International Journal of Pediatric Otorhinolaryngology Extra 13 (2016) 19–22

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology Extra

journal homepage: http://www.ijporlextra.com/

Case report

Sialoblastoma: A very rare cervical mass in neonates: A case report

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ARTICLE INFO

Article history:
Received 21 December 2015
Received in revised form
21 February 2016
Accepted 26 February 2016
Available online 4 March 2016

Keywords:
Alpha-fetoprotein
Submandibulectomy
Behavior
Local recurrence
Metastasis

ABSTRACT

Sialoblastomas are very rare epithelial tumors of the salivary gland that usually occur in the parotid or submandibular gland. Most sialoblastomas present in the neonatal period or early childhood. The biological behavior of these tumors is variable, with a potential for local and systemic recurrences. Only ten cases in the submandibular gland have been reported so far. We are reporting a case of sialoblastoma in a newborn male child presenting in the right submandibular region. Ultrasound visualized a homogeneous solid mass. MRI showed a well-defined soft tissue solid mass in right submandibular region. Alpha fetoprotein was 18.8 ng/ml (*normal 0.4–15*). Fine needle aspiration showed cylindrical connective tissue with groups of basilar cells. Mitosis was not seen. The most probable diagnosis was sialoblastoma.

1. Introduction

Sialoblastoma is the most common epithelial tumor of the salivary gland. Tumors of the salivary gland are rare in childhood, accounting for only 3–5% of all tumors. We reviewed the literature in English and found that only 51 cases of sialoblastoma/embryoma/congenital basal adenoma have been reported so far. The most common site of involvement was the parotid gland, with only ten cases in the submandibular gland. We are reporting a case of sialoblastoma in a newborn male child presenting in the right submandibular region.

2. Case report

A one-day-old male infant presented with a firm, large lump in the right submandibular region. He was born at 40+ weeks by cesarean section delivery due to face presentation and bradycardia. The obstetric history was unremarkable. Soon after birth, he was noted to have a firm, nodular mass in the right submandibular region fixed to the deeper structure but not to the overlying skin [Fig. 1].

In ultrasound, a homogeneous solid mass measuring about 1.8 × 3.5 cm was visualized.

The abdominal ultrasound was unremarkable, the alpha fetoprotein was 18,784 ng/ml (normal 0.4–15).

Magnetic resonance imaging (MRI) of the head and neck showed a well-defined soft-tissue solid mass in the right submandibular region with maximum diameter 3.5 cm, with moderate vascularization, not connected to the tongue or larynx. The mass was similar in texture to the left submandibular salivary gland which was not visualized on the right side [Fig. 2]. The chest X-ray was normal.

Fine needle aspiration (FNA) was performed and showed cylindrical connective tissue with groups of basilar cells. Mitosis was not seen. The most probable diagnosis was sialoblastoma.

A total submandibulectomy was performed on the 16th day of life with no difficulties. The post-operative course was unremarkable. No facial deficit was noted. Alpha fetoprotein decreased to the normal level for the age. The follow-ups have shown no recurrence up to the present time (27 months later).

On pathology examination, grossly, the tumor was well circumscribed, nodular, 12 g in weight, 3.7 × 2.3 × 2 cm in size. Microscopically, the resected tumor showed nests of primitive basaloid cells with variably peripheral palisading, separated by loose fibrous connective tissue [Fig. 3]. The round or basaloid cells had hyperchromatic nuclei, a moderate amount of cytoplasm and demonstrated minimal mitosis (1 mitosis/hpf) [Fig. 4]. In addition to the solid nests of basaloid cells, also present were immature ductular and acinar structures. The tumor had a thin fibrous capsule. Neither lympho-vascular nor neural invasion was identified. Immunostains for P53 and alpha fetoprotein immunostaining were negative. Ki-67, a proliferation marker, was positive in approximately 10–15% of the tumor cells [Fig. 5]. The surgical margins were uninvolved by the tumor; the distance of the tumor

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http://dx.doi.org/10.1016/j.pedex.2016.02.006
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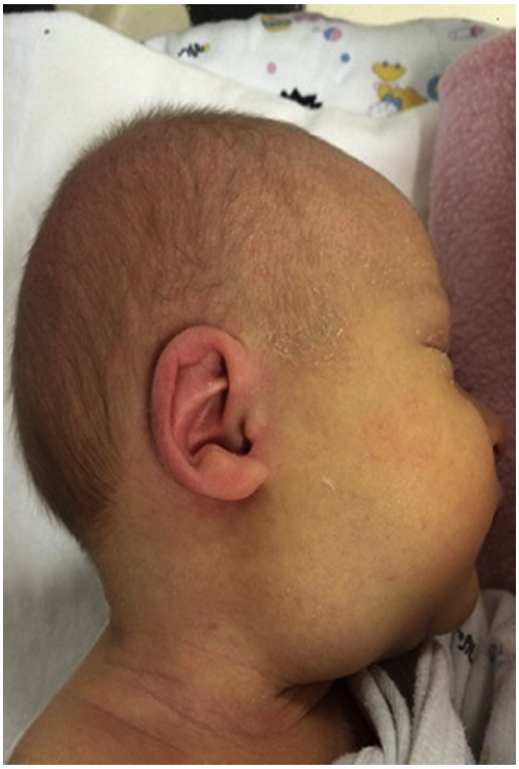

Fig. 1. Neonate with mass in the right submandibular region.

to the closest margin was 4 mm. Based on these findings the mass was confirmed as a sialoblastoma.

