ECTOPIC PAROTID TISSUE WITH CONGENITAL UNILATERAL AGENESIS OF THE PAROTID GLAND: A RARE CASE PRESENTING WITH RECURRENT UNILATERAL CHEEK SWELLING.

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Background

Congenital absence of parotid gland is rare. It usually occurs due to abnormal development of ectodermal tissue of the oral cavity. Aplasia can be either unilateral or bilateral and the degree of aplasia determines the extent of clinical manifestations of the condition. It is usually associated with other craniofacial abnormalities. Ectopic parotid tissue can be seen with unilateral or bilateral aplasia of the parotid gland and undergoes the same pathological processes as normal parotid glandular tissue. We present a case of unilateral aplasia of the parotid gland with associated ectopic parotid tissue without any associated craniofacial abnormalities.

Case report

A 41 year old male patient was referred to us by the surgical unit, with recurrent swelling of the right cheek over a period of one year duration. The swelling presented on and off to a varying degree and was sometimes tender to touch. It did not exacerbate following meals. He does not have any associated co morbidities and does not complain of a history of xerostomia, dry eyes, dysphagia, dental caries or halitosis. He did not develop any systemic symptoms during these episodes, has not sought medical treatment previously and his inflammatory markers when presented to us was only slightly elevated (WBC – 13,000 per mm with 78% neutrophils, CRP 10mg/dl)

On examination there was a soft swelling on the lateral aspect of the masseter muscle with no overlying skin changes. It was tender to touch and fluctuant. Intra oral examination revealed normal opening of the Stenson's duct on the left side, however the right side ductal opening was not identified. Stimulation with a sialogogue did not improve visualization. He did not have any associated craniofacial abnormalities.

Ultrasound scan (USS) was performed (Toshiba 7.5MHz frequency, linear probe) which revealed a well-defined, small hypoechoic rounded lesion over the lateral aspect of the right masseter muscle. Significant internal vascularity was not detected. There was no suspicious cervical lymphadenopathy. Left parotid gland was normal in position and echotexture. Right parotid gland was not normally identified in the anatomical position. Bilateral submandibular glands were identified in normal position and echo texture.

Fine needle aspiration cytology (FNAC) of the lesion revealed neutrophilic infiltrates with few macrophages and blood cells. No atypical or malignant cells.

Patient then underwent a Contrast enhanced computed tomography (CECT) study of the head and neck region. The study was done on a Toshiba Aquilion 16 slice machine with intravenous contrast (Omnipaque) 1ml/kg dose, and images were acquired at 30 seconds in early arterial phase. This revealed absence of normal parotid tissue in the right side with soft tissue stranding and fatty infiltration. There was a 2.3 x 1.2cm structure with hypodense central area and peripheral enhancement noted on the lateral aspect of the right masseter muscle. It appeared to draining in to an orifice in the oral cavity. The left parotid gland, bilateral sub mandibular glands and the rest of the neck was normal. Conclusion was an abscess formation in an ectopic parotid tissue in the absence of an ipsilateral normal parotid gland.

He was subsequently treated with oral antibiotics for 1 week during which the swelling disappeared. A repeat USS was performed in 3 months which showed the previous abscess completely resolved and the presence of a thin stripe of homogenous glandular tissue similar in echogenicity to normal parotid, situated lateral to the masseter muscle. The rarity of the condition and the investigative findings were clearly explained to the patient.

Discussion

Agenesis of salivary glands is rare and its incidence is difficult to ascertain because patients are often asymptomatic.[1] It can occur either as an isolated finding or can be associated with other genetic abnormalities, mainly autosomal pattern of inheritance leading to abnormal development of structures derived from first and second brachial arches. Some of the known associations are hemifacial microsomia, aplasia of lacrimal gland, mandibulofacial dysostosis and CHARGE syndrome.[2,3] Partial or total aplasia of unilateral or bilateral salivary glands, can lead to a wide variety of clinical symptoms from majority being asymptomatic to varying amounts of xerostomia.

Normal development of the parotid gland occurs during the 4^{th} -8^{th} week of embryonic life from the proliferation of ectodermal tissue in the oral cavity. The development of the primordial parotid glands are followed by the development of the submandibular and sublingual glands during organogenesis.[2] In a review of literature, Almadori et el reports that bilateral parotid gland aplasia is more common than unilateral parotid gland aplasia.[1] Usually, when an organ from a paired structure is congenitally absent, compensatory hypertrophy of the remaining organ is

normal. However, there are contradicting findings in literature with regards to the presence of compensatory hypertrophy of the parotid gland in a setting of congenital aplasia.[1,4] Certain metabolic disturbances such as diabetes, hypothyroidism can also give rise to atrophy of salivary glands with subsequent hypertrophy of the contralateral gland. [2] Clinically, the absence of the orifice of the salivary duct can be taken as evidence of developmental defect rather than atrophy subsequent to disease.[1]

Accessory parotid tissue is found in 20% of the population unilaterally.[5,6] It is commonly found in the lateral aspect of the masseter muscle, independent of the normal gland, superolateral to the course of the main parotid duct.[5] One of the main differentiations that needs to be made is with the presence of an anterior process of the parotid gland. This is parotid tissue which is connected to the normal gland and lying anterior to it. It is usually found in 20- 56% of the normal population.[5] Because of histological similarities, pathologies of the main parotid gland can also involve the accessory parotid tissue, including inflammatory processes and neoplastic changes.[5] 1-7% of parotid neoplasms originate in accessory parotid tissue.[6] As per our knowledge, unilateral congenital absence of a parotid gland with ectopic parotid tissue undergoing recurrent inflammation, was reported once before in literature.[2] However, there are four previously reported cases of accessory parotid tissue associated with aplasia of the main parotid gland, which were detected incidentally.[6,7]

