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# The congenital basal cell adenoma of salivary glands

## Contribution to the differential diagnosis of congenital salivary gland tumours

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**Abstract** Congenital epithelial tumours of the salivary glands are very rare. The Salivary Gland Registry maintained in the Department of Pathology, University of Hamburg, contains only three cases among a total of 6,646 salivary gland tumours from the years 1965–1994. The three cases were classified as congenital basal cell adenoma, two of the parotid gland and one of the submandibular gland. Histologically, the three adenomas were similar in structure to the adult counterpart of basal cell adenoma with solid, trabecular or tubular (duct-like) patterns. In some cystic spaces of the duct-like structures PAS- and Astra blue-positive substances were secreted. On immunocytochemistry, the luminal duct-like cells showed membranous expression of cytokeratins 3, 5, 6, 7, 13 and 19. In the isomorphic basaloid cells of the solid and trabecular cell nests few cells expressed cytokeratin. On the outside of the solid cell nests there were smaller elongated myoepithelial-like cells, which expressed cytokeratin 14 and vimentin. Cytokeratins 1, 2, 4 and 18 were not expressed. The pattern of expression reflects the different stages of maturity of the tumour cells and is related to the development of the salivary glands until the end of the 3rd embryonal month with an arrest of further cell differentiation. No acinic cells, invasive growth, recurrence or metastases were observed. The differential diagnosis includes other congenital salivary gland tumours, such as hybrid basal cell adenoma–adenoid cystic carcinoma, sialoblastoma or embryoma, carcinoma, hamartoma and teratoma.

**Key words** Congenital epithelial tumours of salivary glands · Basal cell adenoma · Differential diagnosis

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## Introduction

About 3–5% of salivary gland tumours occur in children [3, 23, 24, 26, 34]. In the first years of life vascular tumours dominate (haemangiomas are the most frequent tumours during the newborn period and the 1st year), but in the second decade solid epithelial tumours are the most significant group.

Congenital tumours can develop in the embryonal or fetal period; congenital epithelial tumours are very rare [1, 4–6, 8–10, 18, 20, 32, 36, 37, 39]. The following three types of epithelial tumours can be differentiated:

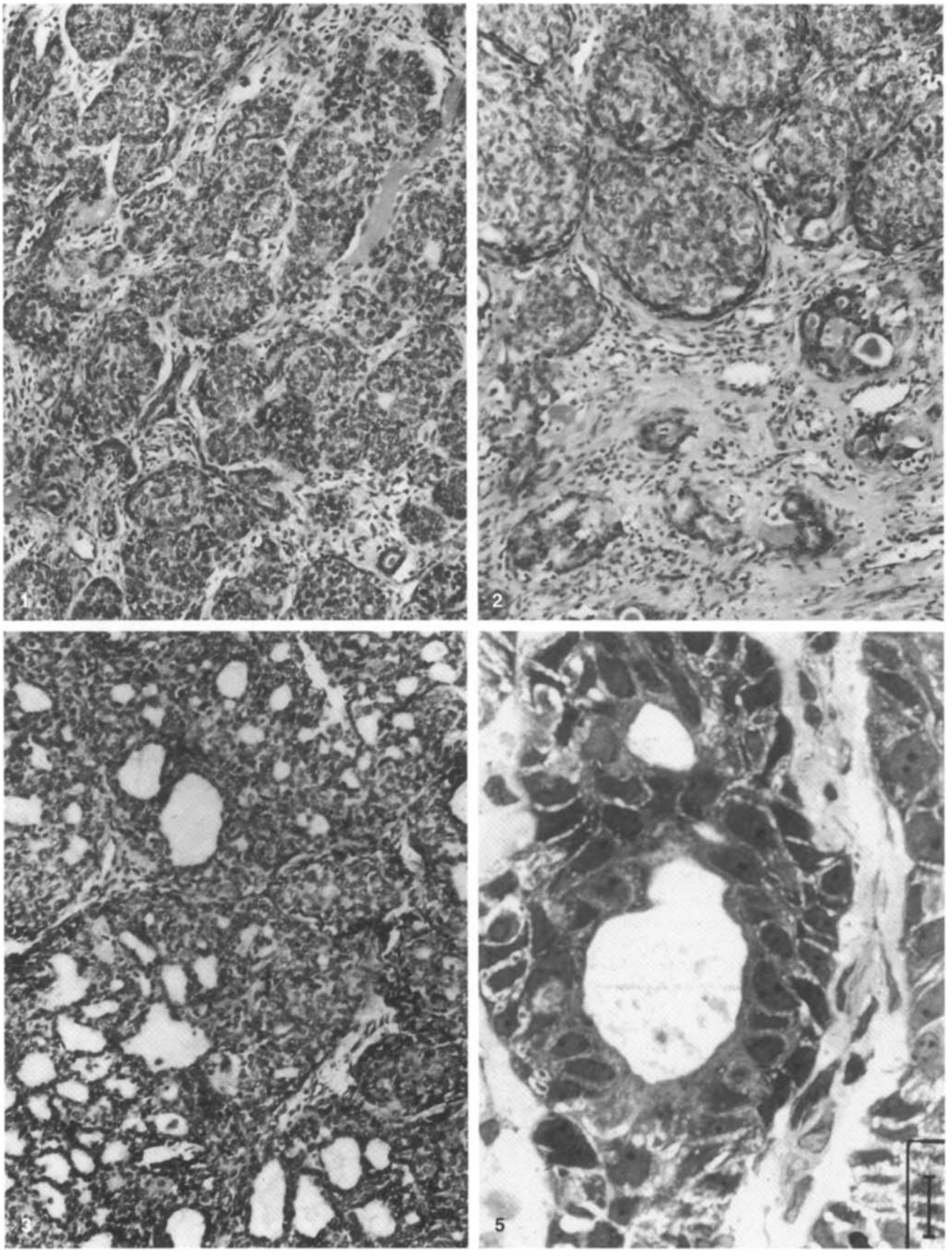
Adenomas, comparable to adult forms
Sialoblastomas or embryomas
Carcinomas

