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Case report

Management and treatment of a sialoblastoma of the submandibular gland in a neonate: Report of one case

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ABSTRACT

Objectives: Report a rare congenital salivary gland tumor and its clinical, radiological and histopathologic aspects. Discuss the differential diagnosis and treatment.

Methods: A case report is described. A literature review of the incidence, disease course, radiological and histopathologic aspects and treatment are presented.

Results: This case report presents a neonate with an asymptomatic left cervical mass. Ultrasounds and MRI showed a circumscribed soft tissue mass measuring 60 mm × 42 mm × 52 mm and extending from the left parotid region to the left submandibular region, but radiological exams could not lead to a diagnosis. A biopsy demonstrated a sialoblastoma. The treatment consisted in a surgical resection with no adjuvant therapy. One year after surgery there is no clinical nor radiological sign of recurrence.

Conclusion: Sialoblastomas are rare congenital epithelial salivary tumors and have been reported to occur predominantly in the parotid gland. They are locally aggressive with a high recurrence rate needing a prolonged follow up. When they are completely resectable, surgical resection is the mainstay for treatment of these tumors and no adjuvant therapy is needed.

1. Introduction

Less than 3% of salivary gland tumors occur in children and the vast majority occur in the second decade of life [1]. During the first year of life, hemangiomas are the most common. Sialoblastomas have been reported to occur predominantly in the parotid gland [2,3]. We report one case of sialoblastoma of the submandibular gland; we describe its clinical, radiological and histopathologic features and discuss the differential diagnosis and treatment.

2. Case report

A 3-day-old girl was referred to our department after birth for a left cervical mass that had already been detected by antenatal echography and foetal Magnetic Resonance Imaging (MRI) (Fig. 1). The prenatal radiological aspect was in favour of a congenital hemangioma.

The little girl weighing 3400 g was born spontaneously at term by vaginal route. Examination revealed a large hard mobile mass going from the left inferior parotid region to the left submandibular region (Fig. 2). The swelling did not have any mass effect or inflammation; the airway at birth was not compromised. There was no facial palsy. An echography was performed by the same radiologist who had already performed the first exams and that is specialized in vascular malformations. As all the echographic and clinical criterias of hemangioma were not met, an MRI was realized. The MRI showed a well circumscribed soft tissue mass which dimensions were 60 mm × 42 mm × 52 mm with a parapharyngeal extension. The tumor was isointense to muscle on T1-weighted images with a post-contrast enhancement; on T2-weighted images the lesion had high-intermediate signal intensity (Fig. 3).

As some of the clinical and radiological aspects were not typical of hemangioma, a surgical biopsy was decided for excluding a sarcomatous lesion like congenital rhabdomyosarcoma or fibrosarcoma. The mass was diagnosed as sialoblastoma by the histopathologists.

A CT scan without contrast injection was realised to better evaluate the relations of the tumor with mandible. It showed a soft tissue mass isodense to muscle with no sign of osteolysis or bone invasion.

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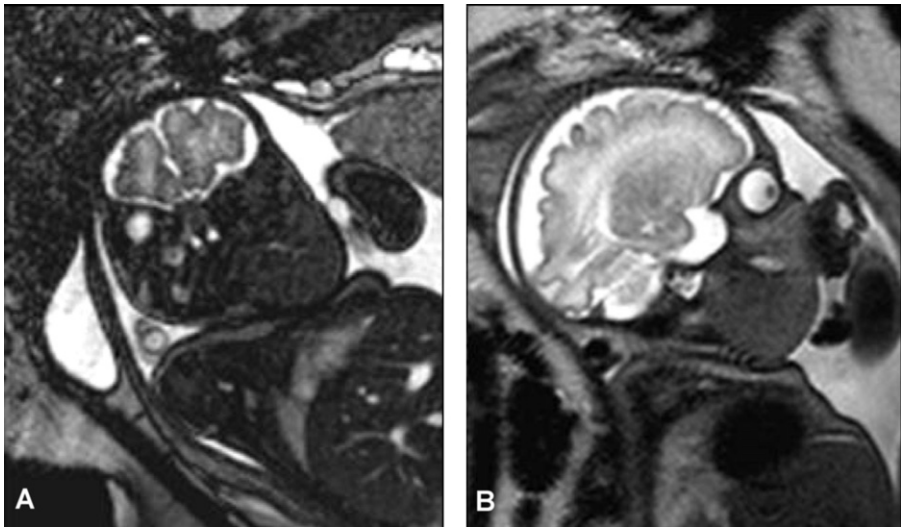

Fig. 1. T2-weighted foetal MRI showing the large left cervical tumor: coronal (A) and saggital (B) views.

