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Case Report

Sialoblastoma: A Very Rare Cervical Mass in Neonates - A Case Report

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Abstract: Sialoblastoma is a rare neonatal epithelial tumor of salivary gland that generally originates in the parotid gland. Previously it was believed that this tumor did not cause systemic metastasis, and only had local recurrence. However recently it was reported to cause systemic metastasis. For this reason, sialoblastomas are long-term follow-up recommended. Our case was initially diagnosed by intrauterine ultrasonography as a cervical mass. Here we discuss the preoperative investigations, surgical management, histopathological examination and long-term follow-up with review of the literature.
Keywords: Neonate, Cervical Mass, Sialoblastoma

INTRODUCTION

Sialoblastoma is an epithelial tumor of salivary gland origin. Tumors of the salivary gland are rare in childhood, accounting for only 3-5% of all tumors. Most such lesions present after the age of 5 years and there is an increasing incidence over the first two decades of life [1, 2, 3]. Perinatal, congenital and neonatal salivary gland tumors are exceptionally rare [4, 5]. The early treatment for sialoblastoma is recommended [6, 7]. We report a case presenting with a prenatal cervical mass during the neonatal period, which was later diagnosed as sialoblastoma.

CASE REPORT

A female neonate was born to a 28 year-old at 36 weeks of gestation and was diagnosed by intrauterine ultrasound as having a cervical mass. A right lateral face, neck and preauricular mass were diagnosed after birth. A mass measuring 10x9x6 cm was present. The mass was covered with thin and erithematous skin. The neck, face, mouth, right eye and ear were distorted by the mass (Fig. 1). At the hematologic examination the ßhCG level was 32000 IU/ L. Magnetic resonance imaging (MRI) scans demonstrated a lobulated, noninfiltrating mass, approximately 8 cm in greatest dimension in the right facio-cervical region. The tumor arose from the parotid gland and extended into the floor of the mouth and the parapharyngeal space. After administration of contrast material, the lesion showed homogenous enhancement. The ramus of the mandibula, the masseter muscle, the carotid arteries, and the jugular veins, showed no evidence of invasion by this tumor. There was no mediastinal or intracranial extension. Abdominal Computed tomography was normal.

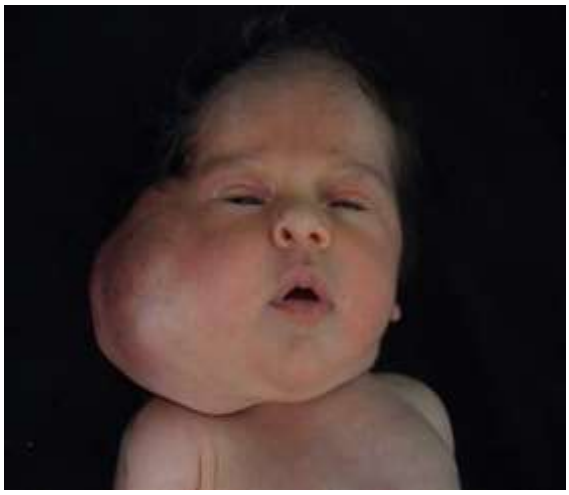

Fig. 1: Anterior view of the tumor. Neonate with extensive mass distorting neck ear, eye and mouth

The tumor was excised under general anesthesia at the 10th day. The tumor was attached to the inferomedial aspect of the mandible, and extended into the floor of the mouth and the parapharyngeal space. The tumor was multilobular and had a thin capsule. There was a clear plane between the tumor and surrounding normal tissues. The facial nerve was preserved during tumor excision. There were no complications early postoperatively. The baby was fed on the 2nd postoperative day and was discharged on the 7th postoperative day. Histopathologic examination was done. Macroscopically, the mass was 9x5x4 cm, nodular, pale brown coloured, well demarcated, with a thin capsule. Histologically, the appearance of the tumor reflected stages during the embryonic development of salivary gland. It was composed of lobulary solid nests of primitive epithelial cells separated by loose fibromyxomatous stroma. Isolated cells with sebaceouse differentiation were found. Formative ducts and pseudoductular spaces, without

acinar diferentiation were seen in a vascular rich loose mesenchyme with an embryonic appearance. There were mitotic activity average 7-8/10 high power fields, focal necrotic and hemorrhagic areas (Fig. 2).

