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# BRIEF REPORT
# Management of Sialoblastoma With Surgery and Brachytherapy

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Sialoblastoma is a rare congenital or perinatal salivary gland tumor with significant variability in histological appearance and biological behavior. We reported one case of sialoblastoma occurring in the parotid gland of an 18-month-old female. The tumor was excised with negative margins, and the sacrificed facial nerve was reconstructed with great auricular nerve graft. The tumor recurred 6 months after operation and was treated with $^{125}$I seed implant brachytherapy. No recurrence was found after 21 months of follow-up. Pediatr Blood Cancer. 2010;55:1427–1430. © 2010 Wiley-Liss, Inc.

**Key words:** brachytherapy; facial nerve; sialoblastoma; surgery

## INTRODUCTION

Less than 5% of all salivary gland tumors occur in children younger than 10-year old and congenital salivary gland tumors or salivary gland tumors in neonates are extremely rare [1,2]. Sialoblastoma is a new type of epithelial salivary gland tumor according to the Third Edition of the World Health Organization tumor classification [3]. It usually occurs in fetuses or neonates, while its cytology and histology have been described, its clinical characteristics and management are not well defined [4,5]. In this paper, we report the case of an 18-month-old patient with sialoblastoma.

## CASE REPORT

An 18-month-old female presented with a left parotid tumor. This tumor had grown from the size of a peanut to that of a table tennis ball within 1 month. Antibiotic therapy was ineffective. Physical examination showed that the mass was below the left earlobe, measured approximately $3.0 \times 3.0 \times 4.0\,cm^3$, was hard to the touch and that mobility was poor. The movements of the facial muscles were normal and symmetrical. A computerized tomography (CT) scan showed that the left parotid region was expanded by a soft-tissue mass that was isodense relative to muscle and occupied almost all the parotid area (Fig. 1A). There were swollen lymphatic nodes around the tumor.

An operation was performed to excise the parotid tumor. The tumor was multinodular and was surrounded by a pseudocapsule. It occupied the superficial and deep lobe of the parotid gland, infiltrating the five branches of the facial nerve. The diagnosis made from the frozen section biopsy was of a sialoblastoma. Considering the aggressive nature of this tumor and its potential for malignancy, the tumor and the lymphatic nodes around it were completely resected, the related branches of facial nerve were also sacrificed in order to get a tumor-free margin. The great auricular nerve on the same side was harvested and anastomosed the distal part to the facial nerve trunk. Proximal parts of donor nerve were anastomosed to marginal mandibular branch of facial nerve in end-to-end and to other four branches in end-to-side nerve graft with opening epineural windows (Fig. 2A,B).

A paraffin section of the tumor confirmed the diagnosis of sialoblastoma (Supplemental Fig. 1). After the operation, the girl experienced left facial palsy, a decline in muscle tension, and asymetrical movements of the facial expression muscles. The postoperative course was uneventful and the patient was discharged 5 days after the operation.

The patient was followed up every month postoperatively. Six months after the operation, a mass reappeared in the left parotid region. A CT scan showed that the normal tissue of the left parotid gland had disappeared and a mass had developed with an obscure boundary. The recurrent tumor infiltrated the retromandibular space and showed a tendency for invasion of the infratemporal fossa (Fig. 1B). The function of grafted facial nerve had recovered very well achieving House–Brackmann scores of II. In the remainder, a slight asymmetry could be seen; the left eyebrow and the left corner of mouth were a little lower than the other side. In motion, the normal movement of the brow and ability to close the eyelid could be achieved symmetrically. The patient was able to move the corners of her mouth with maximal effort and with obvious asymmetry (Fig. 2C–E).

Extensive resection was not chosen at this time because a safe margin would be hard to achieve and the facial nerve would again have to be sacrificed. Instead, $^{125}$I seed implant brachytherapy was used. The target of radiation included the entire left parotid space (Figs. 1C, 2F).