In this paper three cases of congenital basal cell adenoma of the parotid gland will be analysed. In addition, the differential diagnosis against other congenital tumours of the salivary glands will be discussed.

## Materials and methods

Between 1965 and 1994 in all 18,111 tissue specimens of salivary glands were collected in the Salivary Gland Registry, University of Hamburg, 6,646 of which were salivary gland tumours (36.6%). Of all the epithelial tumours, 67% were adenomas and 33% carcinomas. Of all epithelial tumours, 2.5% occurred in children and adolescents younger than 20 years. The adenomas included only three cases of congenital adenoma of the basal cell type.

The biopsy specimens were routinely fixed in formaldehyde and embedded in paraffin wax. Several stains were used, including haematoxylin and eosin, PAS reaction, Astra blue, or Masson-Goldner. In addition, the cases were studied immunohistochemically, by staining for total cytokeratin (CKMNF), cytokeratins 1–7, 13, 14, 18, 19, GFAP (glial fibrillary acid protein), S-100 protein, EMA, CEA, vimentin, muscle-specific actin and desmin. A standardized procedure was followed using the immunoperoxidase method (PAP method) and the alkaline phosphatase method (APAAP method) with application of the following primary antibodies: Dako Diagnostika Hamburg (CKMNF, CK 3, CK 6, CK 8, CK 19, CK low, CK high, GFAP, EMA, CEA, S-100, desmin); Sigma, Deisenhagen (CK 4, CK 7); Boehringer, Mannheim (CK 5,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CK 6); Progen Biotechnik, Heidelberg (CK 13, CK 18, vimentin); Amersham, Braunschweig (CK 1, CK 2); Biogenex, San Ramon (CK 14); Enzo Diagnostics, Farmingdale (actin).

For electron microscopy small tissue blocks were fixed in 3% glutaraldehyde-cacodylate buffer (pH 7.2–7.4; 300 mol/sm) at 4°C for 2 h. After washing, 1.33% s-colloidin-buffered osmium tetroxide was used as another fixative. Then the material was embedded in Epon 812. Thin sections were stained with uranyl acetate and lead citrate, then examined and photographed with a Philips EM 300 electron microscope (beam voltage 80 kV).

## Results

### Case 1

Immediately after birth a tumour was observed in the area of the left parotid gland of a male newborn. The tumour was totally enucleated and was found to have a cartilage-like consistency with a diameter of 3×3 cm. In some areas it was adherent to the oral mucosa and the skin. The branches of the facial nerve were located above the tumour. The lymph nodes were not involved. In the further course no recurrence or metastases were observed.The tumour was encapsulated and had a lobular structure (Fig. 1). The predominant formations were solid and trabecular patterns with inclusion of few duct-like structures. The mostly isomorphic basaloid cell nests were limited on the outside by darker smaller cells (Fig. 2). The nuclei of the tumour cells were large and ovoid with no atypical mitotic activity. No infiltrating growth was observed.

### Case 2

At birth, a female newborn showed a tumour in the area of the right submandibular gland. At the age of 1 month a tumour with diameters of 2×2 cm was removed. In the further course no recurrence or metastases were observed.

The encapsulated and lobulated tumour contained not only solid cell formations, but also many tubular and cystic patterns (Fig. 3). In the cystic spaces of the duct-like microcysts, PAS- and Astra blue-positive substances were secreted. On the outside of the solid cell nests there were flat, darker epithelial cells. There was no infiltrating growth.