A multi radiological approach is used for evaluation of pathologies of salivary glands, where the first line investigation was an USS. Though operator dependent, USS is freely available and easy to perform without the risk of radiation exposure. 7.5MHz linear probe is used to acquire images in two planes. Normal parotid gland shows homogeneous glandular tissue, hyperechoic to surrounding muscle. The absence of normal parotid tissue in the anatomical position of the gland should prompt a search for ectopic tissue in common locations. Ectopic parotid tissue can be identified as homogenous glandular tissue, similar in echotexture to the contralateral normal gland, situated over the masseter muscle. Inflammatory changes of ectopic parotid tissue will have similar radiological appearance to inflammation occurring in a normal parotid gland. In our patient who presented with recurrent cheek swelling, the initial USS showed imaging features suggestive of an abscess in the lateral aspect of the masseter muscle, with absent normal parotid tissue in the ipsilateral side. Follow up imaging following resolution of the inflammation will help confirm the presence of normal parotid tissue in an ectopic location.

We proceeded with a CECT of the head and neck region, as the second line investigation. It is non invasive and widely available in our institution. Another choice of cross sectional imaging would be a Magnetic Resonance Imaging (MRI). This will depend largely on the availability of the study and exclusion of contraindications for MRI. There are no large studies reporting the sensitivity and specificity of CT and MRI in the evaluation of ectopic parotid tissue. In both these modalities we expect to see the parotid fossa be devoid of normal parotid tissue and replaced with fat. The ectopic parotid gland will be identified lateral to the masseter muscle with similar attenuation/intensity to the contralateral normally situated gland. The Stenson’s duct can

be traced from the ectopic gland opening intra orally either at the normal position or in an ectopic position.

Management of ectopic parotid tissue can be either invasive or non invasive, based primarily on the clinical presentation and the pathological process involved. Invasive surgical resection is considered the mainstay of treatment, despite well known post surgical complications.[2] In asymptomatic, or medically managed inflammatory conditions, conservative management is an option.

In conclusion, even though it is rare, unilateral aplasia of the parotid gland with ectopic parotid tissue lateral to the masseter muscle, should be considered as an imaging differential in the assessment of patients with recurrent cheek swelling. Imaging is essential for diagnosis and a multiradiological approach consisting of at least two imaging modalities can help confirm the diagnosis.

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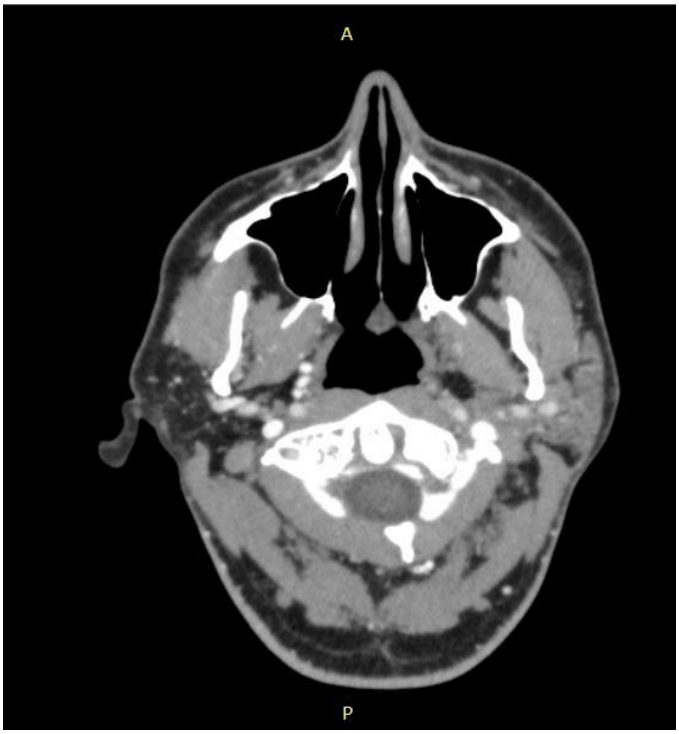

Figure 1: Contrast enhanced axial image shows absent gland in the right parotid fossa

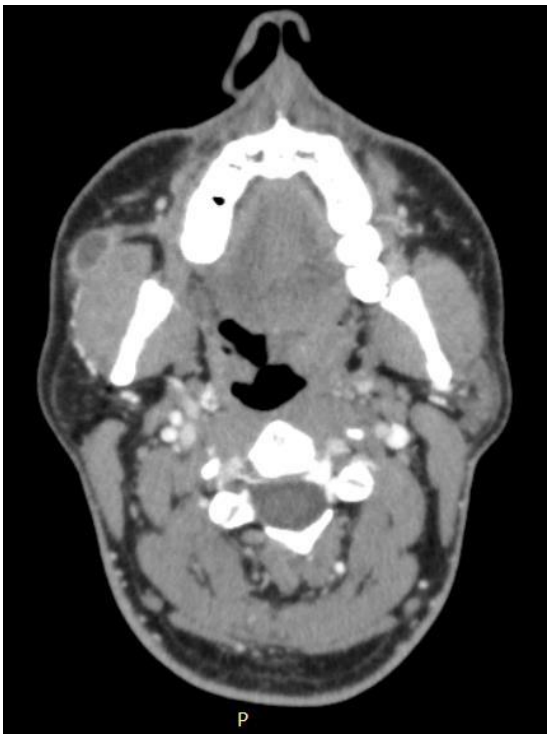

Figure 2: Contrast enhanced axial image shows abscess formation in ectopic parotid tissue, lateral to the masseter muscle. Draining orifice is also noted opening in to the oral cavity