Two months after brachytherapy, the left parotid tumor was no longer evident in clinical examination nor on the CT scan (Fig. 1D). The only side-effect was low-grade dry desquamation on the left cheek, which disappeared 9 months after brachytherapy (Fig. 2G,H). After 21 months of follow-up, no tumor recurrence nor any symptom of developmental anomaly was found.

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Additional Supporting Information may be found in the online version of this article.

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Conflict of interest: Nothing to report.

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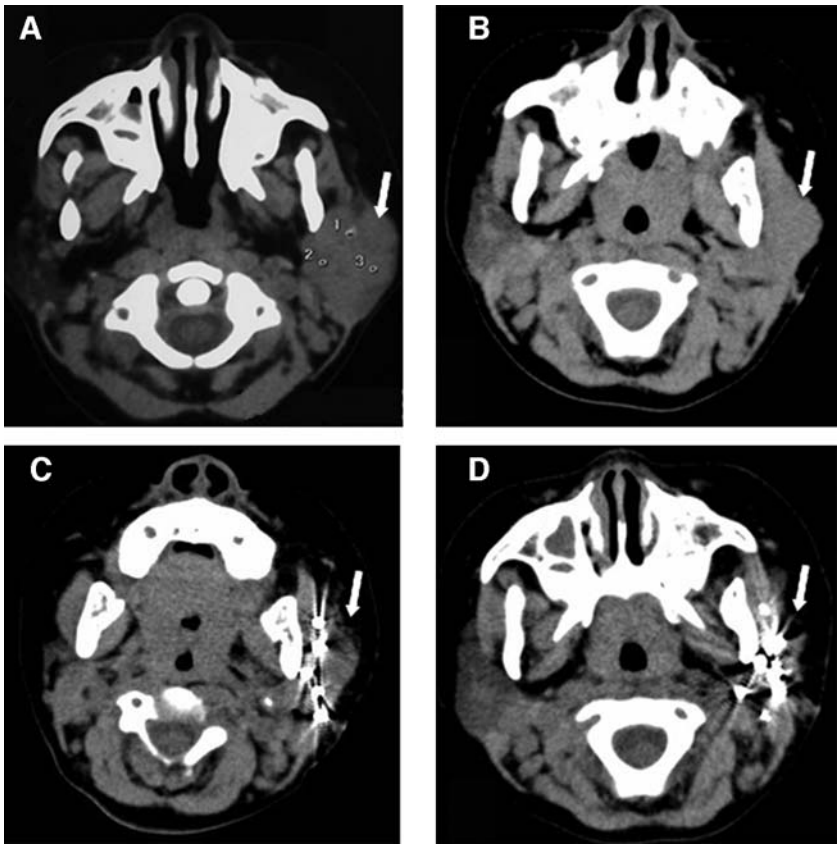


**Fig. 1.** **A**: CT scan showing the parotid tumor before surgery. **B**: CT scan showing the recurrent left parotid tumor 6 months after the initial surgery. **C**: CT scan 2 days after brachytherapy. **D**: CT scan that illustrates the disappearance of the tumor 2 months after brachytherapy. (The white arrows show the parotid tumor.)

## DISCUSSION

Sialoblastoma is a rare, locally aggressive, perinatal or congenital tumor that involves the parotid and submandibular gland [6]. The characteristics of sialoblastoma have been pathologically defined. It qualified as a malignant tumor for being locally aggressive and having the potential for metastasis to the lungs and lymphatics [6,7]. However, in some cases, sialoblastoma is well-encapsulated and no recurrences occur when they are excised with a narrow margin, especially in the submandibular gland [8]. It is difficult to define clearly when these tumors are benign or malignant. This is dependent on the clinical features of the tumor, as well as its histology and cytology. The histological features that suggest malignant potential are necrosis, cellular atypia, and vascular or neural invasion [9]. When recurrences occur, the histological profile becomes more unfavorable. There were four patients in the literature who experienced lung metastasis and one patient who developed metastasis to the neck lymphatic nodes. This result indicates that sialoblastoma is malignant in some cases.