◀ **Fig. 1** Congenital basal cell adenoma of the parotid gland (case 1): lobulated arrangement of mostly solid epithelial basaloid cell complexes. H&E, ×100

**Fig. 2** Case 1: smaller and darker cells on the outside of solid cell complexes; inclusion of some duct-like structures; loose stroma. H&E, ×100

**Fig. 3** Congenital basal cell adenoma of the submandibular gland (case 2): solid basaloid cell complexes with inclusion of duct-like cystic spaces. H&E, ×100

**Fig. 5** Case 3: duct-like structure with an inner layer of ductal epithelial cells and an outer layer of more vacuolated cells. Semithin section, toluidin blue, ×400

### Case 3

At birth, a male newborn had a tumour node on the right cheek. A 5-cm (diameter), encapsulated tumour of the right parotid gland was removed. On the surface some haemorrhages were observed.

The tumour was characterized by solid cell nests and multiple tubular formations with inclusion of PAS- and Astra blue-positive secretory substances in the ductal spaces. Some stroma areas show regressive changes with focal hyalinization and infiltration by leucocytes and histiocytes. The outer layered cells were surrounded by small basal membrane substances. In the more solid cell nests the type of the outer layered dominates.

On aspiration cytology, isomorphic basaloid cell nests were observed with large ovoid nuclei but without duct-like structures (Fig. 4).

In semi-thin sections the duct-like structures were limited by an inner layer of duct epithelium and an outer layer of cells characterized by distinct intercellular spaces and a focally vacuolated cytoplasm (Fig. 5).

The duct-like structures differed in diameter and lining cells. Structures with a large diameter had columnar lumen-lining cells, whereas those with a smaller diameter were limited by cuboidal cells. Both contained flat cells in the periphery. The cord-like strands were composed of four irregular cell layers and showed single-cell necrosis and mitotic figures, which were also seen in the duct-like structures.

All three tumours showed similar structures to the adult counterpart of basal cell adenoma. The stroma was loose with inclusion of dilated blood vessels. No mucoid alterations, acinic cell differentiation, perineural invasion or infiltrating growth were observed. The basaloid cells were occasionally palisaded and surrounded by small basement membrane-like structures.

Immunocytochemically, the epithelial tumour cells, and in particular the luminal duct-like cells, expressed cytokeratin (Fig. 6). The cytokeratin subclasses varied in location and intensity of expression. The expression of cytokeratin 3, 5, 6, 7, 13 and 19 was especially distinct in all duct-like cells (Fig. 7), but only in a few cells of the more solid cell nests. Expression was concentrated at the luminal borders of the duct-like cells and the cell membranes. The cytokeratins of higher molecular weight (type II) showed more emphatic expression (Fig. 8a) than those of lower molecular weight (type I: Fig. 8b). Cytokeratins 1, 2, 4 and 18 were not expressed. The expression of vimentin (Fig. 9) and cytokeratin 14 was localized in the smaller elongated cells in the periphery. Muscle-specific actin was in the same location as vimentin, but was expressed with less intensity. GFAP and desmin were not seen, but CEA and EMA showed a positive reaction in the epithelial duct-like tumour cells.

Ultrastructurally, the cytoplasm of the basaloid cells (Fig. 10) contained free ribosomes, small Golgi regions, rough endoplasmic reticulum and secretory-like granules in occasional tumour cells. Blunt finger-like villi extended into tiny lumina and into segmental dilatations of intercel-

