

Nasopharyngeal Teratoma(Epignathus) —Four Autopsy cases—

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= Abstract =Four autopsy cases with nasopharyngeal teratoma were presented. Three cases were stillborns and one case survived for five minutes. All four cases showed a bulky protruding mass out of the oral cavity which arose in the hard palate. One case showed the destruction of the skull base and intracranial extension. Microscopically, the masses showed predominantly immature neuroepithelial components and some mature and immature elements of the parenchymal organs, such as liver, gastrointestinal tract and pancreas. One of the four cases was associated with unilateral renal agenesis. The characteristics and histogenesis of nasopharyngeal teratoma are discussed.

Key Words: *Nasopharyngeal teratoma, Epignathus, Fetus, Congenital*

INTRODUCTION

Nasopharyngeal teratoma is a rare congenital tumor which often shows an impressive and grotesque appearance of a large solid and cystic mass protruding from the oral cavity(Ahlfeld, 1875; Ewing, 1942; Ehrich 1945; Hatzihaberis *et al.*, 1978). It is commonly called "epignathus," that is, "epi" plus "gnathos" meaning jaw in Greek. There has been a dispute whether it is teratoma or parasitic twin because it shows well-differentiated gross features or histology and occasionally is connected to the host(autosite) by an umbilical cord(Ahlfeld 1875; Ewing 1942; Ehrich 1945). Most of the fetuses bearing a nasopharyngeal teratoma or an epignathous tumor are stillborn and only rarely occur in children over the age of two(Batsakis, 1979; Gnepp 1986). The tumor occasionally extends to the intracranium. Most nasopharyngeal teratomas contain an immature somatic structure, especially primitive neuroepithelial components. However, true malignancy in a child has never been reported(Foxwell & Kelham 1958; Calcaterra, 1969; Hawkins & Park, 1972; Budrawy *et al.*, 1973). In

view of the rarity of this entity, we present four autopsy cases with congenital nasopharyngeal teratoma and also discuss the histogenesis.

CASE REPORT

Case 1

This case was a 32-week-old male fetus who was prematurely delivered vaginally and survived for five minutes. During antenatal care, maternal hydramnios was noted. The birth weight was 2200gm. On autopsy, a huge protruding mass from the oral cavity was seen, measuring 12.5 × 10.6 × 7.5cm (Fig. 1). The mass was multi-lobulated and partially cystic and partially solid. However, no human-like gross appearance was seen. On sagittal section, the mass was attached to the midline of the hard palate. Agenesis of the right kidney, an accessory spleen and an undescended testis were found in the abdominal cavity. Microscopically, the epignathus was teratoma consisting of primitive neural tubes, glial tissue, choroid plexus, liver, spleen, gastrointestinal tract wall, retinal pigment epithelium, salivary glands, bones, teeth and skin (Fig. 2 & 3). There was no axial or appendicular



Fig. 1. Gross picture of Case 1 shows a huge protruding mass from the oral cavity.

skeleton.

Case 2

This 21-week-old male fetus was terminated due to fetal mass and hydramnios detected by sonography. He was the first child of a 23-year-old mother who had taken an unidentified drug for pyelonephritis during the seventh week of gestation. On autopsy, a huge irregular mass was attached to the midline of the hard palate and sphenoid bone, measuring $12 \times 11 \times 10$ cm. The mass protruded from the oral cavity. On midsagittal section, the tumor filled the oral cavity and extended to the intracranium (Fig. 5). The intracranial part of the tumor measured $10 \times 10 \times 9$ cm. The brain tissue could be well demarcated from the tumor and was compressed bilaterally against the parietal and temporal bones. Microscopically, both the oral and intracranial portion of the tumor was covered by squamous epithelium with immature hair follicles

and contained a mature digestive tract, respiratory tract with bronchial cartilages, liver tissue with extramedullary hemopoiesis, pancreas tissue, lung tissue, kidney tissue, bony islands, a finger-like structure containing chondroid tissue (Fig. 6), retinal pigment epithelium and immature and mature nervous tissue. The immature nervous tissue, consisting of primitive neural tubules, were the predominant component (Fig. 7). No other fetal anomaly was associated. There was no vertebral column. The chromosome studies from the peripheral blood of the parents and the cord blood of the fetus were done. The father showed a normal male karyotype with 46,XY. However, both mother and fetus showed pericentric inversion of chromosome 9 with 46,XY, -9, + inv(q) (p11q21).

