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LETTER TO EDITOR

Congenital orofacial teratoma in a 22-week foetus

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Dear Editor,

We report a case of congenital orofacial teratoma also known as epignathus. It is rare and is exclusively seen in neonates. A 34-year-old female, G2P1 at 22 weeks and 2 days period of amenorrhea presented to the antenatal clinic for routine examination. An ultrasound examination revealed a solid-cystic facial tumour at the growing foetus. The tumour measures 7.1 x 4.3 x 5.3 cm, with intracranial extension and occupied the left-brain hemisphere with a midline shift. The mother had a history of gestational diabetes mellitus on diet control and hyperthyroidism on propylthiouracil. She was also found to be anaemic. Termination of pregnancy was performed and the foetus was delivered stillborn.

Examination of the foetus showed a widely expanded mouth due to a large haemorrhagic, solid and cystic tumour protruding from within the oral cavity, measuring 7.0 x 5.5 x 2.0 cm (FIG 1A). The tumour was soft to firm in consistency and weighed 60 grams. A few translucent cysts filled with clear fluids are observed. The cut section of the mass showed variegated, white to greyish and haemorrhagic surface. Bony structure was also present. There was no macroglossia of the tongue. Also identified was a separate intracranial tumour which compressed the left hemisphere of the brain, measuring 6.0 x 5.0 x 3.0 cm and weighed 60 grams (FIG 1B). Intriguingly, the base of skull was intact. We were unable to find any communication with the intraoral tumour. The brain was compressed and distorted by the tumour. Microscopically, both the orofacial and intracranial tumour were similar morphologically and showed immature teratomas, Grade 3, with predominantly immature glial tissue (FIG 1C). There was no other structural abnormality of the foetus. The examination of placenta was fairly unremarkable, except for a mild hypercoiled cord. Congenital orofacial teratoma is seen in about 1:35,000–1:200,000 live births.[1,2] It may be associated with malformations such as cleft palate, bifid tongue, and bifid uvula.[1] It has to be detected early to prepare for intervention, in order to prevent fatal airway obstruction at the immediately postpartum period. The possible differential diagnoses of orofacial tumour include congenital epulis, basal cephalocele, dermoid, lymphatic malformation and rhabdomyosarcoma. On the other hand, a prenatal intracranial tumour includes teratoma, neuroepithelial tumour and craniopharyngioma.[1,3] Based on a literature review by Kirishima *et al*.[1] on 14 cases of epignathus with intracranial involvement, the tumour size ranges from 2 to 23 cm, the mother age ranges from 18 to 32 years old, and 4 cases with intracranial involvement did not have a direct connection between the intracranial and orofacial tumours. As an epignathus with intracranial extension could be encountered and it carries a poor prognosis, it is crucial to perform a careful evaluation in suspected case for any intracranial extension.

Keywords: Congenital, epignathus, head and neck, perinatal postmortem, teratoma

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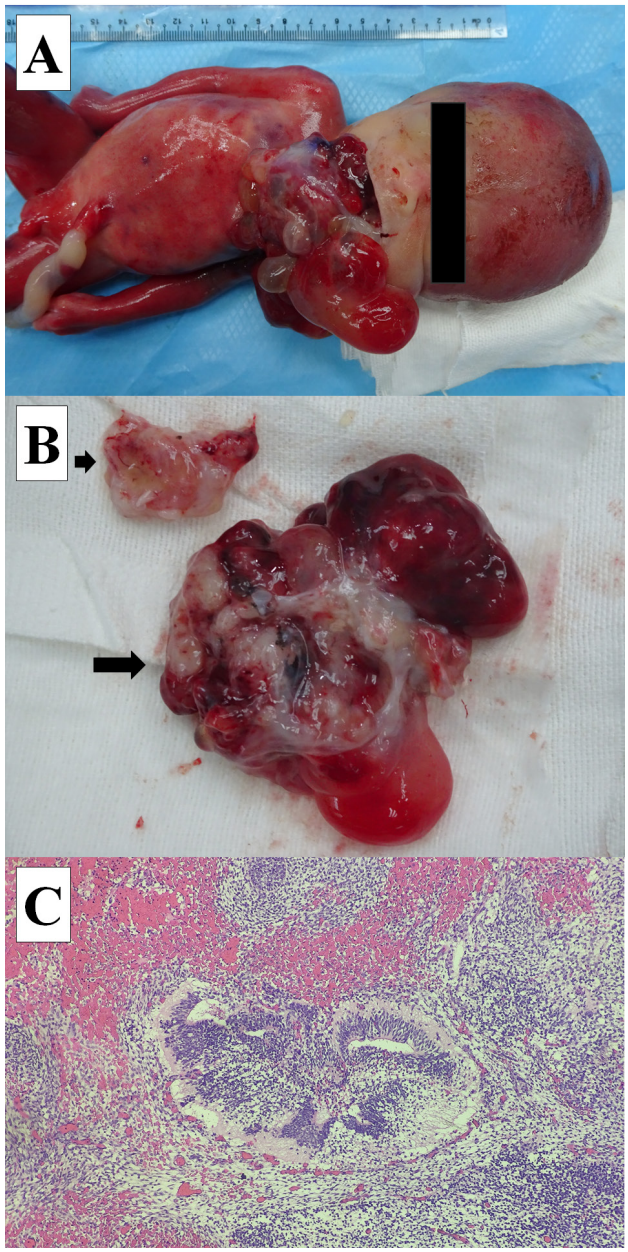

FIG. 1: A) A large solid-cystic intraoral mass protruded from the mouth. B) A solid-cystic tumour from the orofacial region (long arrow) and solid pale pink oedematous tumour from the intracranial region (short arrow). C) The tumour demonstrates multiple foci of immature tissue comprises of primitive neuroectodermal tubules with rosette-like formations.

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