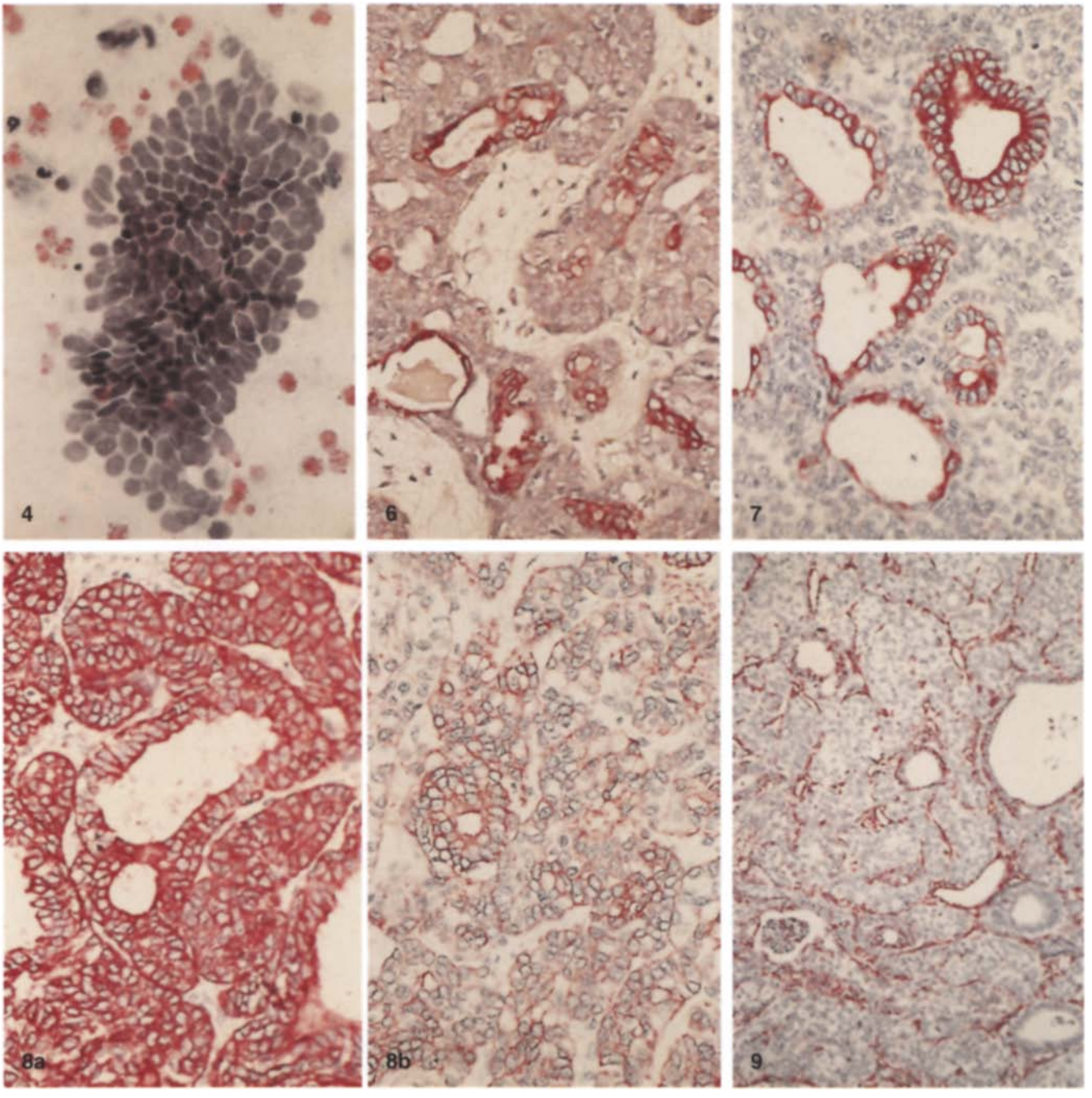


**Fig. 4** Case 3: aspiration cytology with isomorphic basaloid cells. H&E, ×400

**Fig. 6** Case 2: expression of CK3,6 in the duct-lining cells. Immunostaining APAAP, ×250

**Fig. 7** Case 3: expression of CK7 in the luminal borders of duct-like cells. Immunostaining APAAP, ×250

**Fig. 8** Case 3: **a** Marked expression of CK (higher molecular weight) in the duct-like cells and also the solid cell nests. **b** Moderate expression of CK (lower molecular weight) in the cell membranes. Immunostaining APAAP, ×250

**Fig. 9** Case 3: expression of vimentin in the outer layered cells. Immunostaining APAAP, ×100

lular spaces. Tumour cell differentiation was comparable with that in cells of the terminal ducts in parotid gland development in the 20th to 25th weeks of pregnancy.

## Discussion

In the very small group of congenital tumours of the salivary glands three types are seen in most series: congenital basal cell adenoma (Table 1), hybrid basal cell adenoma–adenoid cystic carcinoma (Table 2) and sialoblastoma (embryoma; Table 3). The cases documented thus far comprise 8 cases of basal cell adenoma, 3 cases of hybrid basal cell adenoma–adenoid cystic carcinoma, and

