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Case Report

Pediatric Submandibular Pleomorphic Adenoma: Three Cases and Role of Fine Needle Aspiration Biopsy

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Abstract: Pediatric submandibular pleomorphic adenomas are very rare benign tumors. Fine needle aspiration biopsy is a useful preoperative diagnosis to avoid tumor rupture and seeding. We present three cases of pediatric submandibular pleomorphic adenoma.

Keywords: Pleomorphic Adenoma; Pediatric; Submandibular Gland; Tumor

1. Introduction

Among the pediatric population, salivary gland tumors constitute less than 10% of all head and neck tumors. While 85% of these tumors originate from the parotid gland; only 10% originate from the submandibular gland [1]. Pediatric submandibular pleomorphic adenomas are very rare, and there are few series with a small number of patients in the literature. Therefore, the clinical features of these tumors have not been fully elucidated. In this article, the clinical and histopathological characteristics of three patients who underwent surgery for pediatric submandibular adenoma between 2019 and 2022 are presented.

2. Cases

The study was approved by the University Clinical Research Ethics Committee (Number: 2024-02-02). Parental consent for publication was obtained.

The average age of the patients was 14.2 years, with individual ages of 10, 16, and 17 years, respectively. All patients were admitted due to painless swelling in the neck. Two patients were male, and one patient was female. The swelling was on the right side in two patients and on the left side in one patient. The average duration of symptoms was 6 months. On examination of the patients, there were painless, hard, mobile masses in the submandibular area (Figure 1).

Figure 1. Mobile, painless, hard swelling on the left submandibular region of a 10-year-old female.

Firstly, neck ultrasonography (USG) was performed on the patients as an imaging method. The common feature of the masses on USG was that they appeared hypoechoic, well-circumscribed, ovoid, or round. All patients underwent USG-guided fine-needle aspiration biopsy (FNAB), and the result was compatible with pleomorphic adenoma (Figure 2). Contrast-enhanced neck magnetic resonance imaging (MRI) was performed on all patients. The common features of the masses appeared hypointense in the T1 series and hyperintense in the T2 series on MRI. They showed heterogeneous contrast enhancement (Figure 3). All patients underwent submandibular gland excision, including total excision of the tumor (Figure 4). A facial nerve monitor (NIM Response 3.0, Medtronic, USA) was used during the operation on two patients. No postoperative complications or marginal nerve paresis were observed in any patient. In histopathological examination, sheet forming and single, benign epithelial and myoepithelial cells in the chondromyxoid matrix were revealed. Microscopic findings were compatible with pleomorphic adenoma (Figure 5). The tumor dimensions were 25 × 18 × 15 mm, 35 × 30 × 30 mm, and 30 × 25 × 20 mm. The follow-up periods were 12, 48, and 12 months, respectively. No recurrence was observed during the follow-up.

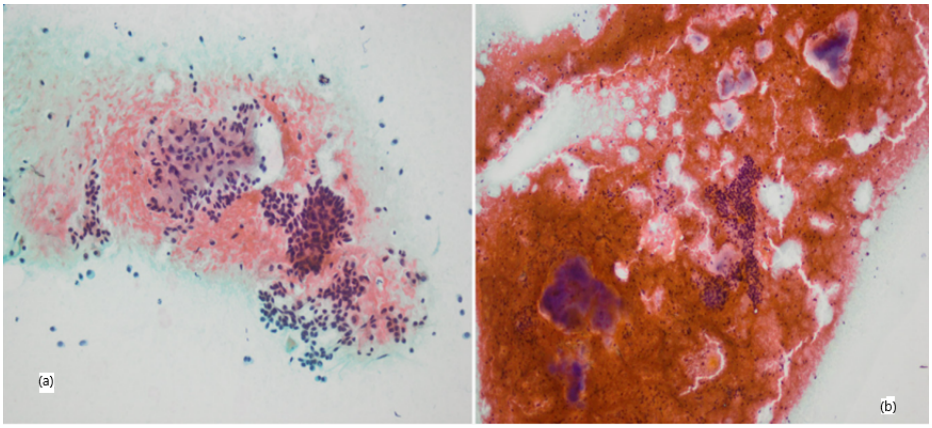

Figure 2. (**a**) In PAP staining, blue-purple colored material compatible with a possible chondromyxoid matrix was observed. (**b**) Cell islands with oval nuclei, narrow cytoplasm, occasionally forming a papillary structure, an occasionally oncocytic appearance, and no cytological atypia. The findings are consistent with pleomorphic adenoma.

