

## Submandibular gland pleomorphic adenoma in a seven-year-old child: a case report

Yedi yaşında bir çocukta submandibüler bez pleomorfik adenomu: Olgu sunumu

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Salivary gland neoplasms are rare in the pediatric age group. Pleomorphic adenomas in the submandibular gland are rarer. In this article, we present a seven-year-old female with a slowly growing mass in her right submandibular area. The firm, mobile and painless mass was about 2x3 cm in size and with bimanual palpation it was indiscriminated from the submandibular gland. Magnetic resonance imaging with contrast revealed a heterogeneous and minimally lobulated mass within the submandibular gland with clearly defined borders. Fine needle aspiration biopsy revealed a diagnosis of pleomorphic adenoma and we performed right submandibular gland excision under general anesthesia. The histopathological diagnosis was pleomorphic adenoma with sparse mitotic figures that may be interpreted as having a potential of malignant transformation. This patient who was followed up for one year without any complication is to our knowledge the youngest case cited in the English-language literature.

**Key Words:** Pediatric pleomorphic adenoma; submandibular gland; tumor.

Tükürük bezi neoplazmaları pediatrik yaş grubunda nadir görülür. Submandibüler bezde pleomorfik adenom ise çok daha nadirdir. Bu yazıda, sağ submandibüler bölgede yavaş büyüyen kitlesi olan yedi yaşında kız olgu sunuldu. Sert, hareketli ve ağrısız kitle yaklaşık 2x3 cm boyutundaydı ve iki elle yapılan muayenede submandibüler bezden ayırt edilemiyordu. Kontrastlı manyetik rezonans görüntüleme ile submandibüler bez içerisinde sınırları belirgin, heterojen ve minimal lobulasyon gösteren kitle tespit edildi. İnce iğne aspirasyon biyopsisiyle pleomorfik adenom tanısı konuldu ve genel anestezi altında sağ submandibüler gland eksizyonu yapıldı. Kitlenin histopatolojik tanısı pleomorfik adenom idi ve görülen seyrek mitotik aktivite malign transformasyon potansiyeline sahip olduğunu düşündürüyordu. Bir yıldır komplikasyonsuz bir şekilde takibimizde olan bu hasta, bilgilerimize göre İngilizce literatürde bildirilmiş en genç olgudur.

**Anahtar Sözcükler:** Pediatrik pleomorfik adenom; submandibüler bez; tümör.

In childhood, most of the lesions occupying the submandibular triangle are inflammatory and usually resolve with medical treatment without surgical intervention.<sup>[1]</sup> In general, salivary gland

tumors comprise about 3-4% of head and neck tumors; less than 5% of these occur in childhood.<sup>[2]</sup> As in adults, salivary gland tumors usually occur in major salivary glands in the pediatric age group.

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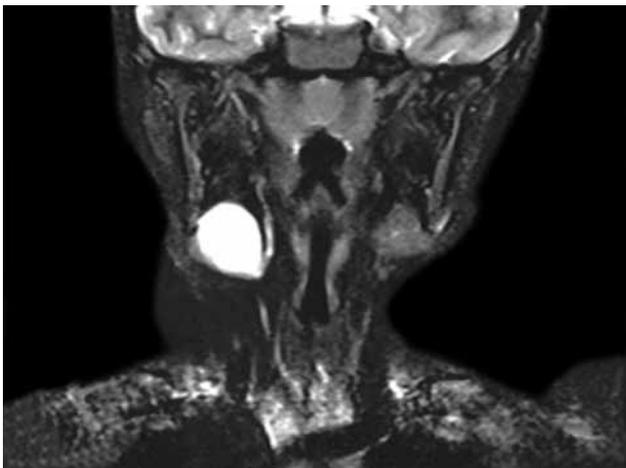
The parotid gland has the highest site of occurrence while the submandibular gland is involved in about 10% of the cases. Submandibular gland tumors are exceedingly rare in childhood, most being pleomorphic adenomas.<sup>[3-6]</sup>

In the largest series published by Krolls et al.,<sup>[7]</sup> only 10 submandibular pleomorphic adenomas in children had been reported up to 1972, and after that publication, only one case was reported by Molina et al.<sup>[8]</sup> In this study we discuss the presentation, diagnosis and treatment of a pediatric case of submandibular pleomorphic adenoma.