We reviewed the English literature and found that only 41 cases of sialoblastoma have been reported until now. The follow-up time was from 7 months to 15 years. Ten cases originated in the submandibular gland and the others arose in the parotid gland. The case total contained 20 males, indicating that the male:female ratio is likely to be approximately 1:1. Some affected individuals also had an extensive congenital nevus [10]. Two children died from sialoblastoma [11].

In 10 cases, the parotid gland tumor recurred between one and four times, suggesting a recurrence rate of 34%. No recurrence of sialoblastoma in the submandibular gland has been reported [1,7]. There are two reasons which may account for this difference. Firstly, facial nerve preservation should be considered as a factor in surgery when the tumors are in the parotid gland and the dissection in this case may break the pseudocapsule or leave a positive margin. When sialoblastoma occurs in the submandibular gland, submandibular glands, and tumors can be resected together. Secondly, when the envelope around sialoblastoma is flaky, microtumors may also be found within the capsule. It has been previously reported that primary excision with negative margins is the most important factor and is sufficient for the cure of sialoblastoma [4]. However, some tumors recurred quickly when resection around the capsule was done, as in the case we described in this report. For safety, extended resections should be performed, especially in the parotid region.

The therapy of recurrent cases was reviewed [6,11,12]. Surgery was the preferred method of the treatment of recurrent sialoblastoma. Chemotherapy and radiotherapy were also used. Eight individuals received chemotherapy, and chemotherapy cured the sialoblastoma in five cases and stopped disease progression in one. Seven patients with recurred tumor were treated with radiotherapy, and three cases were cured after irradiation. The follow-up time of patients who underwent chemotherapy or radiotherapy was less than