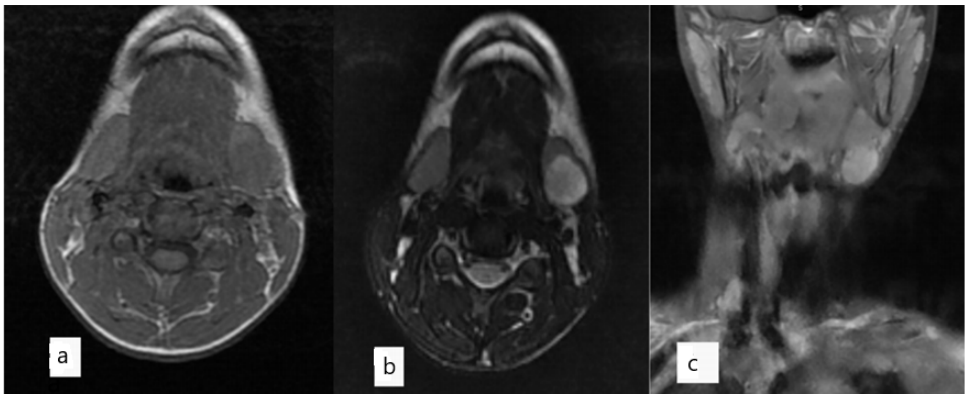

Figure 3. MRI shows right (**a**) hypointense submandibular mass in axial T1-weighted image sequence, (**b**) hyperintense in axial T2-weighted sequence, and (**c**) heterogenous contrast enhancement in contrast-enhanced series.

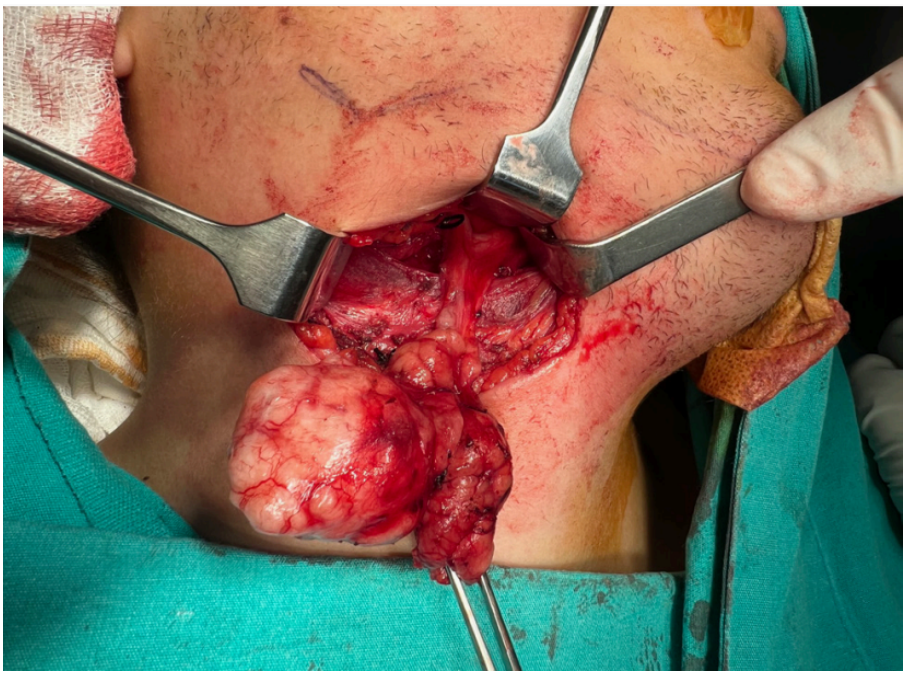

Figure 4. Intraoperative image of the right submandibular gland and tumor of a 16-year-old male.

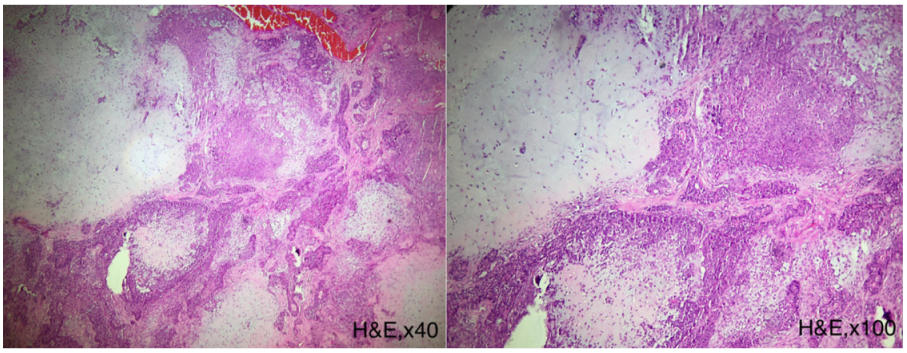

Figure 5. Typical myxoid stroma, next to myoepithelial cells that surround ductal structures and are structured in the form of wide layers (Hematoxylin-Eosin Stain, x40 and 100). Pleomorphic adenoma.