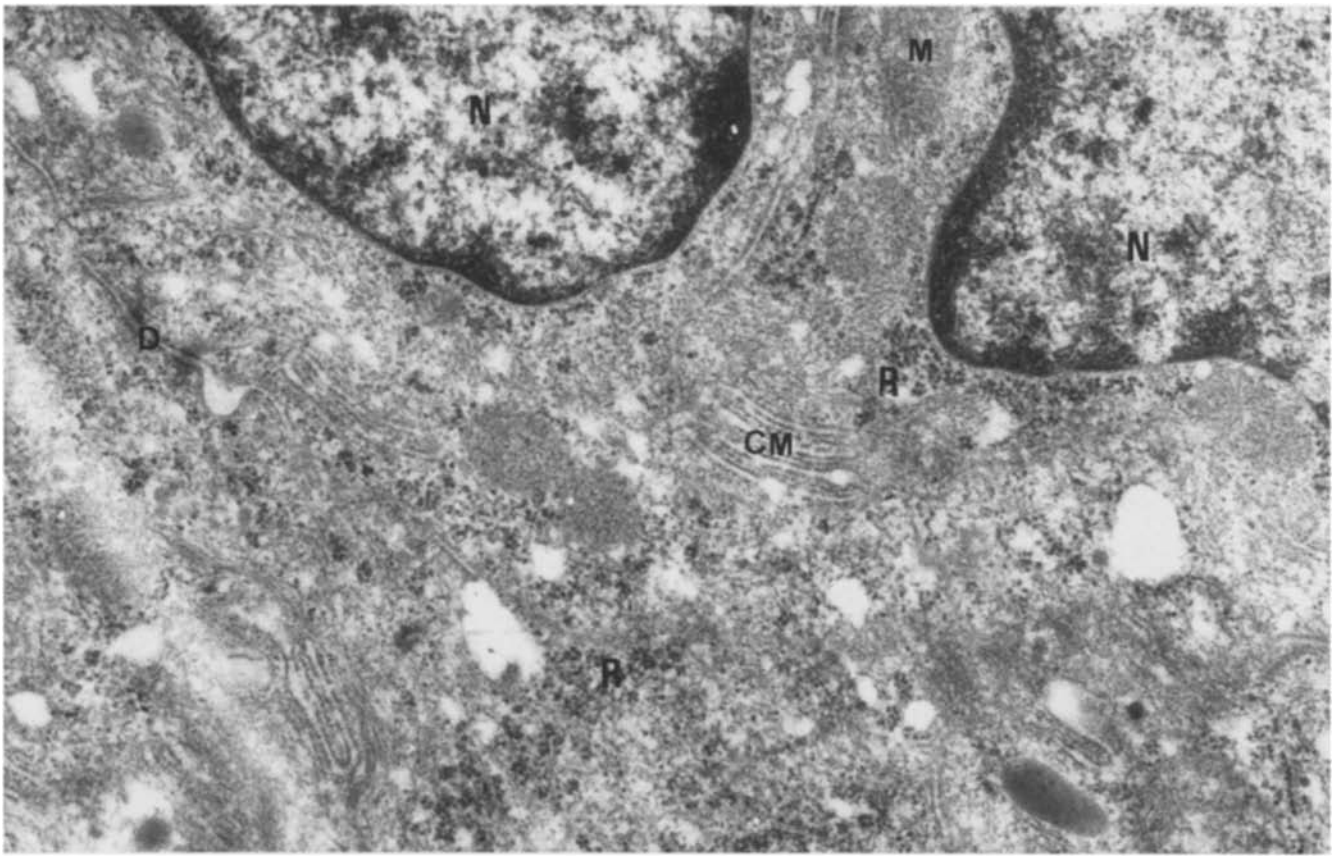


**Fig. 10** Case 3: Ultrastructure of basaloid cells with free ribosomes (*R*), mitochondria (*M*), folded cell membranes (*CM*) and desmosomes (D); nucleus (*N*). ×31,800

**Table 1** Congenital basal cell adenoma of major salivary glands (*Pa* parotid gland, *Sm*, submandibular gland, *nk* not known)

| Reference | Location | Sex | Remarks[a] |
|---|---|---|---|
| [23] | Pa | m | "Basal cell adenoma"; size 5 cm; trabecular and tubular structures (Fig. 10–11); no recurrence 4 years after surgery; no further details |
| [40] | Pa | nk | "Monomorphic adenoma"; tendency to branching in duct-like structures; solid cell nests of basal cell type (Fig. 145); no infiltrating growth; no recurrence; no further details |
| [9] | Sm | m | "Basal cell adenoma"; size 3.5×1×1 cm; excision of a lobulated tumour; solid and tubular-ductal formations with inclusion of mucinous substances; inner cells with whorling tendency; outer cells with palisade arrangement; no recurrence; disease-free 18 months after treatment |
| [32] | Pa | m | Size 8 cm; polylobular; well limited; no recurrence; "congenital basal cell tumour", "embryoma" or "embryonal tumour" |
| [10] | Pa | f | "Monomorphic adenoma"; size 1 cm; firm mobile mass; solid cellular pattern with some myoepithelial cells at the periphery of lobules; focal ductal differentiation with cytokeratin expression; at 6 months of age recurrence in the biopsy scar (size 2×1.5 cm); total parotidectomy; no facial paralysis or recurrence |
| [33] | Pa | m | Case 1: see Results |
| | Sm | f | Case 2: see Results |
| | P | m | Case 3: see Results |

[a] Original diagnoses of authors in quotation marks

11 cases of sialoblastoma (embryoma). Eighteen other congenital tumours with differing pathology are listed in table 4. We observed three cases of congenital salivary gland tumours, which were classified as congenital basal cell adenoma.