### CASE REPORT

A seven-year-old female patient consulted our department with a complaint of painless swelling under her chin on the right side. It was a slowly growing mass presenting for more than a year. There were no other accompanying symptoms.

Otorhinolaryngological examination revealed a firm, mobile, and painless mass in her right submandibular area. The mass was about 2x3 cm in size and with bimanual palpation, it was indistinguishable from the submandibular gland. There was no neural weakness or lymphadenopathy on cervical palpation. The other physical examination findings were unremarkable. Hematological tests were within normal limits. Magnetic resonance imaging (MRI) with contrast revealed a heterogeneous and minimally lobulated mass within the submandibular gland, which was 21x22x26 mm in size. The borders of the lesion were clearly defined (Figure 1). Fine needle aspiration revealed a diagnosis of pleomorphic adenoma. A right



**Figure 1.** Coronal T2 weighted magnetic resonance imaging section reveal a solid mass with a hiperintense heterogeneity in the submandibular gland.

total submandibular gland excision was performed under general anesthesia. On gross examination, the specimen was a grayish white tumoral mass, 3x2x2 cm in size, within the submandibular gland with clear-cut borders differentiating the glandular tissue from the mass.

The patient was discharged from the hospital the day after the operation without any complications. The final histopathological diagnosis was a pleomorphic adenoma with sparse mitotic figures that might be interpreted as having a potential of malignant transformation (Figure 2a, b). The patient was totally normal in the one-year follow-up period.

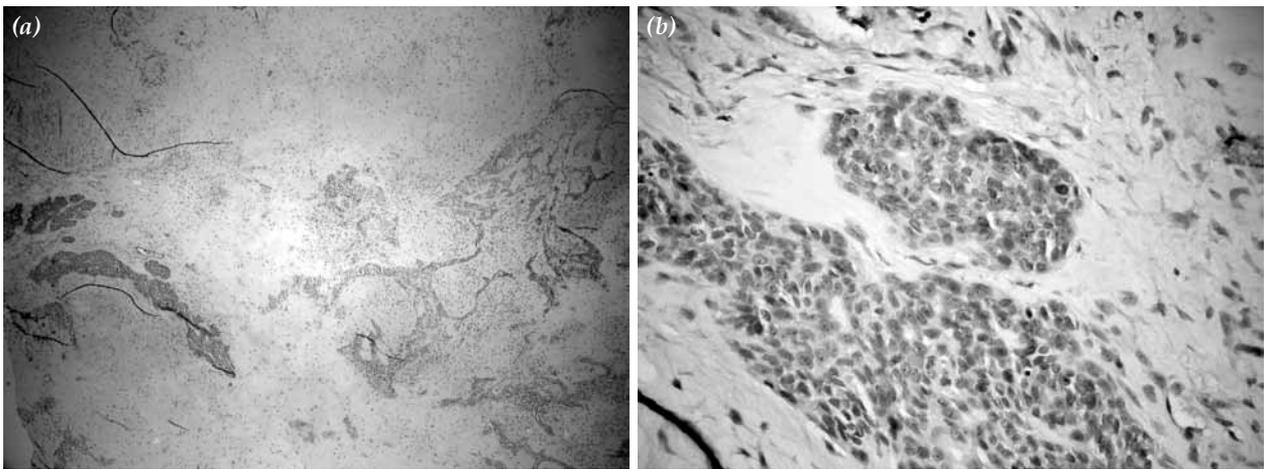
### DISCUSSION

Salivary gland masses in childhood are mostly vascular in origin-hemangiomas or lymphangiomas. As far as solid masses are concerned, mostly pleomorphic adenomas are observed.<sup>[9]</sup> Mostly seen in the parotid gland, pleomorphic adenomas account for 40-60% of all submandibular gland neoplasms.<sup>[9-12]</sup>

