A newborn baby with a tumor protruding from the mouth

Case history

A full-term Caucasian girl with Apgar scores of 9/10 at 1 and 5 minutes and a birth weight of 3,500 g was born to a healthy 24-year-old mother following her second normal pregnancy. Physical examination after birth revealed a firm, broad-based, pedunculated, pinkish-red tumor that measured 25 × 10 × 10 mm and was inserted into the maxillary alveolus to the left of the midline (Fig. 1). The infant was otherwise healthy; hematological and biochemical blood analyses and ultrasound of the head and the abdomen were within normal limits. At age 5 days, the tumor was excised. Hematoxylin- and eosin-stained sections showed large cells with abundant eosinophilic granular cytoplasm, arranged in islands, strands, or syncytial masses with scant intervening fibrovascular stroma (Fig. 2). Excision was incomplete; the tumor was present in resection margins. Healing was uneventful. At a 2-year follow-up there were no signs of local recurrence of the tumor.

What is your diagnosis?

Diagnosis

Congenital gingival granular cell tumor

Discussion

Congenital gingival granular cell tumor (CGCT) of the newborn, also known as congenital granular cell lesion, congenital epulis, congenital myoblastoma (historically), or Neumann’s tumor, is a rare benign tumor (1). Around 200 cases have been described in the literature so far (2). CGCT is usually present at birth and generally does not grow much thereafter. It most frequently occurs as a single tumor, but rarely (in 10%) as multiple ones. CGCT usually arises from the alveolar ridge, the maxillary anterior alveolar ridge being the most common location (2–7). The maxillary to mandibular ratio is 3:1 (3). Additional congenital or underlying bone or dental anomalies are usually not present (5), although there are reports of a hypoplastic or absent underlying tooth (4). There have been a few reports on CGCT tongue involvement (3, 8). The lesion has a striking female preponderance (the female to male ratio is 8–10:1) (3, 9, 10). Spontaneous regression has occasionally been reported (11).

Etiologic factors for CGCT are uncertain. Trauma such as finger sucking in utero is likely not significant. The female preponderance and the cessation of growth or even spontaneous regression of the lesion after birth, when maternal hormones are absent, both suggest maternal hormonal influence (2–5).

The histogenesis is debated, and different studies suggest several origins of CGCT: odontogenic epithelial, pericytic, and fibroblastic (3, 12). Proposed cells of origin also include histiocytes and nerve-related (4, 13), smooth muscle, and primitive mesenchymal cells (2, 14).

Clinically, lesions of the oral mucosa are divided into surface lesions and soft tissue enlargements, which are either reactive enlargements or tumors. Tumors can be solid or cystic, benign or malignant, and congenital or non-congenital. CGCT belongs in the mesenchymal benign tumor group. Reports of spontaneous regression, absence of local recurrence or metastasis after incomplete excision, and lack of a malignant counterpart all imply that CGCT is of non-malignant character (2, 3). CGCT may obstruct the digestive or respiratory tracts. In utero, obstruction of the oral cavity can result in in-
C a s e   p r e s e n t a t i o n

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effective swallowing and can cause polyhydramnios. Postnatally, it may interfere with feeding (4), as in our case, or respiration.

Macroscopically, the CGCT appears as a firm, protuberant pink mass, sometimes pedunculated, of varying size (a few mm to 9 cm) (2, 3). It is covered with normal mucosa and can be macerated on the tip.

Light microscopy demonstrates large cells with abundant eosinophilic granular cytoplasm, arranged in islands, strands, or syncytial masses with scant intervening fibrovascular stroma. Unlike granular cell tumors in adults, the overlying surface epidermis in CGCT does not show pseudoepitheliomatous hyperplasia (2, 4, 5). Immunohistochemically, granular cells in CGCT are typically S-100 protein negative, but are HLA-DR antigen, vimentin, and occasionally NSE and CEA positive (2, 13). Furthermore, they lack immunostaining for alpha fetoprotein, actin, and specific macrophage markers (2).

Prenatal imaging of congenital lesions of oral mucosa is possible by ultrasound and magnetic resonance imaging and can be helpful in planning the delivery (1). Postnatally computed tomography or magnetic resonance imaging of the head is useful in demonstrating the extent and differential diagnosis of congenital maxillofacial mass lesions and for planning surgical treatment.

The clinical differential diagnosis of congenital lesions of oral mucosa is broad and depends on site of involvement, size, velocity of growth, and possible accompanying lesions. The most important lesions to differentiate from CGCT are oral teratoma-epignathus (1, 10) and melanotic neuroectodermal tumor of infancy (11, 17). Other possible diagnoses also have to be taken into account: fibroma (10, 11, 18), lipoma (19), leiomyoma (20), rhabdomyoma (21), rhabdomyosarcoma (22), peripheral giant cell granuloma (11), pyogenic granuloma (10), cysts of oral mucosa, Fordyce’s spots, natal teeth, eruption cysts (11), neurofibroma (3), myxoma (23), hemangiomia, lymphangioma, alveolar lymphangiomia, and congenital ranula (11, 24).

There are reports of spontaneous regression of CGCT, and therefore conservative treatment is sometimes sufficient (11, 25). When the lesion is obstructing feeding or respiration, surgical removal is indicated. There are no reports of local recurrence after incomplete excision (2, 3). In our case, there was no local recurrence of the tumor 2 years after local excision despite positive surgical margins after the procedure. Therefore, a conservative and non-mutilating approach is preferred. There are also reports of conservative surgical removal with CO₂ laser (3).

Learning points

• Congenital gingival granular cell tumor is a rare benign tumor that most often presents on the maxillary or mandibular alveolar ridges.
• About 200 cases have been reported to date.
• The tumor has a striking female preponderance.
• In contrast to granular cell tumors in adults, CGCTs lack pseudoepitheliomatous hyperplasia and are consistently S-100 protein negative.
• No recurrences have been described even after incomplete excision.

REFERENCES