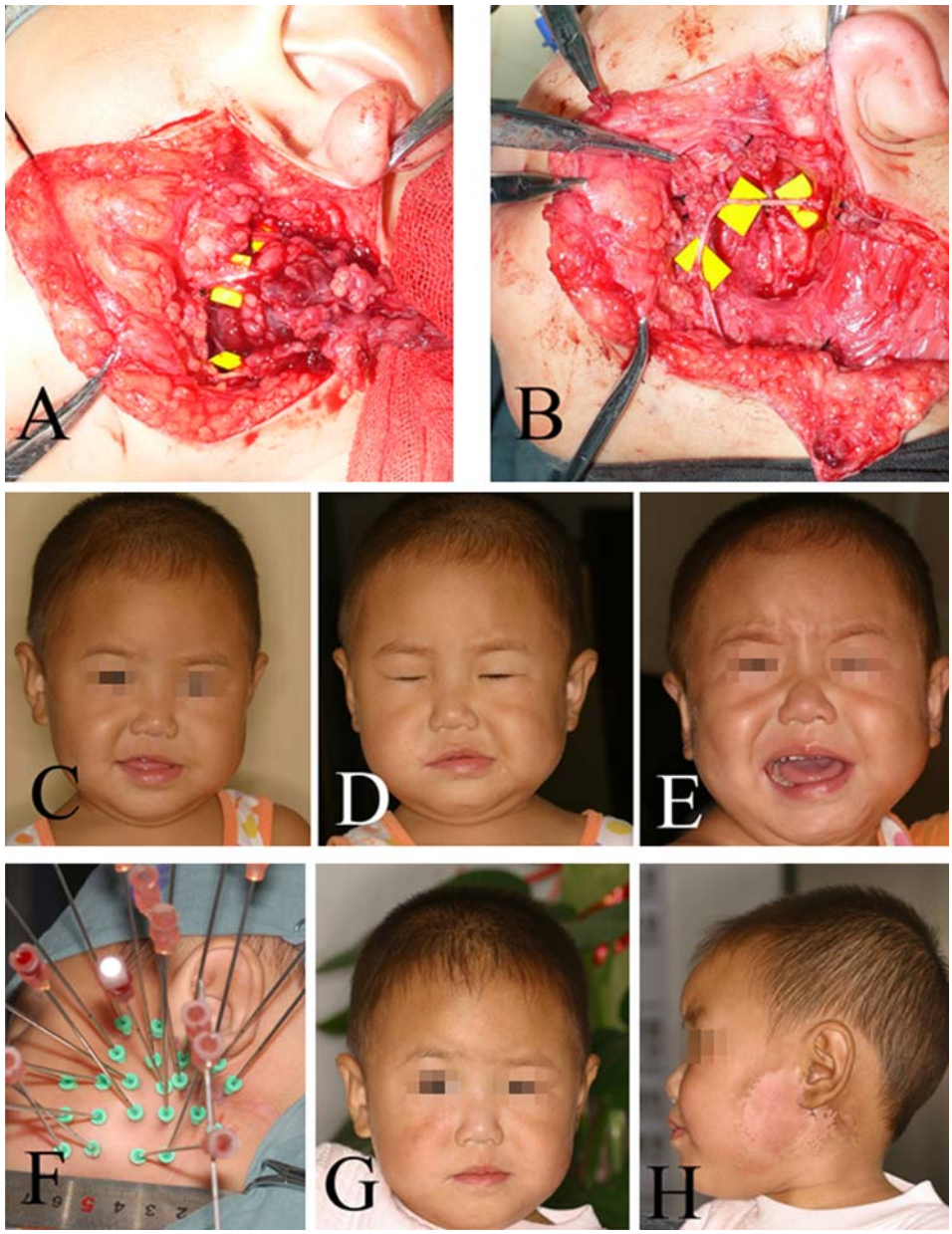


**Fig. 2.** **A**: Facial nerve infiltrated by the tumor. **B**: The defect of facial nerve was reconstructed with great auricular nerve in side-to-end graft after tumor resection. **C**,**D**,**E**: Different emotional expression showed facial nerve function recovered very well in the 6 months after the first operation. **F**: The execution of brachytherapy. **G**,**H**: No tumor and only low-grade dry desquamation in the left cheek that was only present for 9 months after brachytherapy.

2 years and no further recurrence was reported. The adverse effect of chemotherapy and radiotherapy on the development and growth of facial structures could potentially be very profound, although this was not reported.

Our patient received $^{125}$I seed implant brachytherapy when recurrence of the tumor occurred. Brachytherapy is a form of radiotherapy which delivers high-dosage irradiation 1.5 cm around the implanted $^{125}$I seeds and can have a less side-effect on the tissue beyond the area [13]. Brachytherapy appears to offer a high cure rate and to have only minor side-effects on the tissue around the tumor, and on pediatric facial and body development overall. In our patient, surgery was considered the only way to treat the tumor initially. Brachytherapy was used when the tumor recurred because a safe margin would be hard to achieve using surgical methods. After 18 months of follow-up, no developmental anomaly was found, which suggests that $^{125}$I brachytherapy could be a useful way to treat sialoblastoma.

Facial nerve function preservation should also be seriously considered for its significant impact on quality of life and social communication [14]. Due to the fact that sialoblastoma can be locally aggressive and neurally invasive, the facial nerve may need to be sacrificed to preclude recurrence if the nerve is involved. Facial nerve reconstruction has not previously been reported in cases of sialoblastoma. We rehabilitated the facial nerve defect using the great auricular nerve with an end-to-side nerve graft.

The facial nerve branches were sutured with epineural windows in the sidewall of the nerve graft. The facial nerve function of this patient recovered very well 6 months after the operation although the tumor was found to have recurred at this time point. Brachytherapy had no negative effect on the function of the facial nerve.

In conclusion, sialoblastoma is a rare, locally aggressive tumor that occurs perinatally or congenitally. Primary excision with negative margins is very important. $^{125}$I seed implant brachytherapy may be a reasonable alternative for treatment in the event of the tumor recurring, and, importantly, facial nerve function should be considered.

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