

Incidentally Detected Agenesis of Left Submandibular Gland in a Case of Papillary Carcinoma Thyroid - A Case Report

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Abstract: *Congenital agenesis of unilateral submandibular gland is not a commonly seen disorder. Presentations such as dysphagia, problems in dentition or xerostomia may be seen but the patients may also be symptomless. Till now most of the reported cases are associated with other major salivary gland hypertrophy or with some congenital disorders including lacrimoauriculodentodigital syndrome, facial anomalies and ectodermal dysplasia associated with aplasia or dysplasia of lacrimal or thyroid glands. We report a case of unilateral submandibular gland agenesis associated with metastatic Papillary carcinoma thyroid without any compensatory hypertrophy of other salivary glands substantiated by the findings of MR Imaging.*

Keywords: agenesis, submandibular gland, carcinoma thyroid

1. Introduction

Congenital agenesis of salivary glands is a rarely encountered imaging finding and unilateral submandibular gland agenesis without compensatory hypertrophy is even more infrequent in clinical practice [1]. Clinical features such as dysphagia, problems in dentition or xerostomia can be seen, but the patients are mostly found to be asymptomatic in adult age groups [2]. Only about 42 cases of major salivary gland agenesis have been described in English literature [2]. The aetiology remains uncertain and has been postulated to result from abnormalities during organogenesis in foetal life, and association with many congenital anomalies like lacrimoauriculodentodigital syndrome, facial anomalies and ectodermal dysplasia associated with aplasia or dysplasia of lacrimal or thyroid glands [2,3]. To the best of our knowledge, this is the first case of unilateral submandibular agenesis associated with metastatic papillary carcinoma of thyroid without any compensatory hypertrophy of other major salivary glands or association with congenital head/neck anomalies. Our findings were substantiated by MRI and histopathological findings.

2. Case Report

A 65 year old male presented to our institute with complains of painless swellings in infrahyoid neck in left paramedian location and left supraclavicular region since 3 and 2 weeks respectively. On local examination there was a swelling in left thyroid region measuring approximately 5 x 4 cm and palpable left supraclavicular lymphadenopathy. Histopathological evaluation of these swellings was suggestive of papillary carcinoma of thyroid. For assessment of extension and involvements, patient underwent MRI neck. The imaging findings were suggestive of a left thyroid lobe altered signal intensity lesion measuring 4.8(AP) x 4.7(TR) x 6.7(CC) cm seen displacing the trachea medially and left carotid, subclavian arteries laterally. No intrathoracic extension was seen. Right lobe of thyroid & isthmus were unremarkable (Figure 1). In addition, there was absence of left submandibular gland (Figure 2a, 2b). Rest of the major salivary glands were present with normal

size, position and signal intensity. Thereafter, this patient was lost to follow up and could not be managed at our institute.

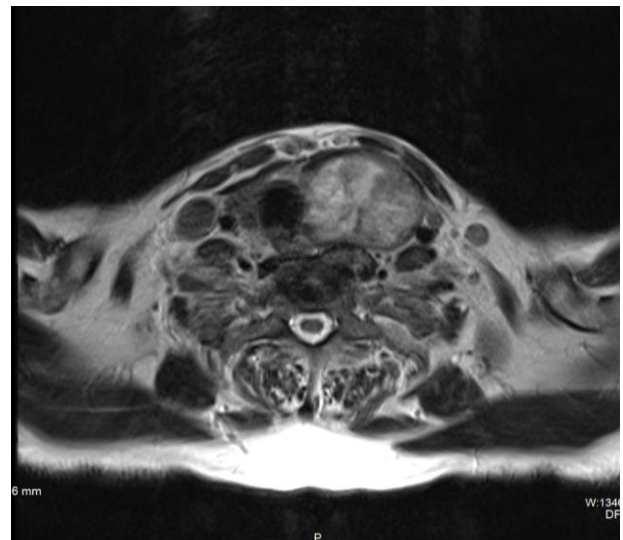


Figure 1: T2 weighted axial MR image showing left thyroid lobe heterogeneous signal intensity mass with areas of cystic necrosis.



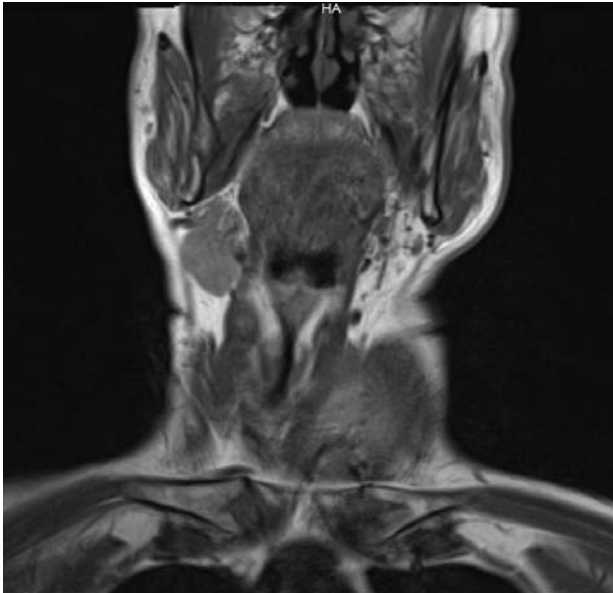


Figure 2a and 2b: T1 weighted axial and coronal images showing absence of left submandibular gland and left thyroid mass.

3. Discussion

Since the first report of bilateral SMG aplasia in 1885 by Gruber, not more than 42 cases have been reported in the English literature [4]. Majority of the previously reported cases in literature were either associated with compensatory hypertrophy of other major salivary glands simulating tumoral masses at imaging or associated with other congenital anomalies [5,6,7,8]. Commonly associated congenital anomalies with salivary gland agenesis, include those of first branchial arch like mandibular dysgenesis (Treacher-Collins syndrome), Orbital anomalies and lacrimoauriculodentodigital syndrome including hypoplasia, atresia of the lacrimal system, deafness and ear anomalies, and dental and digital malformations [3,9]. *Janani Moorthy et al* reported a case of unilateral submandibular gland agenesis associated with capillary hemangioma of the cartilaginous nasal septum [10].

In our case there is unilateral left submandibular gland agenesis associated with papillary thyroid carcinoma with metastatic supraclavicular lymphadenopathy. There was no associated compensatory hypertrophy of other major salivary glands or congenital anomalies. Reports of thyroid dysplasia's associated with submandibular gland agenesis are present in literature, however to the best of our knowledge our case is the first case to be associated with metastatic thyroid neoplasm.

4. Conclusion

The purpose of our case report is to familiarize the radiologists with this rare entity so as to eliminate the chances of overlooking coexisting malignant thyroid neoplasms even in those cases of submandibular gland agenesis where there is no associated congenital anomalies or hypertrophy of other major salivary glands.

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