brownish tumor with necrotic hemorrhagic areas. Microscopic examination showed that this tumor had an alveolar arrangement, the connective tissue was organized into a fibrillar stroma forming connective partitions delimiting closed or communicating cells.

The tumor was classified T1BN1M0 group II. Given the locally advanced nature of the disease, the patient was referred to the hemetological department for

chemotherapy and benefited from Ivado than Iva protocol, than the patient underwent surgery in our department by a left total parotidectomy with preservation of the facial nerve and left lymph node dissection of the territory IIA+ IIB with no tumoral residu in the anatomopathological exam, than he was referred for post-operative radiotherapy.

After one year (surgery + chemotherapy) we noticed a clinical improvement and a notable reduction of the swelling (Figure 3).



Figure 3. A notable amelioration after one year of treatment (chemotherapy + surgery).

3. Discussion

Salivary gland carcinomas are rare in childhood. A survey of salivary tumors from the Armed Forces Institut of Pathology^[5] revealed that 1.3% of benign and 3% of malignant salivary gland tumors occurred in children^[6,7].

Rhabdomyosarcoma represents 13% to 18% of head and neck tumors^[8,9] and less than 1% of salivary gland cancers^[10]. However, its exact frequency remains difficult to determine given the rarity of cases and the heterogeneity of the populations studied. In a series of 168 childhood salivary gland cancers, Krolls *et al.*^[11] reported six cases of rhabdomyosarcoma, i.e., 3.5%. Luna *et al.*^[20] reported two cases of rhabdomyosarcoma out of 11 cases of primary sarcoma of the salivary glands. The largest published series of salivary rhabdomyosarcomas included three to 22 cases^[12,13,6,14].

The parotid gland is the most frequent initial site of rhabdomyosarcoma before the submandibular and

sublingual glands. Involvement of the accessory salivary glands remains exceptional^[15]. Rhabdomyosarcoma is a high-grade malignancy tumor that affects subjects of all ages with a higher frequency in the first two decades of life, with, however, very broad extremes^[16,6,17,18].

Parotid rhabdomyosarcoma usually presents as painful swelling of the parotid gland that rapidly increases in size^[19,18]. Facial paralysis is a frequent sign, testifying to the aggressiveness and degree of tumor extension.

CT scans and magnetic resonance imaging (MRI) of the parotid are an essential contribution in terms of exploration of the parotid gland and evaluation of tumor extension to neighboring structures^[20,21]. MRI seems to perform better with a sensitivity greater than 95% and a specificity greater than 75%^[21]. A staging assessment combining pulmonary, hepatic and bone imaging is mandatory before any therapeutic decision. There is no universally accepted classification. Several are used,

they are generally addressed to rhabdomyosarcomas without taking into account the localization^[22,23].

Because of the aggressive nature of this tumor, treatment combines surgery, radiation therapy, and chemotherapy. Indications and relevance depend on classification, resection quality, and anatomic pathology findings; surgery is the cornerstone of treatment. It includes partial or total parotidectomy, with or without facial nerve preservation, according to the Gustave Rousey Institute classification, which is stage I and II cases and first-line treatment classification according to IRS group I and II cases (Intergroup Rhabdomyosarcoma study)^[24,23], radiotherapy is indicated for incomplete resection and/or histological lymph node invasion, and the radiation field must include the parotid gland where the tumor is located and the cervical-subclavian lymph node region (if involved). For persistent tumors and/or lymph node invasion with capsular rupture, a dose of 60 to 70 Gray is recommended. Due to their activity on salivary gland tumors and sarcomas^[25,5,26]. Adjuvant chemotherapy could improve local control and survival rates, but this remains to be demonstrated. Neoadjuvant chemotherapy could find its interest in locally advanced and/or inoperable tumors (stage III of the classification of the Gustave-Rousey Institute and group III of the IRS) by making them accessible to cardiological surgery.