Case 3

This 24-week-old female was terminated due to a huge fetal mass and maternal polyhydramnios. This was the third pregnancy of this 29-year-old mother. Previously, one of her two older sisters and a congenital hip dislocation. On autopsy, the fetus was 480gm without the mass. A small mass was found in the midline of the palate (Fig. 8 & 9), and a huge multilobulated mass was submitted separately which was detached from the remaining mass attached to the hard palate. It measured $13 \times 12 \times 5$ cm and weighted 330gm. On sagittal section, the mass did not involve the intracranial region. Microscopically, the pharyngeal mass was composed of predominantly primitive neuroepithelial tissue and variously mixed mature tissue including glial tissue, choroid plexus, hepatic parenchyma with gallbladder (Fig. 10), pancreas, gastrointestinal tract, respiratory tract with bronchial cartilages, cartilage islands, bony tissue, squamous cell nests, various loose mesenchymal tissue and skin with dermal appendages. There was no vertebral column. The chromosome study from the cord blood of the fetus and tumor cells were done. The chromosome of lymphocyte of cord blood of the fetus showed 46, XX. However, the tumor cells showed variable chromosomal abnormality such as trisomy, terminal deletion, interstitial deletion and chromosome breakage.

Case 4

This 22-week-old female fetus was terminated on October 28, 1989 due to maternal hydramnios and a huge mass detected by sonogra-



Fig. 2. Low-power view of Case 1 shows predominantly immature neuroepithelial tissue admixed with cartilage islands, respiratory tract, gastrointestinal tract and supporting mesenchymal tissue(H & E, $\times 2.5$).



Fig. 3. In a high-power view of Case 1, immature neuroepithelial tissue appears to be primitive neural tubules(H & E, $\times 100$)



Fig. 4. Gross picture of case 2 shows a huge protruding mass from the oral cavity and bulging cheek.



Fig. 5. Sagittal section of Case 2 shows the tumor filling the oral cavity and almost the entire intracranium, which displaces the brain to both lateral sides. The tumor is attached to the midline of the hard palate and sphenoid bone.

phy. On autopsy, partial disconnection of the head due to dystocia and an irregular-shaped friable mass protruding from the oral cavity and the left zygoma, measuring $16 \times 14 \times 11$ cm, were noted (Fig. 11). On sagittal section of the head, the tumor was attached to the midline of the soft palate (Fig. 12). The tumor extended to and destroyed the left zygomatic region of the face resulting in a protrusion from it. Grossly, it showed friable solid and cystic appearance, and five pairs of foot-like structures with cartilaginous tissue were seen. No other anomaly was noted. Microscopically, the epignathus was immature teratoma, the most part of which was primitive neuroepithelial tissue and glial tissue. Elsewhere, the tumor was intermingled with the respiratory tract wall and bronchial cartilage, hepatic parenchyme, skeletal muscle, gastrointestinal tract (Fig. 13), choroid plexus, retinal pigment epithelium, bone and cartilage. The placenta showed subchorionic and retroplacental

hemorrhage.

DISCUSSION

The most frequent site of congenital teratoma is the sacrococcygeal region (Hawkins, 1972; Dehner, 1986). Other common sites are the neck, nasopharynx, mediastinum and retroperitoneal region. The most common form of teratoid neoplasm in the nasopharyngeal region is the dermoid or hairy polyp. True teratomas of this region are extremely unusual (Felder, 1975; Chaudhry 1978). These tumors usually present as a congenital lesion in the neonatal period and only rarely occur in children over the age of two (Gnepp, 1986; m Batsakis, 1979). They are six times more common in females (Foxwell & Kelham, 1958). The patients commonly present with dysphagia and respiratory distress, and occlusion of the mouth of the host results in

