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Extensive salivary gland choristoma of the pterygopalatine fossa

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A salivary gland choristoma is a congenital heterotopic rest of histologically normal salivary gland tissue. We describe the first case in the English language literature of an extensive salivary gland choristoma that was centered on the pterygopalatine fossa with extension to the infratemporal fossa, cavernous sinus, sphenoid sinus, and middle cranial fossa.

CASE REPORT

A 51-year-old man was referred to the department of neurology with a 5-year history of incoordination of his right hand and tremor. For the previous 3 years he had noticed gait problems, with a tendency to drag his right leg and slight imbalance. Multiple sclerosis was suspected, and an MRI scan was done. This revealed lesions consistent with multiple sclerosis, but an incidental mass was found. This extended from the left middle cranial fossa to the left pterygopalatine fossa and parapharyngeal area. He was therefore referred to the neuro-otology unit for further management. He had no specific otorhinolaryngologic symptoms or signs. His neurologic examination revealed mild dysarthria and mild bilateral rectus weakness with loss of smooth-pursuit eye movements and some unsustained nystagmus at extremes of gaze. His other cranial nerves were intact. He had no limb weakness or any sensory loss, but he was ataxic and hypertonic, more so on the right with an up-going plantar reflex on that side. In view of this unexpected MRI finding, further imaging was carried out. A CT brain scan demonstrated remodeling of bone around the mass with sclerosis of the walls of the middle cranial fossa. A further MRI with gadolinium and coronal imaging showed an extraaxial, heterogeneously enhancing soft tissue mass centered within the left pterygopalatine fossa (Fig 1). It extended laterally into the infratemporal fossa and anteriorly to lie immediately posterior to the left maxillary antrum. It extended medially into the left sphenoid sinus. There was distortion inferomedial to the nasopharynx. It extended superiorly into the left cavernous sinus and was in intimate relation to the inferior medial aspect of the left temporal lobe.

A transseptal sphenoid biopsy specimen was equivocal, and a further transantral biopsy specimen was inconclusive. In view of the uncertain nature of the lesion, he underwent surgical exploration through an extended lateral infratemporal fossa approach involving zygomatic arch displacement. The lateral pterygoid plate was excised to reveal tumor largely medial to this, and large biopsy specimens were taken for frozen-section analyses, which were also inconclusive. The tumor was therefore excised as completely as possible, and an estimated 85% to 90% clearance was achieved. Histology revealed mature tissue elements, the most prominent of which were mixed mucous and serous salivary glands with ducts that contained secretions. These elements were mixed with respiratory epithelium, hyaline cartilage, skeletal muscle, adipose tissue, fibrous tissue, and small nerves. There was no suggestion of malignancy, and this was considered to be a salivary gland choristoma.

DISCUSSION

Salivary gland tissue developing in inappropriate sites is referred to as heterotopia, ectopia, or choristoma. The embryogenesis of choristoma is often unclear. Willis[1] has suggested 3 possibilities: (1) abnormal persistence and development of vestigial structures; (2) dislocation of a portion of a definitive organ rudiment during mass movement; and (3) heteroplasia, which is abnormal differentiation of local tissues. Most salivary gland choristomas, as expected, occur in the head and neck, but they can also occur in remote sites. In the head and neck the paraparotid, middle ear, and lower neck are the most common locations.[2] Less commonly, the following

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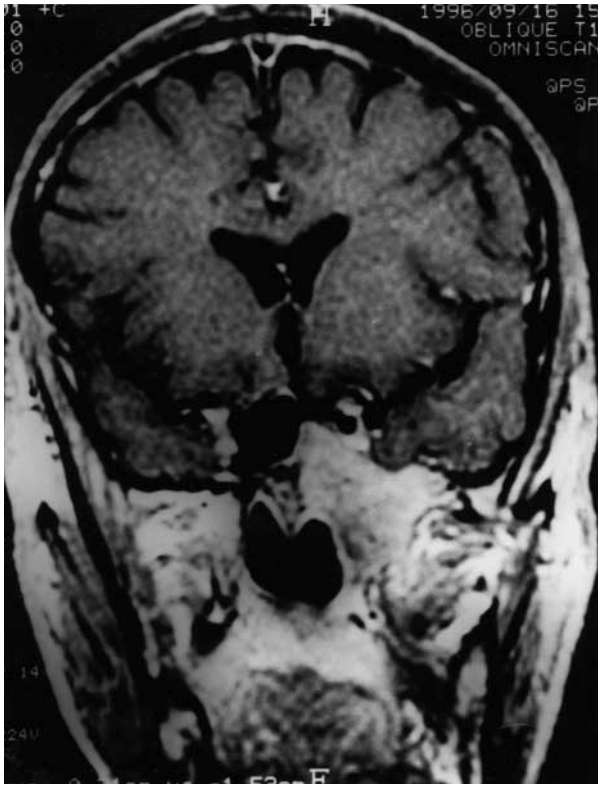