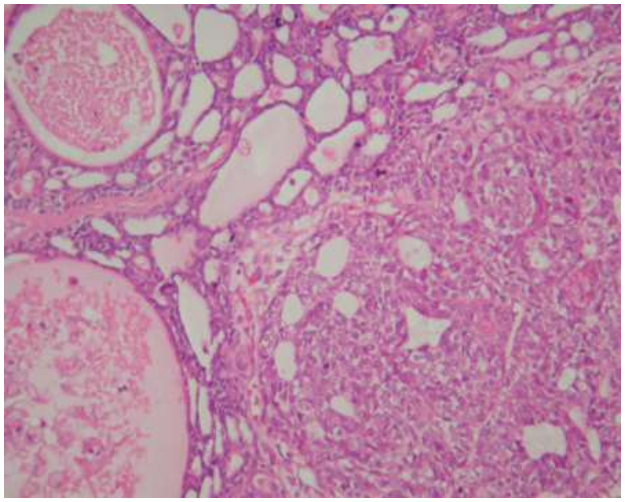

Fig. 2: Microscopic apperance; Embryonic appearing cell groups, basaloid cells, ducts and pseudoductular spaces in a loose primitive masenchyme are seen in a tumor (X200, H-E)

In the first postoperative month, the region of the tumor was examined by USG. Ultrasonographic and physical examininations were normal. At the third month cervical MR, thoracal and abdominal CT, physical examination and ßhCG level were all normal. This baby was followed with physicial examination and chest X-ray at 3 months period in first year. Than she was followed at 6 months period after first year. She is seven years old and still normal that she has not local reccurance and systemic metatsasis.

DISCUSSION

Sialoblastoma, an embryoma of the salivary gland is a rare neoplasm that was initially reported by Vawter and Tefft in 1966 [1, 4]. Developing in the major salivary glands, sialoblastoma has a predilection for the parotid gland [4]. The vast majority of cases present in the second decade of life. This tumor is occasionally rare [2, 4].

Radiologically, a definitive diagnosis of sialoblastoma cannot be made preoperatively by imaging techniques [1, 8]. Hemangiomas should be considered in the differential diagnosis of this tumor, as they also show marked contrast enhancement. Clinical examination and laboratory studies are not definitely diagnostic for sialoblastoma. An exact diagnosis is made with pathological examination. Differential diagnosis should consider other major perinatal neoplasms found in the salivary glands: congenital basal cell adenomas, embryonal carcinoma, and congenital hybrid basal cell adenoma-adenoid cystic carcinoma [4, 8]. Accompanying anomalies have not been reported except hepatoblastoma [4]. For this reason, especially abdominal examination should be carried out before operation. Sialoblastoma can cause respiratory distress by pressure on the trachea and bleeding secondary to tumor eruption [1, 2]. For these reasons, the surgeon must be ready for these emergencies.

Fetal malformations that distort the architecture of the face can be either cystic or solid. Hemangioma, lymphangioma, teratoma, neurofibroma, cystic hygroma, and retinoblastoma should be considered in differential diagnosis [4]. Sialoblastoma is generally of a benign nature so that total surgical excision is adequate. It is reported that the tumor is generally locally agressive hence there is no need for chemotherapy and radiotherapy, and local excision is adequate for local recurrence [1, 2, 4, 6]. However, this benign behaviour is not thought to spread systemically [1], because, it has been reported that sialoblastoma is aggressive and a potentially malignant tumor [7, 9]. A sialoblastoma case with nodal excision from the lung revealed lung metastasis and was treated with chemotherapy and radiation [7, 9]. The long-term biological behaviour of this tumor is uncertain. Although acknowledged cases of this occasionally and infrequent neonatal tumor are very rare, it is thought that total surgical excision is enough and local excision may be required for local recurrence. Nevertheless, the possibility of systemic metastasis should not be forgotten. These tumors have not specific tumor markers therefore ıt is followed physicial examination and chest x-ray. If there are suspect of metastasis or local reccurence, Surgeon or physician should advanced examination.

In summary: Sialoblastoma is a very rare neonatal tumor of the salivary gland, mainly parotid origin. Behaviour of this tumor may vary from the early to the late period. We have planned a long-term follow-up for local recurrence and systemic metastasis.

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