Salivary rhabdomyosarcomas have a tendency to local recurrence, the time to onset is on average nine months^[23]. Pulmonary, hepatic and bone metastases are frequent, tumor dissemination can also occur towards the cerebrospinal fluid after invasion of the meninges. The time to onset is on average nine to 36 months^[27]. Survival rates are variable. From 22 cases, Pappos *et al.* reported a two-year disease-free survival rate of 63% after chemotherapy, radiotherapy and surgery^[13], whereas Flamant *et al.* reported an overall survival rate of 68% and five-year disease-free survival of 53%^[14]. For Cullender *et al.*, five-year survival is 60% in the case of chemotherapy and radiotherapy^[19].

The stages or groups of classifications are major prognostic factors since in the IRS study, the five-year survival rates for groups I to IV were 83, 70, 52 and 20% respectively^[28]. According to the Gustave-Rousey

Institute, the five-year survival rates for stages I and II, III and IV were respectively 80, 53% and almost 0%^[13,23]. Other prognostic factors have been analyzed, in particular the histological type: the more differentiated the tumor, the better the prognosis, the three-year survival rate is 60% in the botryoid forms and 40% for the for- my embryonic and alveolar. Due to more frequent lymph node invasion, the latter form has a 50% prognosis compared to 17% for the other forms^[29]. The most important unfavorable prognostic factors for Cullender *et al.* are age (55% five-year survival rate for children under seven years old and 33% for children over seven years old), alveolar type, and tumor size greater than 5 cm^[19].

4. Conclusion

Rhabdomyosarcoma (RMS) is one of the more common sarcomas of the parotid region occurring during childhood and adolescence, but overall, its incidence remains rare AND often have an unfavorable prognosis.

Improving results requires an early diagnosis and multidisciplinary management based on the results of multicenter trials. The inclusion of these patients in trials must be systematic due to the rarity of cases.

Conclusion: Rhabdmyosarcoma is a rare type of sarcoma that affect children, the early diagnosis combined with appropriate treatment can change the prognosis

References

1. Luna MA, Tortoledo E, Ordóñez NG, *et al.*, 1991, Primary sarcomas of the major salivary glands. *Arch Otolaryngol Head Neck Surg*, 117:302-306.
2. Rogers DA, Rao BN, Bowman L, *et al.*, 1994, Primary malignancy of the salivary gland in children. *J Pediatr Surg*, 29:44-47.
3. Enzinger F, Weiss S, 1995, Soft tissue tumors. Chapter 22: rhabdomyosarcoma. 3rd Edition. St Louis: Mosby.
4. Lussier C, Kljanienko J, Vielh P, 2000, Fine-needle aspiration of metastatic nonlymphomatous tumors to the major salivary glands. A clinicopathologic study of 40 cases cytologically diagnosed and Histologically Correlated. *Cancer Cytopath*, 90:350-356.
5. Douglas JG, Koh WJ, Austin-Seymour M, *et al.*, 2003, Treatment of salivary gland neoplasms with fast neutron radiotherapy. *Arch Otolaryngol Head Neck Surg*,

