

Prenatal diagnosis of epignathus in the first half of pregnancy: A case report and review of the literature

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Summary

Congenital teratomas of the oral cavity commonly present as tissue masses projecting from the mouth. The important prognostic determinants are: the age of presentation, technical problems during surgical removal of the mass, associated anomalies, and the nature of the composing tissues. This study reports one case of epignathus (an extremely rare oropharyngeal teratoma) that was diagnosed by ultrasonography at 19 weeks of gestation and reviews the relevant literature.

Key words: Nasopharyngeal teratomas; Prenatal diagnosis; Epignathus.

Introduction

Teratomas are neoplasms that originate in pluripotent cells and are composed of a wide diversity of tissues foreign to the organ or anatomic site in which they arise [1]. They occur in gonads and extragonadal sites and in various age groups. Head and neck teratomas constitute 5% of teratomas reported in children [2]. The term epignathus is often used for teratomas originating in and around the oropharynx, although etimologically implies a relationship to the jaw [1].

Prenatal ultrasonographic examination has resulted in the recognition of teratomas in the fetus.

Case Report

A 34-year-old healthy woman in her third pregnancy was referred to our unit at 19 weeks of gestation for ultrasound examination because the maternal serum α -fetoprotein (MS-AFP) was 6.1 multiples of the median (MoMS). Ultrasound examination demonstrated a singleton pregnancy and the fetal measurements were compatible with the gestational age calculated from the menstrual history. There was a large echogenic mass protruding from the fetal mouth (Fig. 1) but there were no other obvious fetal or placental abnormalities and the amniotic fluid volume was normal. Amniocentesis was performed and normal fetal karyotype was obtained. However, the amniotic fluid AFP was found to be 5.1 MoMS.

Following counselling the parents opted for termination of the pregnancy which was carried out by intravaginal administration of prostaglandins and intravenous oxytocin. Pathological examination of the aborted female fetus demonstrated a large (6x3x2 cm) solid mass protruding from the mouth and attached to the soft palate (Fig. 2). Histology showed a teratoma containing loose connective tissue, cartilage, bone, smooth muscle, seromucinous glands, squamous and gastrointestinal-type epithelium and neuroepithelial elements. There were no other external or internal abnormalities.



Figure 1. — Ultrasound picture demonstrating a large solid mass projecting from the fetal mouth.

Discussion

Congenital teratomas of the oral cavity commonly present as tissue masses projecting from the mouth. The obvious manifestation of large tumors related to the facial orifices is airway obstruction, which may be life-threatening. The tumors may attach to the roof of the pharynx, palate, maxillary bone or mucosa and may be associated with an intracranial teratoma. Some tumors exhibit striking organ development [1].

Ultrasonography can lead to prenatal diagnosis in about half of the cases, allowing for planned delivery and immediate neonatal respiratory support with subsequent surgical excision [4]. Appropriate surgical management has resulted in cure of even massive tumors of this area. The important prognostic determinants are the age of presentation, technical problems at the time of birth and surgery, associated anomalies, and the nature of the composing tissues [1]. It appears that the occurrence of



Figure 2. — The epignathus was extending through the open mouth and attached to the soft palate.

immature somatic tissues is not an indication of potentially malignant behaviour [2, 6].

A literature search identified 19 previous cases of epignathus that were diagnosed prenatally [7-9] and in five of these cases the tumor was detected during the first half of the pregnancy. In only three (15.8%) cases the diagnosis was made before and in 17 (84.2%) after 20 weeks. There were six terminations, three intrauterine and six neonatal deaths, and four survivors. The differential diagnosis of oropharyngeal masses includes gingival granular cell tumour, teratoma, salivary glands cyst,

hemangioma, lymphangioma, terinoblastoma, and neurofibroma [11]. Elevated amniotic fluid AFP, as found in our case, has previously been described in association with craniofacial teratomas [12].

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