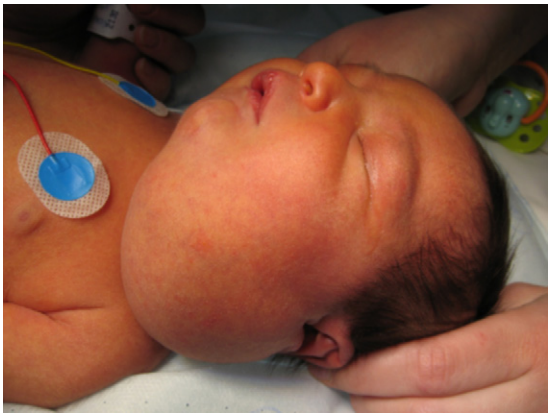

Fig. 2. Photography of the 3-day-old girl showing the large swelling going from the left parotid region to the left submandibular region.

A surgical treatment was decided during a multidisciplinary consultation including a pediatric cancerologist, two ENT surgeons, a radiologist and a neonatologist, indeed, based on MRI and CT scan findings, the mass appeared suitable for total resection. Excision was made under general anaesthesia through a parotid-type incision with facial nerve monitoring at 10 days of age. The mass was found to be well encapsulated, distinct though adjacent to the left parotid gland. The facial nerve trunk was identified and preserved, so was its buccal branch that was adherent to the tumor. Tumor was found to arise from the left submandibular gland; therefore a monobloc resection was performed (Fig. 4).

Macroscopic examination revealed a circumscribed, firm, multilobulated, orange or beige solid mass measuring 7.5 cm × 4.5 cm × 6 cm and weighing 69 g (Fig. 5). Microscopic examination after hematoxylin eosin saffron staining showed a tumor composed of islands and nests of epithelial cells with basaloid appearance separated by bands of fibrous stroma (Fig. 6). The mitotic rate was 10 per 10 high power fields and many apoptotic bodies were also present. No marked cytologic atypia, no necrosis nor perineural or vascular invasion were identified. Tumor cells were immunohistochemically reactive for Cytokeratin AE1/AE3, 7, 18 and 19 and Vimentin. Epithelial membrane antigen (EMA) and S100 protein also showed a week staining. The cells were negative for α-foetoprotein, smooth muscle antigen (SMA), glial acid fibrillary protein (GFAP) and Cytokeratin 20. The Ki67 index was approximately 30% (Fig. 7).

The postoperative course was uneventful and the baby was discharged 1 week after surgery. No adjuvant therapy was

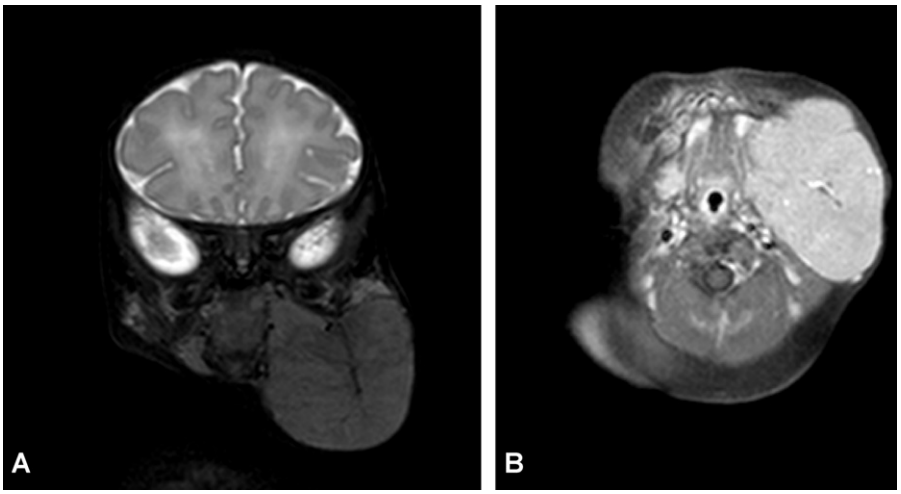

Fig. 3. (A) T2-weighted coronal view showing an intermediate signal intensity of the tumor; and (B) axial post-gadolinium T1-weighted images showing a rather high contrast enhancement of the mass.