Congenital basal cell adenoma (Table 1) shows similar structures to the adult counterpart of basal cell adenoma. The lesions are encapsulated, lobulated and mostly localized in the parotid gland. The basaloid epithelial cells are arranged in solid, trabecular or tubular forma-

**Table 2** Hybrid congenital basal cell adenoma–adenoid cystic carcinoma of major salivary glands

| Reference | Location | Sex | Remarks |
|---|---|---|---|
| [43] | Sm | nk | Size 4 cm; ducts and solid lobules; focal adenoid cystic pattern (glandular type); no recurrence after 3 years; "basal cell adenoma–adenoid cystic carcinoma"; no further details |
| [36] | Pa | f | Size 1.5×1.5 cm; facial nerve invasion with facial paralysis; total parotidectomy; at 8 months of age regional lymph node metastasis; neck dissection; chemotherapy (Adriamycin); free of tumour 4.5 years after presentation; basaloid cell nests with peripheral palisading and duct-like structures; focal areas resembling adenoid cystic pattern; "hybrid basal cell adenoma–adenoid cystic carcinoma" |
| [1] | Pa | m | After birth slowly growing tumour; at 12 months superficial parotidectomy; unencapsulated tumor (size 2.5×2×1 cm); solid cell nests with single neoplastic lumina and mitotic figures; peripheral palisading cells of basal cell type; invasion of the adjacent connective tissue, but no neural or vascular invasion; 8 years after presentation no recurrence or metastases; diagnosis as "low-grade basaloid adenocarcinoma" |

**Table 3** Congenital sialoblastoma/embryoma of the major salivary glands

| Reference | Location | Sex | Remarks |
|---|---|---|---|
| [42] | Pa | f | Size 5×7 cm; movable, encapsulated and lobulated; haemorrhages and degeneration centrally; excision; courses of irradiation and chemotherapy; at 3 years of age free of tumour; trabecular and ductal pattern with PAS-positive deposits in the lumina; peripheral palisading cells with basal membrane substances; structure similar to that of basal cell adenoma; "embryoma" |
| | Pa | m | Size 5×3×3 cm; recurrence at 6 months of age (size 3 cm) and at 10 months (size 3 cm) beneath the scar; one course of irradiation; 12 years after presentation free of tumour; identical pathohistology, similar basal cell adenoma; "embryoma" |
| [8] | Pa | f | "Embryoma", extensive excision; one course of radiotherapy; at 4 years free of tumour; no further details |
| [6] | Pa | m | Size 4.5×4.0×3.5 cm; semicystic; parotidectomy; no recurrence; no facial paralysis; solid areas with peripheral palisading cells; duct-like structures; no acinar cell; cytoarchitecture analogous embryonic salivary gland anlage with arrest of maturation; similar basal cell adenoma; "embryoma" |
| [39] | Pa | f | Size 12×15 cm (weight 120 g); one lymph node metastasis; recurrence at 6 months (size 2×1 cm), at 13 months and over 30 months; no haematogeneous metastases; solid cell nests and ductal structures with expression of cytokeratin; myoepithelial cells surrounding the ductal structures with expression of actin; no acinar differentiation; "sialoblastoma", but similar to basal cell adenoma |
| [18] | Sm | f | Size 2 cm at birth; slowly increasing growth at the age of 10 months; excision of multilobulated tumour (size 4 cm); no recurrence; composition of basal type cells with ductal and acinar differentiation; intracellular secretory granules; expression of cytokeratin and EMA in the ductal cells; expression of S-100 protein and vimentin in the peripherally located myoepithelial cells; "sialoblastoma" |
| [20] | Pa | f | Size 2.2 cm; at 17 months recurrence; parotidectomy; no further recurrence; no metastases; solid nests of epithelial cells and ductal structures; no acinar cells; expression of cytokeratin in the ductal cells; expression of actin, vimentin and S-100 protein in the myoepithelial cells of the outer layer of ducts; "sialoblastoma" similar to basal cell adenoma |
| [5] | Pa | nk | Three parotid gland "embryomas" without further details; solid and apparently embryonic cell groups evocative of the embryonal epithelial anlage of salivary glands at various stages of its branching morphogenesis; solid nests and duct-like structures with early ductal spaces |
| [37] | Pa | f | "Sialoblastoma"; size 12 cm; association with hepatoblastoma (AFP positive); no further details |

tions with inclusion of some microcysts. The included duct-like structures contained secretory material, but no acinar cell differentiation, foci of squamous cells or atypical mitoses were observed. In our cases no infiltrative growth, lymph node metastases or recurrence were present.

