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Sialoblastoma: a congenital epithelial tumor of the salivary gland

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Abstract Sialoblastoma is a rare congenital epithelial tumor of the salivary gland that is diagnosed at birth or shortly thereafter with significant variability in histologic range and clinical course; hence, for an individual case, it may be difficult to predict the most appropriate therapy [*Cancer* 1972;30:459-69; *Pediatr Pathol* 1988;8:447-52; *Br J Plast Surg* 2000;53(8):697-699]. We report the case of a 4-year-old girl who had a widely spreading sialoblastoma of the left cheek. We were obligated to widely resect the tumor including the trunk of the facial nerve, superior part of the left maxilla, and the zygoma. Although the patient was operated 3 times in 4 years, invasion of the tumor could not be stopped. The patient died because of respiratory insufficiency and deterioration of her general health.

Sialoblastoma, which is also called "embryoma," represents a neoplastic proliferation of cells of organ rudiments [1], and it is the most frequent type of congenital epithelial tumor of the salivary glands [2]. Sialoblastoma have been reported to occur predominantly in the parotid gland and very rarely in the submandibular gland [3]. Although it is a slowly spreading tumor, late diagnosis can cause the tumor to spread widely and enlarge the limits of the operation to resect the tumor [1,4].

1. Case report

We report the case of a 4-year-old girl. First, her mother noticed a soft, 2-cm mass in the left cheek at the age of 1. When she was admitted to another plastic surgery clinic, her mother was told that the mass was a benign lesion that was probably a hemangioma, and no treatment was needed until the age of 5; the mass was expected to become smaller and spontaneously resolve eventually. At the age of 3, blunt trauma to the mass caused purplish color changes and some enlargement. When we examined the patient at 4 years of age, the mass was 5 cm in diameter. On examination, the mass was infiltrating the left cheek and elevating the skin overlying it. It was fixed to the maxilla, hard, and lobulated. Two lymph nodes were palpated, 0.5 and 1 cm in diameter, moderately hard, and mobile at the left submandibular region (Fig. 1A, B). Computerized tomography and magnetic resonance imaging (MRI) evaluation showed that the tumor infiltrated the left cheek, maxilla, infraorbital region, zygoma, and left temporal fossa (Fig. 2). We performed an incisional biopsy to the tumor under general anesthesia, and the diagnosis was sialoblastoma. Microscopic evaluation revealed that the tumor mass was surrounded by a tensile, connective tissue and consisted of epithelial cells with small ducts (Fig. 3). The tumor was

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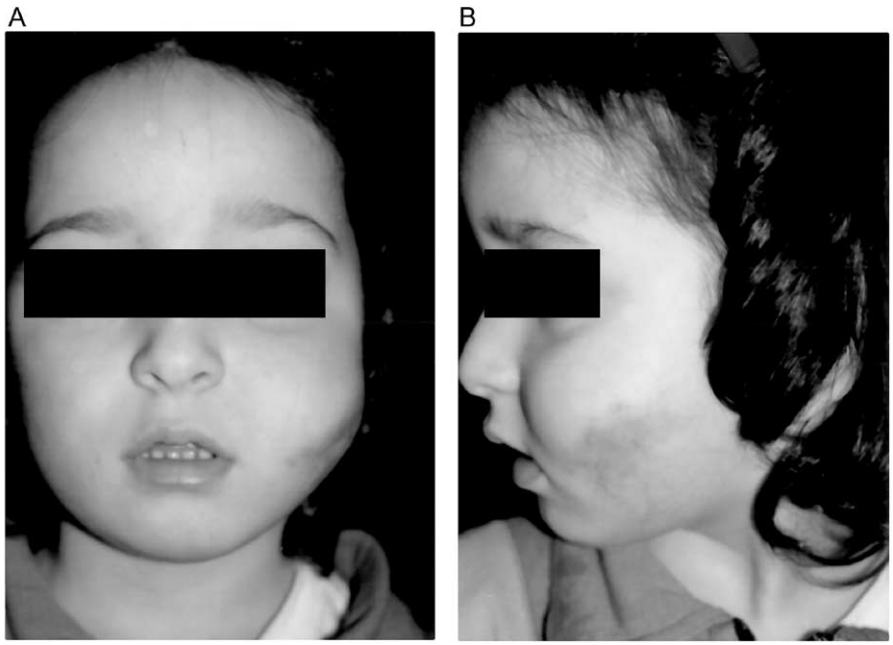

