

SPOT DIAGNOSIS (IMAGE GALLERY)



Neonatal Palatal Mass

Kumaravel KS, Pugalendhiraja KV, Rameshbabu B, Rameshkumar V, Karthick NR

Department of Pediatrics, Government Dharmapuri Medical College, Dharmapuri, Tamilnadu, India.

Address for Correspondence: Dr Kumaravel KS, Department of Pediatrics, Govt Dharmapuri Medical College, Dharmapuri, Tamilnadu 636701, India. Email: kumaravelks@rediffmail.com

A term female baby born to non-consanguineous parents, delivered by normal vaginal delivery was detected to have a huge mass protruding from the mouth. The mass was pedunculated, arising from the hard palate and was with associated cleft palate. There was a maternal history of polyhydramnios. On palpation, the mass had a variegated consistency. It was predominantly hard with interspersed cystic to soft areas with an epithelial covering on the entire mass. The mass was highly vascularized but had no abnormal pulsations. Swelling was not compressible. Arterial pulsations were felt in the pedunculated stalk. Examination of other systems was normal and there were no dysmorphic features. Total surgical excision of the tumor was done. The weight of the mass was 400g. The histopathological examination revealed presence of all three germ cell tissues, cartilage and neural tissue.

What is the diagnosis?

Epignathus. It is a tumor arising from the jaw, though it is commonly used to describe tumors or teratomas of the mouth in the newborn. (1) In the year 1940, Ewing classified these nasopharyngeal tumors into 3 types: a) dermoids - consisting of epidermal and mesodermal germ cell layers, attached to soft or hard palate and, or pharynx near midline, b) teratomas - consisting of all three germ layers, with an indifferent degree of organization and c) epignathus, consisting of teratomas with high degree of organization and recognizable structures. (2) The overall incidence of teratomas is about 1 in 4000 live births. Among them oropharyngeal lesions account for less than 2 percent. These epignathus tumors arise from the Basisphenoid at the Rathke's pouch and descend into the oral cavity causing palatal defects. (3) There is a female predisposition. The cleft palate of mechanical origin is the most common associated anomaly. They are a rarity as so far since 1918 there are only totally 117 cases reported worldwide. (1) Maternal polyhydramnios is a classic feature in epignathus, which occurs due to reduced swallowing due to mechanical obstruction. There can be elevated levels of maternal alpha fetoprotein (AFP). Moreover AFP levels are used as indicators for detecting the post-operative tumor clearance. Differential diagnosis includes encephalomeningocele, hygroma, and oral granular cell myoblastoma. (4) Early sonographic diagnosis is possible with very rare 1st and 2nd trimester reporting of these cases. Confirmation of the diagnosis is done by histopathology which shows - ganglion cells, cartilage, bone, respiratory epithelium, with fibrous stroma. Prognosis is poor in these babies. Surgical excision is the treatment of choice. Prior to surgery, plain X-ray and computed tomography (CT) scan must be performed to rule out intracranial extension of the tumor. (3)

References

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