Some cases documented in the literature show structures reminiscent of congenital basal cell adenoma but have been classified as "embryoma" [40, 42] or "monomorphic adenoma" [10, 40]. Only in 1 case [42] was local recurrence at 6 and 10 months of age observed beneath the upper portion of the scar, but no lymph node metastases.

The morphology of congenital basal cell adenoma is similar to that seen at particular stages in the developing parotid gland [14]. The recapitulation of embryogenesis is an attractive model, which reflects the architecture of congenital basal cell adenoma. The lumina of the duct-like structures are limited by undifferentiated embryonal cells, and myoepithelial cells are arranged between these cells. Immunocytochemical results suggest that undiffer-

**Table 4** Other congenital tumours of the salivary glands (*Np* nasopharynx)

| Reference | Location | Sex | Type of tumour | Remarks |
|---|---|---|---|---|
| [32] | Pa | m | Congenital sebaceous cell adenoma | Size 5 cm (50 g); well encapsulated; at 9 years of age no recurrence |
| [23] | Pa | m | Congenital pleomorphic adenoma | Autopsy case |
| [5] | Sm | – | Hamartoma-like tumour | Tubuloductal phenotype with acinar differentiation; lobular arrangement; resemblance to monomorphic adenoma; no further details |
| [31] | Sm | f | Congenital teratoma | Size 2 cm with few cysts (0.2–0.5 cm); infiltration of skin; inclusion of heterotopic tissue (neuroglia, bone, cartilage, other somatic tissues) |
| [27] | Pa | f | Congenital carcinoma | Undifferentiated adenocarcinoma; size 4×2 cm; infiltration of skin, ear and orbita; multiple irradiations; death with invasion of the brain and lung metastases |
| [13] | Pa | m | Congenital carcinoma | Undifferentiated small cell carcinoma; size of a pigeon's egg; excision 12 days post partum; recurrences at age of 5 weeks and 12 months; multiple lymph metastases; at age of 18 months free of tumour |
| [43] | Sm | m | Congenital carcinoma | Adenoid cystic carcinoma (glandular type); size 4.5×3.2×3 cm; no recurrence 3 years after removal |
| [11] | Sm | m | Congenital carcinoma | Adenoid cystic carcinoma (glandular type); size 3.5×2.8×2.8 cm (weight 16 g); perineural invasion; excision at age of 8 months; no recurrence |
| [16] | Pa | m | Congenital carcinoma | Epithelial–myoepithelial carcinoma; size 9 cm; recurrence at 4 months after surgery; death 2 months later with regional spread |
| [12] | Np | 7m 2f | Congenital pleomorphic adenoma | "Salivary gland anlage tumour"; pediculated; size 3 cm; biphasic pattern of squamous nests and duct-like structures; similar to the developing salivary glands; probably hamartoma of minor salivary gland derivates |

entiated embryonal basaloid cells are the major cell compartment in these adenomas and that myoepithelial cells are a smaller, peripherally located, compartment. The epithelial tumour cells show a combination of the two cytokeratin subfamilies [28, 29] with expression of cytokeratin polypeptides typical of stratified squamous epithelia and their maturation stages (CK 3, 5 and 6 of type II) but with the additional expression of cytokeratin polypeptides typical of simple columnar epithelia (CK 7 of type II and CK 13 and 19 of type I). The expression is mainly in the luminal duct-like cells, especially at their luminal borders and cell membranes. The other basaloid cells of the more solid cell nests show cytokeratin expression only in a few cells. Some myoepithelial cells are located at the periphery of the epithelial structures and express CK 14 and vimentin and moderately muscle-specific actin.

Congenital hybrid basal cell adenoma–adenoid cystic carcinoma is an association of two tumour components, which are defined as a hybrid [36] or composite [4] tumour or as the evolution of adenoid cystic carcinoma from a basal cell adenoma [7]. In contrast to basal cell adenoma, congenital hybrid basal cell adenoma–adenoid cystic carcinoma is characterized by recurrence, facial nerve invasion and lymph node metastases in about 25% of the reported cases [23, 36]. In spite of mitotic figures and neural invasion as markers of aggression, however, haematogenous metastases are not observed.