Fig. 1 The mass infiltrated the left cheek and elevated the overlying skin. A, Anterior image. B, Lateral image.

resected with the skin (8 cm in diameter), left parotid, superior part of the left maxilla and the left orbital floor, left zygoma, and left buccal mucosa. The part of the tumor infiltrating the temporal fossa through a passage under zygomatic arch was resected with the zygomatic arch as well. The facial nerve was cut before entering the parotid. On frozen section evaluation, margins of the resection were revealed to be tumor-free. Reconstruction was done by a free scapula fasciocutaneous and serratus muscle combined flap while reconstructing the orbital floor by cortical iliac bone graft stabilized by microplates. The left buccal mucosa was formed by the epimysium of the serratus muscle. We have not observed any complication, except partial graft loss at the donor side of the scapular flap.

The biopsy specimens were routinely fixed in formaldehyde and embedded in paraffin wax. Several stains were used, including hematoxylin-eosin (H&E), periodic acid-Schiff reaction, and Alcian blue (pH 2.5). In addition, the cases were studied immunohistochemically, by staining for myosin, muscle-specific actin, periodic acid schiff (PAS), cytokeratins, epithelial membrane antigen (EMA), vimetin, S100 protein, and neuron-specific enolase (NSE). H&E–stained sections showed that the tumor was characterized by solid cell nests of epithelial cells intermingled with proliferating ductal structures and multiple trabecular formation. All cells contained small nucleus, eosinophilic cytoplasma with atypia. Both ducts and acini were surrounded by fibrous stroma. These appearances were uniform throughout the lesion; hemorrhage and necrosis were present. Cytokeratin immunoperoxidase staining showed strong positivity off cells forming ductal structures. Antibodies to EMA showed weak staining of ductal tumor

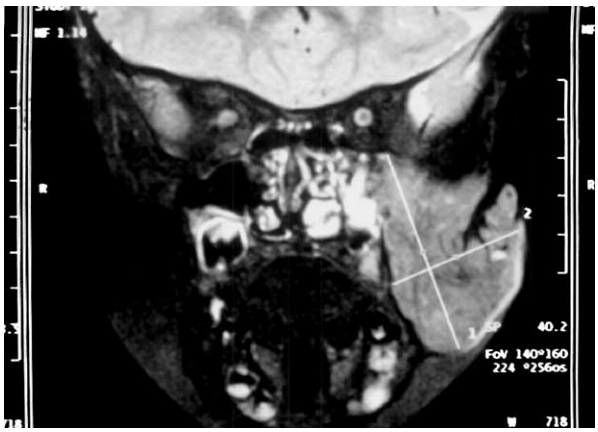

Fig. 2 Sialoblastoma infiltrated the left cheek, maxilla, infraorbital region, zygoma, and left temporal fossa.

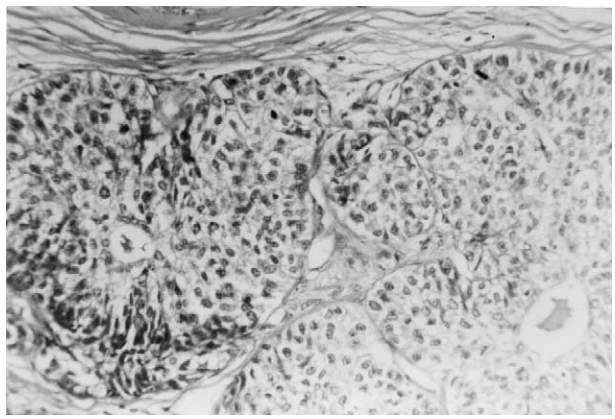

Fig. 3 The tumor mass was surrounded by a tensile, connective tissue and consists of epithelial cells with small ducts (H&E, 125).