- 129:944-948.
6. Rogers DA, Rao BN, Bowman L, *Et al.*, 1994, Primary malignancy of the salivary gland in children. *J Pediatr Surg*, 29:44-47.
7. Taylor RE, Gattamaneni HR, Spooner D, 1993, Salivary gland carcinomas in children: A review of 15 cases. *Med Pediatr Oncol*, 21:429-432.
8. Callender TA, Weber RS, Janan N, *et al.*, 1995, Rhabdomyosarcoma of the nose and paranasal sinuses in adults and children. *Otolaryngol Head Neck Surg*, 112:252-257.
9. Douglas EC, Shapiro DN, Valentine M, *et al.*, 1993, Alveolar rhabdomyosarcoma with the t(2, 13): cytogenetic findings and clinicopathologic correlations. *Med Pediatr Oncol*, 21:83-87.
10. Luna MA, Tortoledo E, Ordonez NG, 1991, Primary sarcomas of the major salivary glands. *Arch Otolaryngol Head Neck Surg*, 117:302-306.
11. Krolls SO, Trodahl JN, Boyers RC, 1972, Salivary gland lesions in children. A survey of 430 cases. *Cancer*, 30:459-469.
12. Almedia M, Sastny JF, Wakfly PE, *Et al.*, 1994, Fine needle aspiration biopsy of childhood rhabdomyosarcoma: reevaluation of the cytologic criteria for diagnosis. *Diagn Cytopathol*, 11:231-236.
13. Pappas AS, Etcubanas E, Santana VM, *Et al.*, 1993, A phase II trial of ifosfamide in previously untreated children and adolescents with unresectable rhabdomyosarcoma. *Cancer*, 71:2119-2125.
14. Spraggs PDR, Rose DSC, Grant HR, *et al.*, 1994, Pathology in Focus: Post-irradiation carcinosarcoma of the parotid gland. *J Laryngol Otol*, 108:443-445.
15. Kapadia SB, Rajiv D, Hiroaki F, 1996, Botryoid embryonal rhabdomyosarcoma of Stensen's duct. *Am J Otolaryngol*, 17:127-132.
16. Enzinger F, Weiss S, 1995, Soft tissue tumors. Chapter 22: rhabdomyosarcoma. 3rd Edition. St Louis: Mosby.
17. Silverman JF, Joshi W, 1994, FNA biopsy of the small round cell tumors of the childhood: Cytomorphologic features and the role of ancillary studies. *Diagn Cytopathol*, 10:245-255.
18. Valencerina G, Dauterman J, 2000, Fine-needle aspiration biopsy of alveolar rhabdomyosarcoma of the parotid. A case report and review of the literature. *Diagn Cytopathol*, 24:249-252.
19. De M, Banerjee A, Graham I, *et al.*, 2001, Alveolar rhabdomyosarcoma of the parotid gland. *J Laryngol Otol*, 115:155-157.
20. David M, 2000, Major salivary gland imaging. *Radiology*, 216:19-29.
21. Zrounba PH, Bolot G, 1997, Apport de l'imagerie devant une tuméfaction de la parotide. *J Fr ORL*, 46:110-112.
22. Feldman BA, 1982, Rhabdomyosarcoma of the head and neck. *Laryngoscope*, 92:424-439.
23. Flamant F, Rodary C, Rey A, *et al.*, 1984, Treatment of non metastatic rhabdomyosarcoma in childhood and adolescents results of the second study of the international society of pediatric oncology, MMT. *Eur J Cancer*, 34:1050-1062.
24. Flamant F, Gerard-Marchant R, Schwaab G, *et al.*, 1978, Rhabdomyosarcomas de l'enfant. *Med Infant (Paris)*, 85:797-816.
25. Breteau N, Wachter T, Kerdraon R, *et al.*, 2000, Utilisation des neutrons rapides dans le traitement des tumeurs des glandes salivaires: rationnel, revue de la littérature et expérience du centre d'Orléans. *Cancer Radiother*, 4:181-190.
26. Noel G, Feuvret L, Ferrand R, *et al.*, 2003, Le traitement par neutrons: hadron thérapie partie II: bases physiques et expérience clinique. *Cancer Radiother*, 7:340-352.
27. Kwan WH, Choi PM, Li CK, *et al.*, 1996, Breast metastasis in adolescents with alveolar rhabdomyosarcoma of the extremities: report of two cases. *Pediatr Hematol Oncol*, 13:277-285.
28. Maurer HM, Gehan EA, Beltangady M, 1993, The intergroup rhabdomyosarcoma study II. *Cancer*, 71:1904-1922.
29. Newton WA, Hamoudi A, Webber B, *et al.*, 1991, Pathology of rhabdomyosarcoma and related tumors: experience of the intergroup rhabdomyosarcoma studies. In: Maurer HM, Ruymann F, Pochedly C, editors. Rhabdomyosarcoma and related tumors in children and adolescents. Boca Raton, FL: CRC Press:19-47.

Publisher's note

Accscience Publishing remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.