Fig 1. Coronal image on MRI scan showing superior, medial, lateral, and inferior extensions of the salivary gland choristoma.

sites are reported: the upper neck, mandible, external ear canal, mediastinum, cerebellopontine angle, pituitary gland, vulva, rectum, thyroglossal duct, thyroid gland, parathyroid capsule, tongue, tonsil, and tonsillar fossa.

In an audit of 20,000 cases of salivary gland lesions at the Armed Forces Institute of Pathology 110 cases of choristoma were identified, of which 64 had their sites documented.[3] Of these 64 cases, 30 (47%) were in lymph nodes of the neck, and 18 of these were in the paraparotid lymph nodes. Twenty cases (31%) were in the lower neck, 4 (6%) in the middle ear, 2 (3%) in the thyroid gland, 2 (3%) in the lacrimal gland, 1 (1.6%) in the upper neck, 1 (1.6%) in the stomach, 1 (1.6%) in the mandible, 1 (1.6%) in the mediastinum, 1 (1.6%) in the cerebellopontine angle, and 1 (1.6%) in a thyroglossal duct cyst.

Occasionally these ectopic rests may undergo neoplastic transformation, with mucoepidermoid carcinoma being the most commonly observed, followed by pleomorphic adenoma, Warthin's tumor, and other adenomas, in that order of frequency.[2] In the Armed Forces Institute of Pathology review, there were 13 neoplasms (12%), with 6 being mucoepidermoid carcinoma (46%), 3 pleomorphic adenoma (23%), 3 Warthin's adenoma (23%), and 1 unspecified adenoma (8%).[3]

Our patient was asymptomatic with no evidence of eustachian tube dysfunction. All the tissue elements of this choristoma are found in the eustachian tube, and in view of their close relation, the eustachian tube is the most likely site of developmental origin. Symptoms may occur at other sites. In the middle ear a unilateral conductive hearing loss is the most commonly described symptom. Many patients have multiple middle ear deformities, and a syndrome of conductive hearing loss, choristoma, ossicle anomalies, and abnormal facial nerve has been described.[4] Errant development of the first and second branchial arches before the fourth uterine month is the suggested embryogenesis.[3] In the lower neck salivary gland choristoma present as small draining sinuses that secrete a mucoid, creamy, or saliva-like fluid.[5] These sinuses are often bilateral and usually medial to the lower border of the sternomastoid muscle. The pathogenesis is unclear, but it has been proposed that overgrowth of the second branchial arch, which overlaps the second, third, and forth branchial clefts, is responsible.

Salivary gland choristomas are most commonly of mixed mucous and serous gland type, as was found in our case. Choristomas are nonneoplastic, hamartomatous-like lesions that do not require excision, unless recent growth is observed in adults or disproportionate growth occurs in childhood, but biopsy is needed for diagnosis. In our case about 90% of the lesion was excised, and indeed a complete resection was considered inappropriate in the absence of a histologic diagnosis and with tumour extension into the cavernous sinus. The intraoperative frozen section of the biopsy specimen also showed only mature tissue elements with no evidence of malignancy, and a subtotal resection was therefore undertaken. With the increasing use of diagnostic imaging, these rare developmental aberrations will increasingly be discovered incidentally and continue to be enigmatic radiologic lesions. Tissue biopsy is mandatory, but as in our case, may not be diagnostic. Unfortunately, formal excision is sometimes the only way to obtain a precise diagnosis.

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