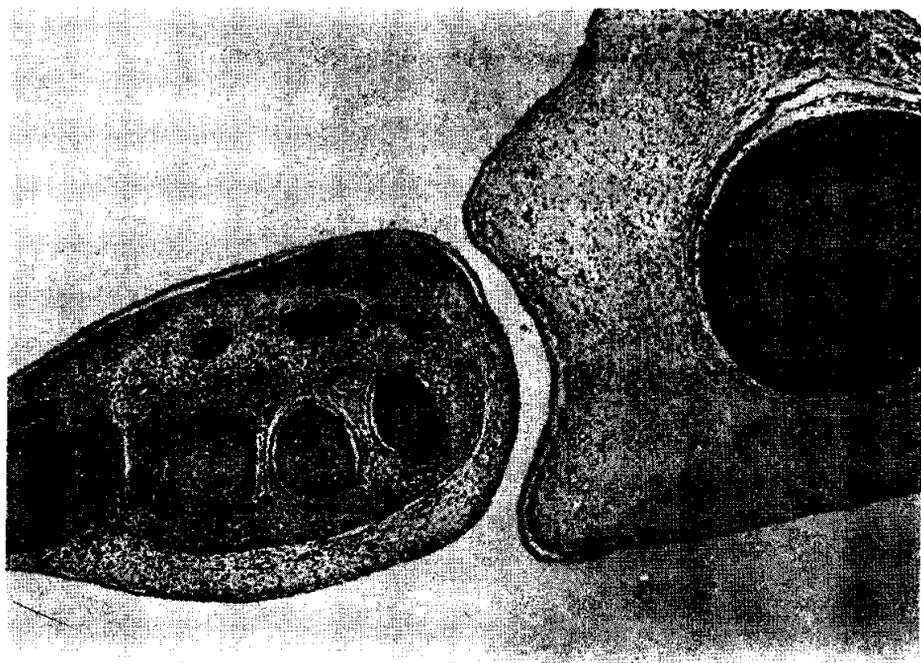


Fig. 6. Microscopic view of Case 2 shows a digit having a chondroid skeleton and covered by primitive skin(H & E, $\times 2.5$).



Fig 7. Low-power view of Case 2 shows predominantly primitive neuroepithelial tissue and glial tissue intermixed with mature tissue elements(H & E, $\times 2.5$).



Fig. 8. Gross picture of Case 3 shows a residual pharyngeal mass.



Fig. 9. Sagittal section of Case 3 shows tumor attached to the midline of the hard palate.



Fig. 10. Microscopic view of Case 3 shows well-differentiated hepatic parenchyma with gallbladder and extramedullary hemopoiesis(H & E, $\times 40$).



Fig. 11. Gross picture of Case 4 shows protruding mass from oral cavity and the left zygomatic area.

maternal polyhydramnios (Rosenfeld, 1979). Nasopharyngeal teratomas may arise from the sphenoid bone, hard or soft palate, or the jaw (Sollee & Memphis, 1965).

The history of the teratoid parasites of the pharynx and mouth has been well described by Ahlfeld (1875). In 1880, he found 40 cases in the literature; some were attached to the autosite by an umbilical cord. Since the more highly developed parasites showed brain tissue in the upper pole and extremities in the lower, Ahlfeld thought that they were true fetal implantations and the aberrant growth of a totipotent blastomere. Schwalbe (1907) described nasopharyngeal teratoma as an epignathus which he considered to be a various type of asymmetrical double monster. He thought that they were due to dislocation of germ material and classified them into four subgroups according to the degree of organization of the parasite. (1) Class I: parasitic monster attached to autosite through umbilical

cord; (2) Class II: ill-defined tumor mass that may be attached to head, neck or thorax; (3) Class III: organized trigermal oral teratoma; (4) Class IV: mixed tumors generally having the shape of hairy pharyngeal polyps. According to this classification our cases would be categorized into Class III.

Meanwhile, Ewing (1942) classified them as follows: (1) Dermoids: containing tissue of epidermoid and mesodermal origin; (2) Teratoids: containing tissue from the three primary germ layers but poorly differentiated; (3) True teratomas: similar to teratoid tumors but differentiated into tissue resembling organs histologically; and (4) Epignathi: trigermal in origin and differentiated grossly into organs. However, these ideas of the possibility of a parasitic twin had been abandoned. We never found such reported nasopharyngeal epignathus having a vertebral axis. In our cases, no vertebral axes were identified either. From the late 1950's to 1960, there was some dispute as to whether it was a congenital malformation or a true neoplasm (Foxwell & Kelham; 1958). Today a teratoma is regarded as a true neoplasm because it displays progressive and uncoordinated growth.

