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Solitary neurofibroma arising from the infratemporal fossa in a child☆

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Abstract Neurofibromas are derived from the nerve sheath and are commonly located in the head and neck region. They usually occur between the ages of 30 and 50 years. Neurofibromas arising from the infratemporal fossa are quite rare, especially in children. We describe a solitary neurofibroma arising from the infratemporal fossa in an 8-year-old boy who presented with a painless mass in his right cheek. Computed tomographic scan showed a soft-density, not well-circumscribed mass located in the right infratemporal fossa. The tumor was resected via the transmandibular approach with an excellent outcome. The histologic examination with immunohistochemical staining yielded the diagnosis of neurofibroma.

Pediatric tumors of head and neck usually present as congenital and malignant lesions. In the sinonasal region, the most common malignancy is rhabdomyosarcoma, which often extends to the orbit and the face. In the neck, the most common cause of primary malignant disease is lymphoma [1]. Neurofibroma, which is usually associated with neurofibromatosis type I (NF-1), is a common neurologic tumor of the head and neck region. Here, we report a solitary neurofibroma arising from the infratemporal fossa in a child without the stigmas of NF-1. The lateral mandibular osteotomy approach was used to resect the tumor.

1. Case report

An 8-year-old boy was admitted to our hospital because of a painless right cheek mass for 1 month. Five years ago, he received surgical treatment of a mass in the same region, which was diagnosed as neurofibromatosis. However, other signs of neurofibromatosis were not detected and no family history of neurofibromatosis was obtained.

On physical examination, the patient presented with facial asymmetry (Fig. 1). Intraorally, a nontender mass in right pterygomandibular ligament was noted. The buccal mucosa was normal without any signs of inflammation. Computed

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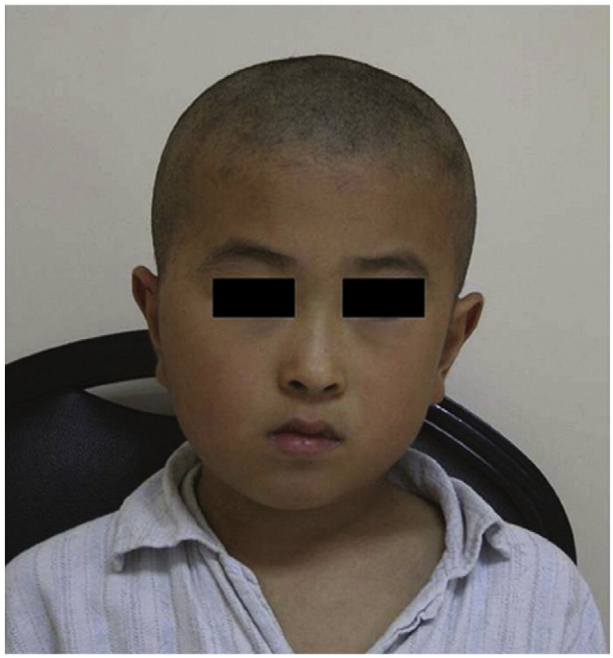

Fig. 1 Preoperative view of an 8-year-old boy with facial asymmetry.

tomographic (CT) scan detected a soft tissue density mass located in the infratemporal fossa. This tumor extended superiorly to the skull base, inferiorly to the level of the angle of the mandible and masseter muscle, anteriorly to the lateral wall of maxillary sinus, and medially near the lateral pterygoid and lateral pterygoid process (Figs. 2 and 3). The initial clinical diagnosis was myofibroblastic disease.

