Case Report

Sialoblastoma of parotid gland: A rare case report and review of literature

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ABSTRACT

Sialoblastoma is a rare congenital tumor of salivary gland. Herein, we report a case in a 5-year-old child who presented with a painless swelling in parotid region since birth and also review the literature. Surgical excision of parotid gland was done. Further, histopathological and immunohistochemical examination was performed and a final diagnosis of sialoblastoma was made. Patient is on follow-up 6 monthly.

Key words: Congenital tumor, parotid gland, sialoblastoma

INTRODUCTION

Fewer than 5% of all salivary gland tumors occur in children.[1] Tumor that originate in the salivary gland ductal or secretory epithelial cells are exceedingly rare in children. The following three types of epithelial tumors can be differentiated - adenomas comparable to adult forms, sialoblastomas or embryomas and carcinomas. A group of tumors, sialoblastoma has been recognized that usually present at birth or shortly thereafter.[2] It represents a neoplastic proliferation of cells of organ rudiments and it is the most frequent type of congenital epithelial tumor of the salivary glands.[3] A total of 30 cases have been reported in the literature. We describe in this report the clinical and pathological findings in a 5-year-old child and review the literature.

CASE REPORT

A 5-year-old boy presented for the first time at our hospital with the complaint of a gradually increasing painless swelling in left parotid region, which appeared soon after birth. The mass measured 6 cm in diameter and had a firm consistency. Ultrasonography of the left parotid region was performed, which showed an expansile and enhanced mass lesion with well-defined irregular borders. A fine-needle aspiration was performed from the mass. Smear revealed variably arranged, tight solid clusters of atypical appearing basaloid like cells in a background of dispersed epithelial and myoepithelial cells. The clusters contained admixed benign ductal cells and dense metachromatic magenta hyaline globular material with smooth rounded outlines. Differential diagnosis included neoplasms composed of either basaloid cells and/or admixed hyaline matrix material such as pleomorphic adenoma, basal cell adenoma, sialoblastoma, and adenoid cystic carcinoma. A presumptive diagnosis of undifferentiated neoplasm with closest resemblance to sialoblastoma was given. The patient underwent surgery. Frozen section was requested. Multiple grey white soft tissue pieces were received measuring together 1.0 × 0.5 × 0.5 cm. Frozen sections were examined and a diagnosis of sialoblastoma with reactive lymphadenitis was given. Total parotidectomy with lymph node excision was done and the specimen sent for histopathology. On gross examination, there was a single large multilobulated firm grey white to grey brown tissue mass measuring 6 × 4.5 × 3 cm. External surface was congested. Cut section revealed grey white to grey brown tumor along with rim of normal salivary tissue. Microsections examined show a circumscribed tumor consisting of solid basaloid aggregates of epithelial cells surrounded by basement membrane deposition and separated by fibrous septa. The epithelial cells contained round to oval hyperchromatic

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nuclei, dispersed granular chromatin and inconspicuous nucleoli [Figure 1]. The cytoplasm was amphophilic with distinct cell borders. In some of the nests, the ductal lumens contained dense basophilic secretory material, which showed positive staining for PAS [Figure 2]. The cells showed positivity for cytokeratin [Figure 3]. There was no necrosis and mitotic activity was minimal as evident by Ki 67 negativity [Figure 4]. Areas of ductal differentiation showed strong positive staining with cytokeratin. Final diagnosis of sialoblastoma was made.

