BÖLÜM 27

BURNUN KONJENİTAL MALFORMASYONLARI

Hasan DEMİREL [1]

GENEL BAKIŞ

Burnun ve yüzün konjenital malformasyonları nadir görülen anomalilerdir. Belirgin deformiteler şeklinde görülebileceği gibi, farkedilmesi güç anomaliler olarakta karşımıza çıkabilir.

NAZAL EMBRİYOLOJİ

Nazal plakodlar üçüncü ve dördüncü gestasyon haftasında ortaya çıkarlar. Gestasyonun beşinci haftasında bu plakodlar invajine olup olfaktör çukuru oluşturur, ince nazobukkal membran ile oral kaviteyi nazal kaviteden ayırırlar. Bu invajinasyon, altıncı haftada posteriorda rüptüre olup koanaları oluşturur. Maksiller proses, medial ve lateral prosesler ile birleşerek nazolakrimal oluğu şekillendirir. Frontonazal prosesten nazal septum ve premaksilla oluşur (1,2).

KOANAL ATREZİ

Koanal atrezi, nazal koanaların kemik ya da kemik-membranöz kombinasyonu tarafından, başarısız rekanalizasyon nedeniyle tıkandığı konjenital bir hastalıktır. En sık görülen konjenital burun anomalisidir. Tek taraflı veya iki taraflı olabilir; hastaların %65-75'i tek taraflıdır (3,4). Tek taraflı vakalarda, sağ tarafta atrezi görülme sıklığı sola göre iki kat daha sıktır. Anomali 5000-8000 canlı doğumda bir görülür ve kadınlarda iki kat daha fazladır. Atrezik plak, vakaların %30'unda tamamen kemiksel, %70'inde kemik-membranöz yapıdadır (3,4,5). Atrezi, vakaların %50'sinden fazlasında diğer konjenital anomalilerle ilişkili olabilir. En sık koloboma, kardiyovasküler malformasyon, koanal atrezi, büyüme ve mental gelişim geriliği, genitoüriner anomaliler ve kulak anomalilerin eşlik ettiği CHARGE sendromu ile birliktelik gösterir (5). Diğer anomaliler arasında Treacher Collins, Pfeiffer, Apert, Mandibulofasiyal disostoz ve Crouzon sendromları bulunmaktadır (6).

Koanal atrezide iki ana osteolojik deformite tanımlanmıştır; medial pterygoid plakların medializasyonu ve arka vomerde kalınlaşma. Bu deformasyonlardan herhangi biri koanada daralmaya yol açarak koananın tamamen tıkanmasına neden olabilir (7,8). Koanal atrezi oluşumunda birkaç gelişim teorisinden bahsedilir: (i)

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