Sialoblastoma: A historical review of diagnosis and management

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Abstract

The Sialoblastoma is a rare malignant tumor of the epithelial cells of the salivary glands, with less than 100 cases reported. Its main location is in the parotid gland, but it may affect other salivary glands. The diagnose should be investigated in cases of a congenital tumor in the topography of these salivary glands and may be confirmed by open biopsy, or after complete excision. Due to rarity of the disease, there is no consensus for the conduct in the literature, but surgical treatment shown better outcomes. The adjuvant treatment with chemotherapy may be feasible to advanced metastatic cases.

Sialoblastoma: A historical review of diagnosis and management

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Abstract

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Introduction

Sialoblastoma is a rare malignant tumor originating from the epithelial cells of the salivary glands, with most of the cases having a congenital origin and occurring in the neonatal period or early childhood, with a few cases reported in adults[1]. Only 77 cases are described in the literature. It is a rare neoplasm, being estimated by The Surveillance Epidemiology database and End Results[2] an incidence of 0.1 per 100,000 salivary gland tumors in children under 15 years old. This tumor affects the major salivary glands, especially the parotid gland, where it presents more aggressive behavior. The second most common occurrence site is the submandibular gland, with a less aggressive behavior[3], however it can affect minor salivary glands and even ectopic salivary glands[4,5]. The first record of sialoblastoma was in 1966, by Vawterand Tefftin, in which

the author believed to be the first parotid tumor recorded in a newborn, classifying it as a benign tumor[6]. In 2005, the World Health Organization (WHO) reclassified it as a malignant tumor of salivary glands[7].

Due to the rarity of this tumor, there is no consensus in the literature regarding the management of this pathology. In the present study, we report a case of a five-month-old patient who presented a malignant tumor of the minor salivary gland in the right cheek mucosa since birth and with rapid growth and development of metastases. She was operated on at the National Cancer Institute and had a diagnosis of Sialoblastoma, continuing her treatment in the same institution.

Case Report

L. F. R, female, since birth, a nodular lesion was observed on the right side of the face. An ultrasound scan (USS) of the face was performed when she was 22 days old. A solid hypoechogenic lesion was observed, with well-defined contours, measuring 3.10 x 2.86 x 2.29 cm, located in the subcutaneous cellular tissue and muscular planes on the right side of the face. The initial hypothesis was a congenital hemangioma, based on which treatment with beta-blockers (propranolol) was prescribed for 4 months, but there was no response. Then she was referred to the Brazilian National Cancer Institute at five months of age. Magnetic resonance imaging (MRI) of the face was performed, showing a lesion approximately 3.90 x 3.60 x 3.30 cm. Also, a computed tomography (CT) scanned the chest, which showed diffusely distributed nodules and soft tissue density highly suggestive of metastasis. Further exams were performed, but inconclusive, including fine needle biopsy (FNA).

Surgical treatment was performed by a transoral approach with complete excision of the lesion (Fig. 1). The histopathological anatomy report the sialoblastoma. The macroscopy showed a rounded tumor measuring 5.0 x 3.5 x 2.0 cm, partially covered by a capsule with a vascularized surface with an epicenter on the right cheek mucosa. The immunohistochemical report showed diffuse positivity for AE1 / AE3 with ductular enhancement; Positive S-100; p63 positive; Calponin, Actin 1a4, negative desmin and myogenin; and cell proliferation rate by ki67 of about 80%.

In follow up, the patient showed a good postoperative evolution, removing the nasogastric tube after 5 days. A new MRI of the face was requested (Fig. 2: B and D), without signs of residual disease or recurrence. As the patient presented pulmonary nodules, the therapeutic intervention of chemotherapy with etoposide, mesna and vincristine was chosen. The patient is still monitored by the pediatric oncology.

The parents agreed to the publication and a form was signed authorizing the use of the image.

Discussion

Sialoblastoma is a rare malignant tumor that is most common among the pediatric population and it originates from the epithelial cells of the salivary glands. According to Batsakis' "salivary gland unit" theory, this neoplasm results from the activation and proliferation of reserve multipotent cells in rest[11]. The most commonly affected sites, according to the literature, are the major salivary glands, such as the parotid gland[3]. In the case studied here, the minor salivary gland in the jugal mucosa was the location affected.

Its main feature is the presence of a mass with accelerated growth in the topography of the affected gland. The typical presentation is a fibroelastic tumor in the parotid or cervical-facial region in the child, which commonly leads to confusing diagnosis with other congenital and neonatal tumors, such as hemangioma[12]. As reported in the literature, in the case described, the main symptom was, since birth, a mass with progressive and continuous increase in size in the right nasogenian region. The diagnosis is confirmed by biopsy, however fine needle aspiration is an alternative that aims to reduce morbidity and accelerate the investigation[3].