The sialoblastoma as an embryonal tumour or "embryoma", represents a neoplastic proliferation of cells of organ rudiments [42] and is the most frequent type of congenital tumour of the salivary glands [5, 6, 8, 18, 20, 37, 39, 42]. The cases documented have been localized chiefly in the parotid gland, only one case being localized in the submandibular gland [18]. The characteristic structure is a tubulo-ductal phenotype with inclusion of acinar cell differentiation. The cells are arranged in a lobular structure with some similarity to basal cell adenomas. The epithelial structures include cell groups with an embryonic appearance, some sebaceous cell clusters, and foci of squamous differentiation or myoepithelial cell nests [18, 39]. The areas with ductal differentiation show a distinct expression of cytokeratin 8 (CAM 5,2) and a positive EMA reaction at the luminal surface, whereas the myoepithelial cells express S-100 protein [18]. A loose immature stroma develops between the epithelial cells. Less than 25% are characterized as malignant with elevated mitotic activity, recurrence and local lymph node metastases [20, 39]. The occurrence of mitotic figures alone does not indicate malignancy.

There is an indistinct histopathological border between sialoblastoma and congenital basal cell adenoma. Only one case report of sialoblastoma mentions acinic cell differentiation with intracellular secretory granules [18]. All other reports describe solid and ductal epithelial structures without acinic cell elements [6, 20, 39, 42]

and PAS-positive substances in the duct-like structures [6, 42], and some mention peripheral palisading cells with basal membrane substances [6, 42]. In comparison with congenital basal cell adenoma, there are also myoepithelial cells present in sialoblastoma, with expression of vimentin, actin or S-100 protein [18, 20, 39]. The architecture of some tumours classified as sialoblastoma may mirror various stages of branching morphogenesis of salivary gland development, but does not contain acinic cell complexes as part of a salivary gland anlage.

When the differential diagnosis of congenital basal cell adenoma and sialoblastoma is considered, it is evident that these two entities can be regarded as relatively similar tumours. In spite of the similar architecture, clinical data reveal differences in behaviour between basal cell adenoma and sialoblastoma. Basal cell adenomas have a mean diameter of 4 cm, a male predominance, and evidence of only one recurrence in a scar, whereas sialoblastomas are characterized by a mean diameter of 7 cm, a female predominance, and recurrences and/or regional lymph node metastases in about 25% of the documented cases. In follow-up studies, however, many cases of both tumour entities are free of tumour many years after presentation.

A pleomorphic adenoma [23] and a congenital sebaceous cell adenoma of the parotid gland [32] have been reported, and hamartomas occur; an acinar pattern may predominate, although every component seen in normal tissue is present. In one report a well-encapsulated and lobulated congenital hamartoma of the submandibular gland containing tubulo-ductal structures and acinar differentiation was described [5]. In another case, in a 16-year-old boy [41], the hamartoma was localized in the superficial lobe of the right parotid gland and was composed of serous and mucous gland acini, salivary ducts, myoepithelial cells, adipose and lymphoid tissue. The term "adenolipomatous hamartoma" characterizes this special histological composition.

The salivary gland anlage tumour or congenital pleomorphic adenoma, which is localized in the midline of the posterior pharyngeal wall of the nasopharynx [12, 17, 38], is probably also a hamartoma. This type derives from minor salivary glands and has a biphasic histological pattern of squamous nests and duct-like structures in the periphery, blended into solid, predominantly mesenchmal nodules centrally. The histological and architectural features are similar in some aspects to developing salivary glands.

Only one congenital teratoma, in a submandibular gland, is recorded in the literature [31]. The female newborn had a 2 cm diameter cystic swelling in the left submandibular region. In all other cases, congenital teratomas of the oronasopharyngeal area have been located in the palate, tongue and floor of the mouth, or in the vicinity of the thyroid gland. One case of a well-differentiated teratoma is reported in the parotid gland of a 24-year-old woman [35].

Congenital carcinomas of the salivary glands are extremely rare. Only five cases are reported in the literature [11, 13, 16, 27, 43] which are listed in Table 4. Other rare carcinomas in the first year of life are mucoepidermoid carcinomas [19, 23], adenocarcinomas without exact histological subclassification [22] and undifferentiated carcinomas [21]. The most common carcinoma in children is the mucoepidermoid carcinoma, followed by adenoid cystic carcinoma and acinic cell carcinoma [16, 26, 34]. All carcinomas of this age group are characterized by infiltrative growth, recurrence and lymph node or haematogenous metastases. An embryonal carcinoma of the parotid gland was observed in a 12-year-old boy with infiltrative growth, lung metastases and a lethal course [15].

There are no reports of congenital sarcomas of salivary glands. In the first decade a few cases of embryonal rhabdomyosarcomas have been described [2, 3, 25, 26].

In the group of benign non-epithelial tumours the congenital capillary haemangioma (juvenile haemangioma or benign infantile haemangio-endothelioma) predominates [30].

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