DISCUSSION

Less than 5% of all salivary gland tumors occur in children and fewer than 0.25% are found in children under 10 years of age.[4] Batskis classified perinatal salivary gland tumors into four categories. The first category consists of histologically benign tumors comparable to adult counterparts e.g., pleomorphic and monomorphic adenomas. The second category includes the hamartomatous tumors. The third group which includes sialoblastoma is described as tumors histologically reminiscent of the embryonic epithelial anlage of the major salivary glands at various stages of development. Thus, these tumors demonstrate varied branching morphogenesis and cytodifferentiation. The fourth group consists of tumors which are biologically and histologically malignant.[1,4]

The first description of sialoblastoma dates back to 1966 when Vawter and Tefft who called it embryoma because of the similarity to embryonal salivary gland tissue. They suggested that these lesions may have a local recurrences and regional high mitotic activity.[5] The "salivary gland unit" theory of Batsakis hypothesized that neoplasia within salivary gland results from the activation and proliferation of resting multipotent reserve cells. While this may be true in adults, Taylor in 1988 attributed the histogenesis of congenital tumors to the disordered proliferation of blastemous cells rather than to activation of resting reserve cells. He also suggested the term sialoblastema to convey both the dysontogenic character and salivary origin of these tumors.[6]

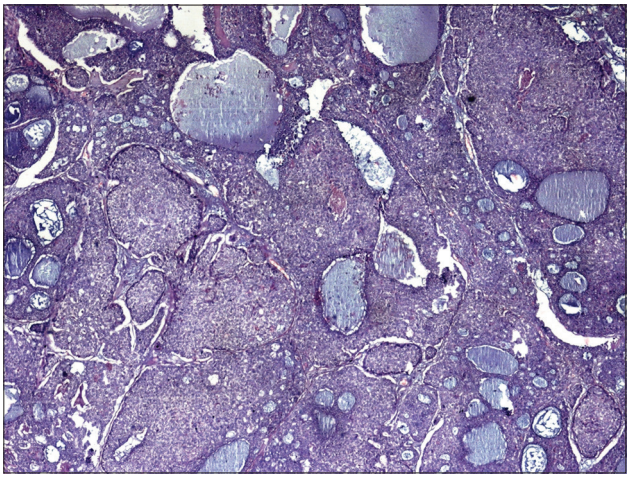

Figure 1: Tumor cells arranged in nests (H and E, ×40)

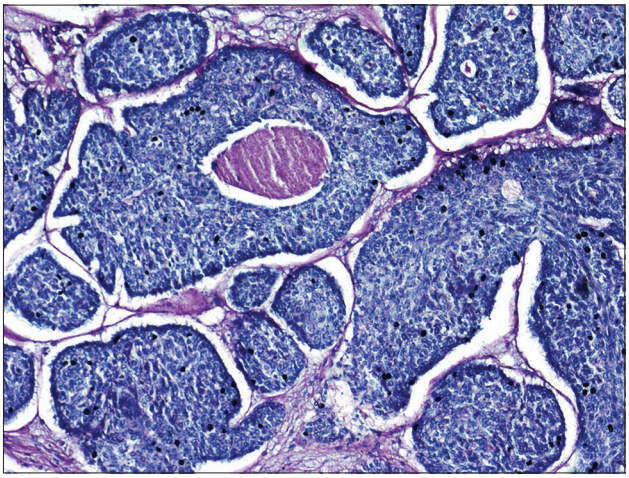

Figure 2: PAS positive material in the lumen (PAS, ×100)

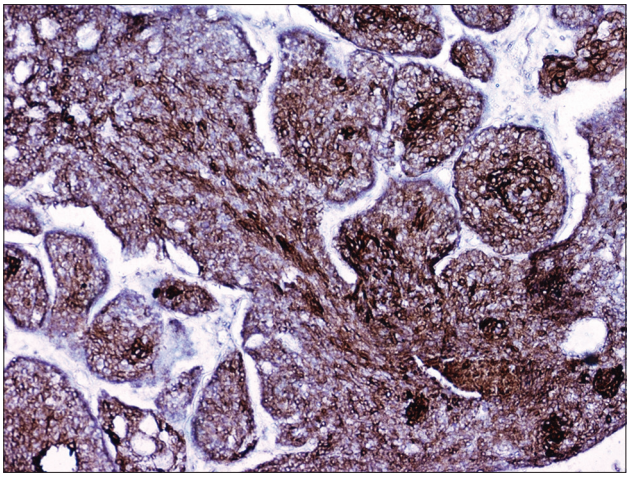

Figure 3: Cytokeratin positivity in tumor cells (IHC, ×100)