3. Discussion

Sialoblastoma, a tumor of the salivary gland is a rare neoplasm initially reported by Vawter and Tefft in 1966 as 'embryoma' [1].

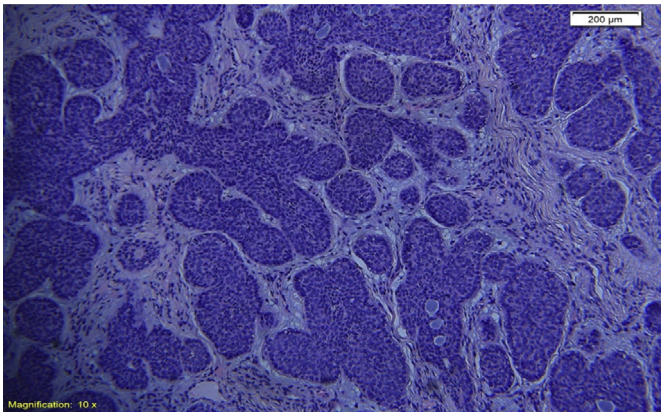

Fig. 3. Most of the tumor showed a solid nest of basaloid cells separated by loose connectivetissue. Some ductular structures were also present.

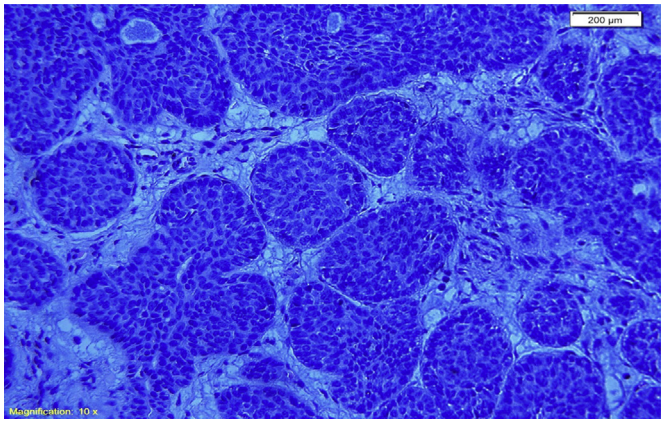

Fig. 4. The round or basaloid cells had hyperchromatic nuclei, a moderate amount of cytoplasm, and demonstrated a small number of mitosis (1 mitosis/hpf).

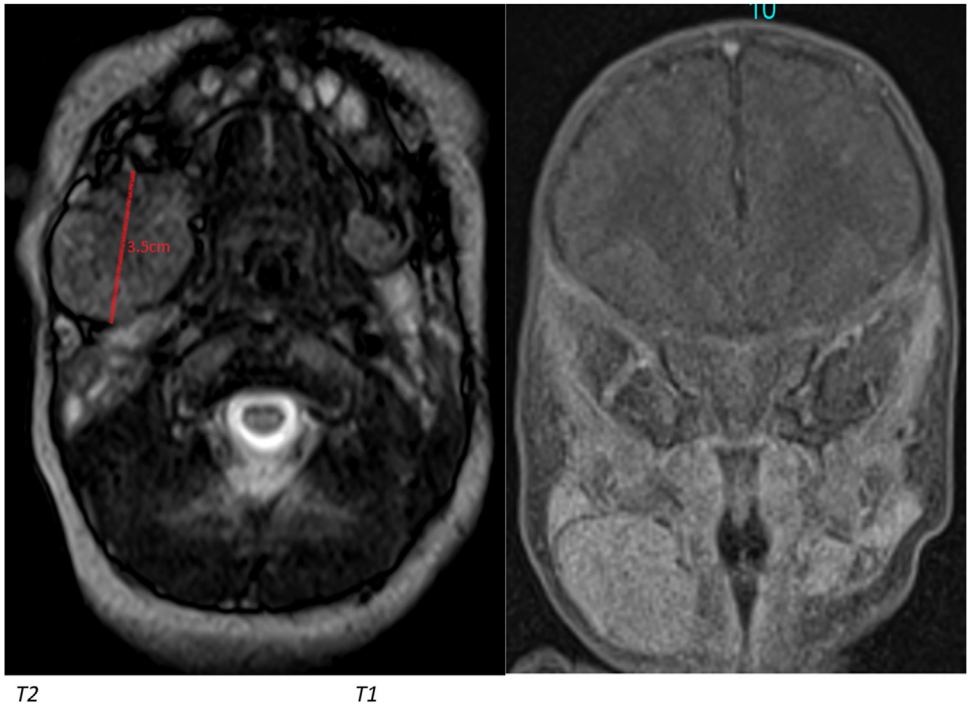

Fig. 2. Magnetic resonance imaging (MRI) neck. Well-defined soft tissue mass in right submandibular region.