Among the four cases presented (Table 1), two were male and two were female. Three cases were artificially terminated due to fetal mass and polyhydramnios detected by ultrasonography during antenatal care. One case died five minutes after birth due to severe respiratory difficulty. All four cases showed a typical protruding epignathus from the oral cavity. In all cases it was also found that the mass was attached to the midline of the hard palate. One case extended to the intracranium and filled half of the cranial cavity, and the other one case destroyed the left zygoma. All four cases were associated with maternal polyhydramnios. Microscopically, all were true teratomas and the most portion was composed of primitive neuroepithelial components. According to the grading system of Norris et al., applied to the ovarian teratoma, our cases were grade III immature teratoma. Although the nasopharyngeal teratoma is composed of an immature somatic element, malignant transformation has never been reported (Dehner; 1986).

The histogenesis of the nasopharyngeal teratoma is controversial. These lesions commonly ori-

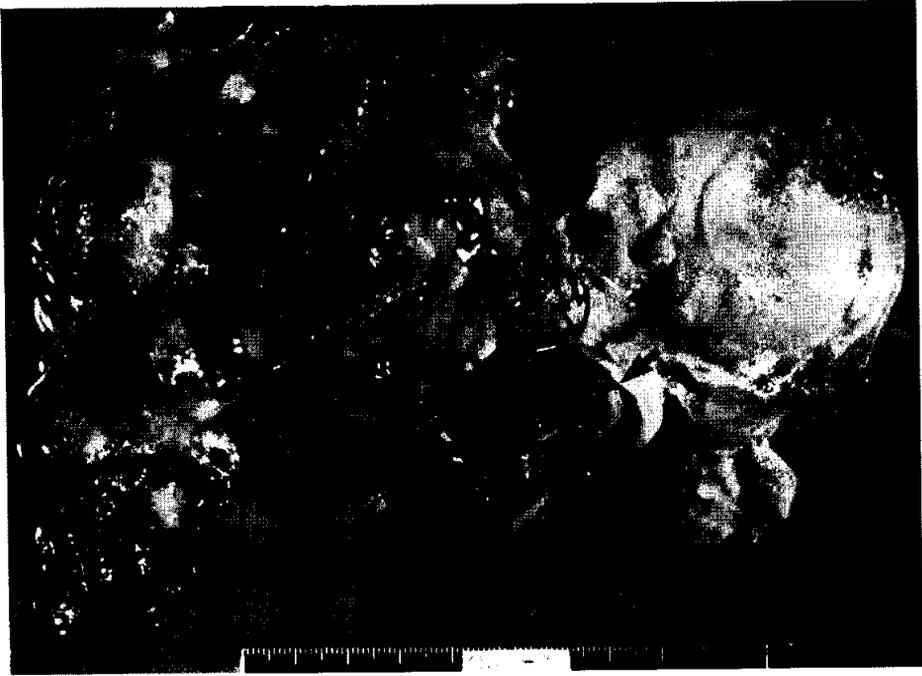


Fig. 12. Sagittal section of Case 5 shows the tumor attached to the midline of the soft palate(curved arrow). The oral cavity and mandible are seen(arrow).