The histological findings of sialoblastoma are based on hematoxylin and eosin staining and are corroborated by immunohistochemical findings (S100, cytokeratin, P63, Her 2 Neu, hight ki67)[13]. In the described report, the histopathological findings resulted in sialoblastoma and the immunohistochemical ones were S-100 positive; p63 positive, and cell proliferation index by ki67 of around 80%. In macroscopy, it was evidenced that areas of necrosis are related to a worse prognosis of the disease[14]. In the literature, it is described that p63

positivity is usually confirmed, with focal positivity related to a favorable prognosis, whereas diffuse positivity to unfavorable outcomes[15]. Positivity for ki67 is associated with a worse prognosis and a greater chance of recurrence[14]. Another important marker for the diagnosis and course of the disease is the alpha-fetoprotein, which at high serum levels is also considered a factor of aggravated evolution[3].

Image exams are important before surgeries and in the post-treatment follow-up[3]. The description of these exams in the literature regarding sialoblastoma is limited and there isn't a defined protocol. Computed tomography, in general, shows a hypodense soft tissue mass concerning the brain and isodense regarding muscles[12,16]. In nuclear magnetic resonance, an isodense lesion was observed with muscle tissues in T1[17]. Di Micco et al described a well-defined mass with increased echogenicity on ultrasound[3].

Sialoblastoma is considered a locally aggressive tumor, but it is also described in the literature as regional recurrences, lymphatic dissemination, and distant metastases[5]. In the review carried out by Kanaram from 1966 to 2011, of the 46 analyzed cases, ten had local recurrence in 24 months of follow-up and no metastasis or residual disease resulted in death. Analyzing the total number of cases with recurrences, two had only pulmonary metastasis and two others presented pulmonary metastasis and dissemination to cervical lymph nodes[12]. Regarding the staging of sialoblastoma, in 1992, a histopathological criterion was proposed by Batsakis and Frankenthaler, which define the stage of tumor malignancy and if the surgical resection was effective[11,18]. The characteristics evaluated include perineural and perivascular invasion and cell necrosis with cytological atypia.

There is still no well-established protocol for first-line treatment. The literature suggests that surgical resection with free margins is sufficient for the treatment of low malignancy tumors[3,19]. Qian Wang et al described that chemotherapy plays an important role in the treatment of residual disease, with extensive metastasis or recurrences[1]. There is still no defined consensus regarding the indication for prophylactic neck dissection. If there is clinical and radiological evidence of metastasis, therapeutic neck dissection is indicated[20]. In 2010, Xiao-Feng Shan described the possibility of brachytherapy as an adjunct treatment to surgery[9]. Thus, treatment must be individualized, according to the patient's age, the possibility of locoregional disease, tumor aggressiveness, and the potential to develop metastasis

In the article we reported a rare case of a Sialoblastoma of the minor salivary gland, with pulmonary metastasis, that was surgically treated and underwent adjuvant chemotherapy. Due to rarity the diagnosis was difficult at first and the treatment was delayed. The patient is still on an ambulatorial and radiological follow up and a multidisciplinary team is carrying out follow-up. We aim to contribute to the literature of Sialoblastoma diagnosis and treatment.

The authors have no conflicts of interest to declare.

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Abbreviations

WHO	World Health Organization
USS	Ultrasound scan
MRI	Magnetic resonance imaging
CT	Computed tomography
FNAB	Fine-needle aspiration biopsy

Legend List

Figure 1: A: Incision in the tumor mucosa B: Dissection of the lesion without rupture of the capsule. C: Aspect after complete excision of the lesion with visualization of Bichat Ball D: Final surgical aspect.

Figure 2: Comparison between pre (A and C) and postoperative (B and D) imaging exams in the axial and coronal planes, respectively.

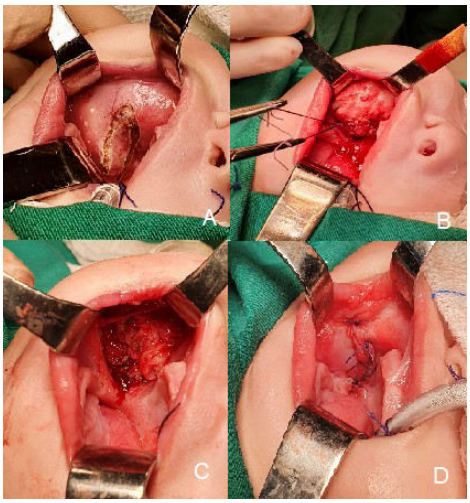

Figure 1

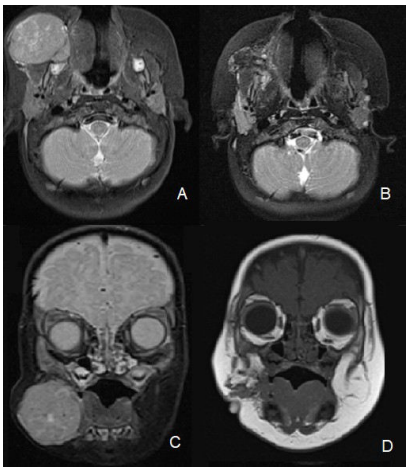

Figure 2