3. Discussion

Pediatric submandibular pleomorphic adenomas are very rare tumors. They present as a slowly growing, mobile, painless swelling in the submandibular region and mostly occur between the ages of 10 and 14 [2]. It is important to eliminate infectious and inflammatory causes in differential diagnosis. In a review by Masumoto et al. [2], only 22 patients were reported in the Japanese literature. The average age was 12 years, and the symptoms lasted 40.5 months. The average diameter was 2.9 cm. The authors stated that local recurrence mainly occurs as a result of rupture of the tumor capsule during the operation or because the tumor borders are not clearly distinguished. In this study, local recurrence was observed in 2 of 8 patients whose tumor capsule ruptured. A malignant component was observed in only one patient. Xu et al. [3] reported only 3 submandibular pleomorphic adenomas in their study examining 53 salivary gland tumors aged < 20 years from 1992 to 2016. All of our patients were diagnosed with preoperative FNAB. With the preoperative diagnosis of pleomorphic adenoma, careful total excision of the glandular tissue without rupturing the tumor's capsule is very important to prevent recurrences, considering the long life expectancy in children.

USG, computed tomography (CT), and MRI can be used for imaging. Ultrasonography is the most important method in diagnosis. It is used to determine whether the lesion originates from the gland, to differentiate between solid and cystic, and, when necessary, for fine needle aspiration biopsy. On USG, it appears hypoechoic and well-circumscribed. CT and MRI may be used. MRI is more reliable in distinguishing benign from malignant and detecting its relationship with surrounding soft tissues. On MRI, pleomorphic adenomas appear well-circumscribed, round, and hyperintense on T2 images. It is mostly hypointense or isointense on T1 images. Additionally, enhanced capsule structure and lobulation are distinctive for pleomorphic adenoma. Most preoperative diagnoses of PA are based on FNAB due to its high diagnostic accuracy, as reported in the literature, up to 95% [4].

Fang et al. [4] reported 122 patients who underwent surgery for epithelial salivary gland tumors between 1987 and 2011. Benign tumors were observed in 105 patients, and malignant tumors in 17 patients. 21 cases were diagnosed as submandibular pleomorphic adenomas. Out of 41 patients under 20 years of age operated on by Dombrowski et al. [5] for salivary gland pleomorphic adenoma between 1998 and 2017, 7 (21.9%) had tumors originating from the submandibular gland. Preoperative FNAB was performed in only 8 patients. USG was used in 9 patients, CT in 15 patients, and MRI in 23 patients. In our cases, preoperative USG and MRI imaging were performed, but CT was not used. MRI is especially useful for assessing the tumor's relationship with the gland and surrounding structures. In addition, it is superior to CT in that it does not cause radiation exposure in children.

The main treatment for submandibular pleomorphic adenoma involves the complete excision of the tumor along with the submandibular gland. When enucleation is performed, the recurrence rate can range from 30% to 80% [5]. Therefore, enucleation should be avoided. In our series, all patients underwent total submandibular gland excision via a transcervical approach. No instances of capsular rupture were observed in any of the patients, and they remain under follow-up without any signs of recurrence. Despite being controversial, FNAB is useful in diagnosis in the preoperative period. In cases of doubt, incisional or excisional biopsy should be avoided to prevent the risk of tumor transplantation.

Author Contributions

Concept—H.C.İ., M.Ö., G.I.; design—H.C.İ., M.Ö., G.I.; supervision—H.C.İ.; resources—H.C.İ., M.E.Ö.; materials—H.C.İ., M.E.Ö.; data collection and/or processing—H.C.İ., M.Ö., G.I.; analysis and/or interpretation—H.C.İ., M.E.Ö., M.Ö., G.I.; literature search—H.C.İ.; writing—H.C.İ., M.Ö., G.I.; critical review—H.C.İ.

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Institutional Review Board Statement

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. The study was approved by Health Sciences University Bursa Yuksek

Ihtisas Training and Research Hospital Ethics Committe (Protocol number: 2024-TBEK 2024/02-02, date: February 2, 2024).

Informed Consent Statement

Verbal informed consent was obtained from the parents of the patients who agreed to take part in the study.

Data Availability Statement

The data will be available upon request to the corresponding author.

Conflicts of Interest

The authors declare that they have no conflict of interest.

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