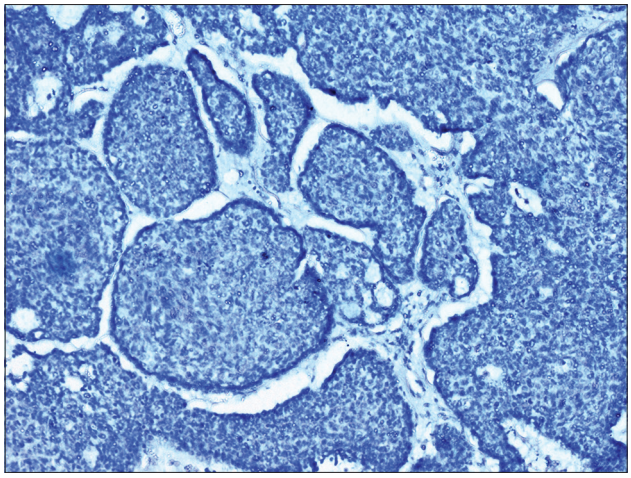

Figure 4: Ki67 negativity in tumor cells (IHC, ×100)

The term "sialoblastoma" is currently favored and accepted by clinicians and pathologists. They usually present within the first few months of life as subcutaneous masses from 2 to 8 cm in size in the parotid region. They are locally aggressive but are not thought to spread systemically. The long term biological behavior of these tumors is uncertain.[7] As Brandwein *et al*., pointed out the distinction between benign and malignant sialoblastoma may not be as well-defined as originally thought. The ultimate tumor behavior will be defined by the histologic grade as well as tumor stage and the extent of resection.[2] Batsakis and Frankenthaler advocated that approximately 25% of these rare tumors have been histologically and/or biologically malignant. He also proposed histologic criteria for assessment of malignancy in sialoblastomas, which included invasion of nerves or vascular spaces and ancillary findings of necrosis of cells with cytologic atypia beyond that expected or presumed for an embryonic epithelium.[1,4]

The various other tumors, which resemble to sialoblastoma are: Congenital basal cell adenoma, embryonal carcinoma, basaloid monomorphic adenoma, and adenoid cystic carcinoma. Congenital basal cell adenoma shows many similarities to sialoblastoma, but shows clear sebaceous differentiation and lacks myoepithelial cells.[8] Embryonal carcinoma consists of solid and medullary areas, some showing acinar patterns, with epidermoid foci and keratinization.[9] Basaloid monomorphic adenomas are characterized by prominent peripheral palisading of the nuclei, lack of pleomorphism or mitotic activity, and a benign, generally nonrecurrent clinical course.[6] The presence of a cribriform pattern developed within the solid sheets of cells in adenoid cystic carcinoma is different from sialoblastoma. Ultimately, it is the grade, stage and extent of resection that will dictate the prognosis and possible efficacy of alternative treatments.[10]

For an individual case, it may be difficult to predict the most appropriate therapy as advocated by Tatlidede *et al*.,[3] Surgical excision is the primary treatment of choice. They can be treated with early conservative surgery alone provided that free margins are obtained. Local recurrence or persistence is not uncommon. Sialoblastomas may become more anaplastic over time and this histologic change may predict local recurrence and possibly the risk of distant metastasis. In the presence of persistent or locally recurrent tumor, chemotherapy or radiotherapy can be used to prevent distant spread.[2,5]

CONCLUSION

To conclude, Sialoblastoma is a rare tumor of salivary gland and should be included in the differential diagnosis of the childhood facial tumors and an early diagnosis should be made to prevent tumor spread.

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