Sialoblastoma- long-term follow-up and remission for a rare salivary malignancy

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Abstract

Sialoblastoma is a rare salivary neoplasm which presents either congenitally or during early infancy. It was originally considered a benign neoplasm, however a number of reported cases have documented locoregional recurrence and distant metastases. Currently, there is no consensus on the appropriate treatment for this neoplasm. We report on long term follow-up of a patient with metastatic sialoblastoma, and a brief discussion of the possible treatment modalities currently being considered.

Introduction

The pediatric population, specifically under 15 years of age, accounts for only about 4% of all patients with salivary gland tumors.[1] In a review of the Salivary Gland Register of the University of Hamburg, only 2.5% of all epithelial neoplasia occurred in patients younger than 20 years.[2] Sialoblastoma is a very rare salivary tumor initially reported in 1966 by Vawter and Tefft under the nosology of embryoma. This tumor has also been referred to as congenital basal cell adenoma, basal cell adenoma, and basaloid adenocarcinoma. It may present congenitally or perinatally.[3] Most commonly, it originates in the parotid but may also arise in the submandibular gland.[4,5]

Sialoblastomas were initially classified as benign. However, increasing experience, including reports of distant metastasis, has lead to the reclassification of sialoblastomas as malignant.[6,7] Although the mainstay of treatment is surgical resection with adequate margins,[8] there are reported patients who have received alternative treatments such as neoadjuvant chemotherapy, radiation therapy, and even brachytherapy. We have previously reported a young child who presented with a parotid sialoblastoma at the age of 3.5 months.[9] This report extends the follow-up on this patient who is now 17 years old and in remission for 7 years. This represents the longest published follow up for a patient with metastatic sialoblastoma, and the first report of prolonged remission.

Case Report

We originally reported a patient with a sialoblastoma of the left parotid diagnosed at 3 months of age.[9] The tumor was locally invasive and recurred multiple times. Increasing tumor mitotic index and cytological pleomorphism was noted with recurrences (Figure 1). She was treated with incomplete surgical resection, chemotherapy (vincristine, adriamycin, cytoxan, etoposide and cisplatinum) and high-dose radiation therapy at 3.5 years of age. Orbital recurrence developed 1.5 years later, which was completely resected with negative margins; adjuvant radiation therapy was administered. At 6.5 years of age, she developed metastasis to the right lung. There was complete radiologic response after six cycles of Ifosfamide, Carboplatinum and Etoposide; however the patient developed metastasis to the left lung two years later, which responded to two cycles of Cytoxan and Topotecan (Figure 2). The left lung metastases were resected with negative margins, and she received six more courses of adjuvant chemotherapy with the same agents. Unfortunately 6 months later, she developed a third pulmonary recurrence in the right upper lobe, this metastasis was treated by a wedge resection.

Presently, this patient is 17 years old; she remains in disease-remission seven years from the last intervention. This remission certainly seems indicative of a durable response, as all recurrences had occurred at most 24 months from the end of previous treatment.

Discussion

Primary surgical resection with negative margins is considered the mainstay therapy for sialoblastomas.[5,8,10,11] In cases complicated by metastases, recurrences, or difficulties in surgical access or resectability, additional chemotherapy and/or radiation may be necessary.[12,13] Neoadjuvant chemotherapy and/or radiation may result in good response. Saribeyoglu *et al.* reported on a patient with locally recurrent sialoblastoma who was treated with chemotherapy only, obviating the need for surgical intervention.[13] Optimistically, that patient was in remission for 4 years at the time of publication. However, this is an isolated report of a durable response; most of the published reports lack long term follow-up. Scott *et*

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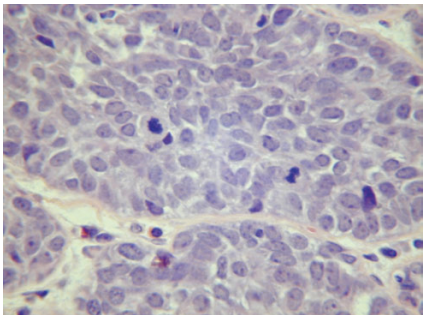

Figure 1. Recurrent sialoblastoma, which forms nests of pleomorphic basaloid cells with brisk mitotic activity.

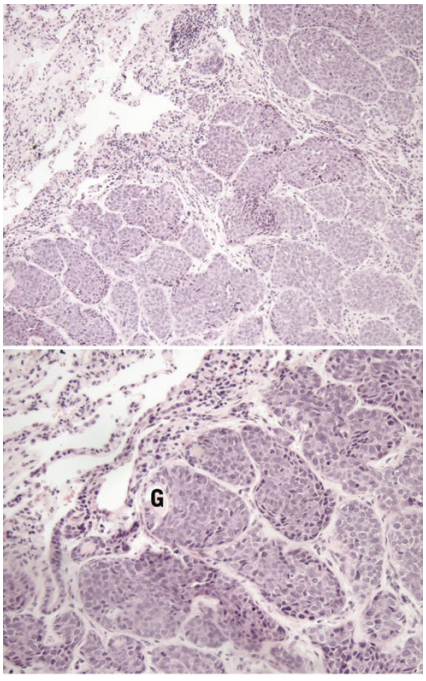

Figure 2 Upper panel: low power photomicrograph of pulmonary metastasis. Lower panel: high power photomicrograph demonstrating primitive gland formation (G).

al. reported a young girl with a sialoblastoma which had also metastasized to the lungs. The tumor was resected after a good response to neo-adjuvant chemotherapy; pulmonary metastases were treated by partial lobectomy.[14] Only six months of follow up was reported for this patient. Ersoz *et al*. describe a patient still receiving chemotherapy for pulmonary relapse at the time of the report.[6] Likewise, Prigent *et al*. described a patient with sialoblastoma and pulmonary metastases who was in remission one year following neoadjuvant chemotherapy and resection.[12] A novel approach to tumor recurrence was recently reported by Shan et al. In an effort to preserve facial nerve function and avoid a second extensive resection, brachytherapy with a 125I seed implant was utilized, which lead to a complete clinical response after two months. The patient was disease-free 21 months after this intervention; however the durability of this response cannot is unknown.[15]

In our case, the increased tumor mitotic index and cytological pleomorphism preceded development of distant metastases. Despite the initial response to Ifosfamide, Carboplatinum, and Etoposide, and Cytoxan and Topotecan, chemotherapy and radiation, these modalities did not prevent ultimate disease-progression. Importantly, surgical resection of pulmonary metastasis has lead to a durable remission.

While treatment planning for children with sialoblastomas should be individualized for each patient, surgical resection of recurrent and metastatic disease provided our patient with a long-term remission.

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