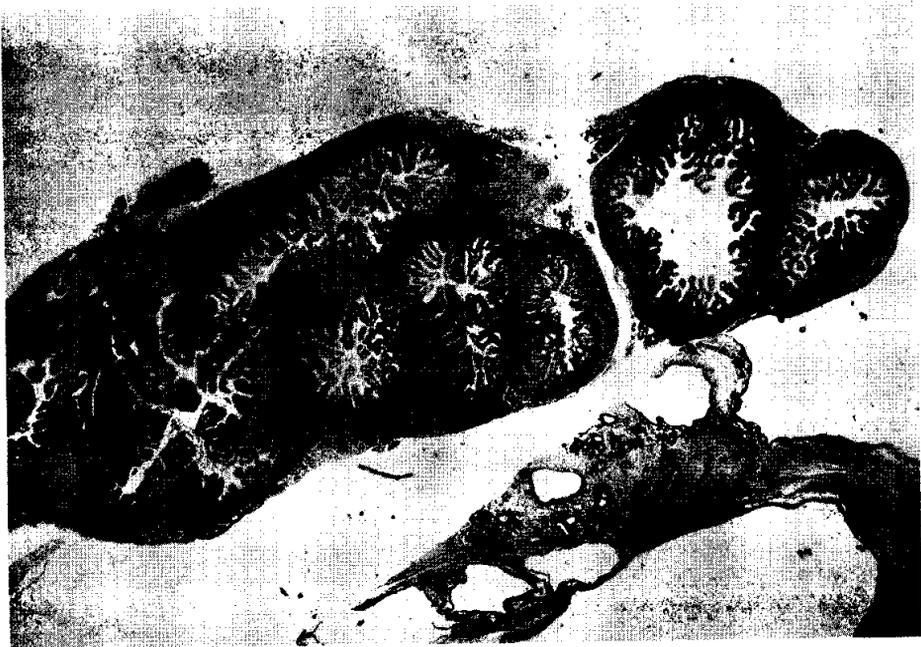


Fig. 13. Microscopic view of Case 4 shows well-differentiated gastrointestinal tract(H & E, $\times 2.5$).

Table 1. Clinical and pathological findings of pharyngeal teratoma

Case	Gestational Age	Sex	Mass size (cm)	Attached site	Associated anomalies	Maternal polyhydramnios	Karyotype of fetus
Case 1	32 wks (Survived for 5 minutes)	M	12 × 10 × 7	Midline of the hard palate	Agensis of right kidney Accessory spleen Undescended testis	Present	Not done
Case 2	21 wks	M	12 × 11 × 10	Midline of the hard palate & sphenoid bone	Absent	Present	46,XY,-9, +inv(q)(p11q21)
Case 3	24 wks (Stillborn)	F	13 × 12 × 5	Midline of the hard palate	Absent	Present	46,XX
Case 4	22 wks (Stillborn)	F	16 × 14 × 11	Midline of the soft palate	Absent	Present	Not done

ginate at the central site as in our cases where, in early embryonic life, the buccopharyngeal membrane, Rathke's pouch and the rostral end of the notochord meet. Chaudhry(1978) suggested that these might be the result of misplaced totipotential cells of one or more germinal layers, which escape the physiological-organizing influences that control growth and development under normal circumstances. Such cells retain the ability to proliferate, differentiate and form tumor-like conglomerates of various tissue that are present in a disorganized fashion.

Clinically, the differential diagnosis of nasopharyngeal teratomas includes encephalomeningoceles, gliomas, hemangioma, neurofibromas and heterotopic brain tissue(Heroman et al., 1980). Since heterotopic brain tissue is occasionally found in the newborn nasopharynx(Low et al., 1956; Zarem et al., 1967, Broniatowski et al., 1981), the differentiation cannot always be made by a biopsy alone since pharyngeal teratomas contain an abundance of central nervous system tissue. It should also be noted that occasionally massive teratoma of adjacent anatomic regions can involve the nasopharyngeal region (Ehrlich, 1945).

In this series 2 cases underwent chromosomal study. One case showed pericentric inversion of chromosome 9 in the fetus and the mother, while the father was normal. The other case showed normal karyotype.

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= 국문초록 =

비인강 기형종(상악체)

-4부검 증례-

서울대학교 의과대학 병리학교실 및 제일병원 병리과

박성혜 · 강경훈 · 고경혁 · 김희숙 · 지제근

비인강 기형종은 일명 상악체라고 알려진 대단히 드문 질환으로 신생아기에 발견되는 중요한 기형종이다. 그 모양이 크고 괴상할 뿐 아니라 구강내에서 밖으로 돌출하는 특이한 양상을 띄고 있고 또 육안적으로나 조직학적으로 분화가 잘 되어 있어 예전부터 그 발생기전에 대하여 많은 논란이 있었던 병변이다.

우리나라 문헌상 거의 보고례를 찾을 수 없었는데 서울대병원 부검기록을 검토한 결과 4례가 있었고 또 이들중 1예는 두개강내로 자라 들어가는 특이한 모양을 나타내어 이들을 부검소견과 함께 보고하였다.

병리학적 검사상 실구 상악체는 비인강과 구강을 포함한 두부를 제외하고는 기타 장기의 기형을 별로 동반하지 않고 있음을 알 수 있었고, 예외 없이 구개에 붙어 있기 때문에 중앙부 구개부가 그 기원이라 여겨졌다. 현미경 소견을 종합컨데 미숙 중추신경조직이 주종을 이루고 있었으나 기타 기형종에서는 흔히 관찰되지 않는 실질 장기조직 특히 간 조직이 흔히 관찰됨을 알 수 있었다.