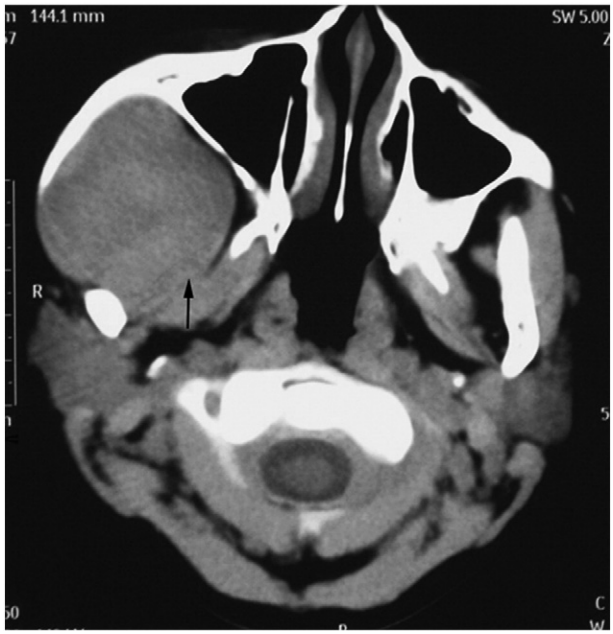

Fig. 2 Axial CT scan showed a soft-density, not well-circumscribed mass located in the right infratemporal fossa. The mass did not have a clear margin separate from the lateral pterygoid (arrow).

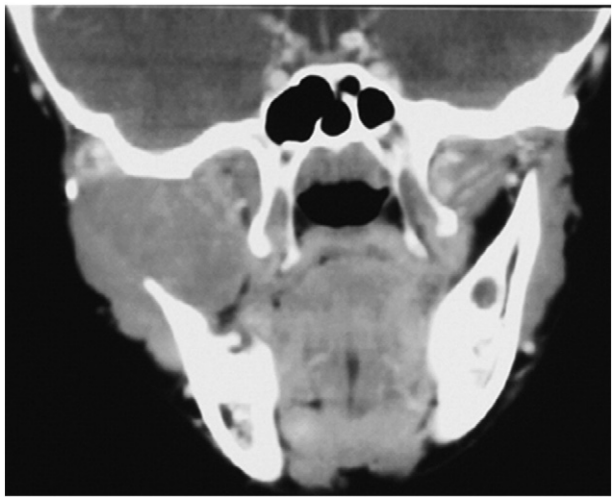

Fig. 3 Coronal CT scan demonstrated that the mass was medial to the external pterygoid process and extended to the skull base, no intracranial extension.

The tumor was successfully removed via a transmandibular approach under general anesthesia. A curvilinear incision was developed from the median lower lip to the preauricula through the submandibular region (Fig. 4). A right lower lip and buccal soft tissue flap were elevated. An osteotomy was performed in front of the angle of the mandible to provide wide exposure of the mass. The mass and coronoid process were resected (Fig. 5). After removing the tumor, the mandible was returned to the original position and fixed by a titanium plate.

Grossly, the mass was yellowish in color when cut and not well circumscribed (Fig. 6). The histologic examination on paraffin sections indicated that this was a neurofibroma (Fig. 7). Immunohistochemical examination showed that the tumor cells were strongly positive for vimentin staining and part of the cells were positive for S-100 and CD57, which was consistent with a neurogenic origin.

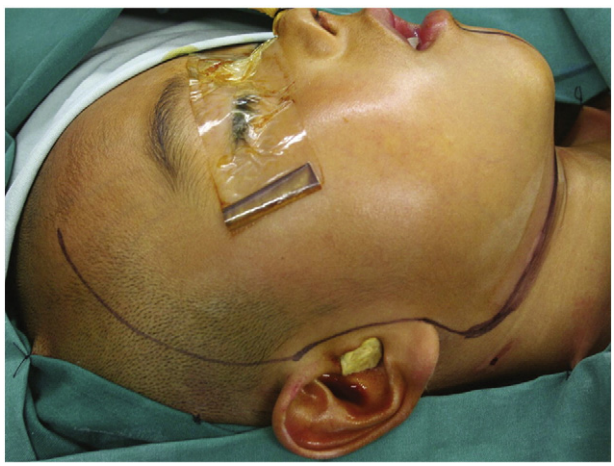

Fig. 4 A curvilinear incision was developed from the median lower lip to the preauricular through submandibular region.