Pediatric salivary gland neoplasms usually occur in puberty or post puberty in contrast to our patient, who was only seven-years-old.<sup>[4,5]</sup>

Other than the very young age, another interesting finding of our case was the presence of sparse mitotic figures pointing to malignant transformation (although it was reported as pleomorphic adenoma). Salivary gland neoplasms in children have a malignancy rate of about 50%, in contrast with rates in adults which range between 15-25 percent.<sup>[3,5,13]</sup> The most common malignant salivary gland tumor in children is mucoepidermoid carcinoma, and it is usually encountered in parotid gland. To our knowledge, malignant mixed tumor in the submandibular gland has not been cited so far in that age group. Left untreated, malignant transformation might have taken place in such a young girl.

The radiological evaluation of submandibular neoplasms usually includes MRI, computed tomography (CT) and ultrasound (US). Although the usual concept is that MRI and CT are nearly equally effective in diagnosing masses in head and neck area, a recent study points out that MRI is better in diagnosing pleomorphic adenomas.<sup>[14]</sup> Magnetic resonance imaging is better in defining the soft tissue invasion. Other than radiation exposure, disadvantages of CT include the use of iodinated contrast material, and poorer soft



**Figure 2.** (a) A microscopic view of pleomorphic adenoma with the epithelial cell rows and clusters in myxocondroidal stroma. The cell islands located on the left are recognized to stained more hyperchromatic than the rest (H-E x 40). (b) A closer look at the area that stained darker in Figure 2a. The nuclei are more prominent and mild coarsening of the chromatin is recognized. There are some mitosis in the central area (H-E x 400).

tissue distinction. Computed tomography may also be problematic in cases of benign pleomorphic adenomas when the outer margin of the tumor appears indistinct and may falsely suggest malignant invasion of the surrounding tissue. Magnetic resonance imaging shows the well-defined outer borders in these instances. Occasionally, however, CT may be the only study needed to guide the surgeon. In such tumors, ultrasonography is usually performed with poorer diagnostic confidence so we did not perform US. We preferred MRI to prevent radiation exposure of the patient and for its better soft tissue identification.

Although the sensitivity and specificity is not 100%, fine needle aspiration is considered one of the major diagnostic tools in head and neck masses. Fine needle aspiration (FNA) cytology is reported to well demonstrate most of the histological features of pleomorphic adenoma of the salivary gland and may be considered a useful tool in initial assessment of the tumor despite its histological diversity.<sup>[15,16]</sup> Accordingly, to complete our preoperative work and to have more confidence in the diagnosis we performed a FNA of the mass. It revealed a diagnosis of pleomorphic adenoma. That the sparse mitotic figures as a sign of malignant transformation were shown in histopathological examination of the specimen but not in FNA cytological examination may be attributed to the paucity of the material of the aspirate.

Surgery is the treatment of choice. Enucleation is not recommended because of the tumoral growth

pattern which shows small protrusions beyond the capsule and the risk of leaving capsular remnants during the enucleation. The risk of recurrence with enucleation increases to as high as 50% in contrast to en-bloc resection of the tumor with the gland that ranges between one to five percent.<sup>[3]</sup> The studies performed so far clearly show that total resection of the tumor with the gland but not enucleation is the treatment of choice since it is associated with lower recurrence rate.<sup>[11,17]</sup> We performed a total en bloc resection of the tumor with the submandibular gland. There was no remarkable lymphadenopathy near the specimen and the follow-up period was free of recurrence, despite being relatively short.

In conclusion, submandibular gland pleomorphic adenomas are very rare. When diagnosed they must be evaluated as in adults and en bloc resection of the tumor with the gland is the treatment of choice.

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