

Progressive Metastatic Sialoblastoma in a Young Child: Challenges in Treatment

Sialoblastoma is one of the rare tumors of the salivary gland. It is a congenital tumor arising most often from the parotid gland. This tumor is also known as basaloid adenocarcinoma, basal cell adenoma, and congenital basal cell adenoma. It has significant variability in biologic behavior. It was originally considered a benign tumor, but locoregional and distant metastasis have been described. Here, we describe a child with metastatic progressive sialoblastoma.

A 1-year 7-month-old Indian girl presented to us with complaints of swelling of the left cheek for a year. She was treated with multiple courses of antibiotics, despite which the swelling persisted. She was noted to have a 5 cm × 6 cm swelling over the left parotid area. Ultrasound of the parotid region revealed lobulated hypoechoic lesion in the superior lobe of the left parotid gland. Magnetic resonance imaging neck showed enlargement of the superficial lobe suggestive of parotid neoplasm. Fine-needle aspiration cytology was suggestive of rhabdomyosarcoma elsewhere, and she underwent superficial left parotidectomy. Histopathological evaluation of the specimen was suggestive of Grade III sialoblastoma with immunohistochemistry positive for S100, vimentin and patchy staining for EMA. She developed a 6 cm × 8 cm firm swelling in the parotid area extending from the inferior orbital margin to the mandible and from the angle of the mouth to the retroauricular region 1 month after surgery [Figure 1]. Fluorodeoxyglucose positron emission tomography–computed tomography (PET CT) scan revealed multiple lung nodules. She underwent left radical parotidectomy [Figure 2]. Chemotherapy and radiotherapy were offered as treatment options, but parents refused treatment. PET CT was repeated one-month post surgery. It showed new lesion in the infratemporal fossa

and increase in the size and number of lung lesions. The family was counseled regarding repeat surgery followed by radiation and chemotherapy; however, they did not opt for any treatment. She was followed up telephonically for 10 months postsurgery during which she was doing well. She was lost to follow-up after 10 months.

Sialoblastomas are rare salivary gland tumors usually present at birth or recognized shortly after birth. Age of presentation is reported from the prenatal period to 4 years.^[1] Sitthichaiyakul *et al.* observed that the median age of diagnosis was 9.8 years.^[2] There have also been isolated case reports of sialoblastoma in adults.^[1]

The incidence is extremely rare with only single case reports being reported. Choudhary *et al.* did a literature search and reported 46 cases from 1966 to 2011.^[1] Irace *et al.* reviewed 62 cases of pediatric sialoblastoma in 2016.^[3]

There is no clear consensus regarding the pathogenesis of sialoblastoma. It is thought to arise from the retained blastemal cells in the salivary gland.^[2]

Sialoblastomas usually tend to progress locally, with local recurrences and rarely metastasize.^[2] Irace *et al.* did a literature review of 62 cases, of which only nine patients had metastatic disease.^[3] In a review by Choudhary *et al.*, 10 cases out of 46 recurred. Most cases recurred within 24 months.^[1]

Due to the rare incidence of the tumor, there are no standard treatment protocols.^[4] Wang *et al.* reported three cases of congenital sialoblastomas, which did not recur after surgery and after a mean follow-up of 34 months. They concluded that surgical resection of the tumor is the safest modality of treatment.^[5] Chemotherapy has



Figure 1: Preoperative picture showing massive swelling on the left cheek



Figure 2: Postoperative picture showing considerable decrease in the size of the swelling

also been tried as a treatment modality as this is a very chemosensitive tumor. Saribeyoglu *et al.* published a case of recurrent sialoblastoma who was treated with vincristine-, actinomycin D-, and cyclophosphamide-based chemotherapy. They suggested chemotherapy for unresectable or recurrent sialoblastoma.^[6] Neoadjuvant chemotherapy has also been used with good outcome by Prigent *et al.*^[7] Radiation has also proved beneficial in treatment of sialoblastoma with incomplete removal.^[2]

Conclusion

Sialoblastoma is a rare tumor in the pediatric age group that arises from the salivary gland. Our case highlights the aggressive nature of the disease with local and distant metastasis. Surgery remains the cornerstone of treatment with radiation and chemotherapy considered useful in metastatic disease. However, as there are no standard protocols, treatment has to be individualized.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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