

The Cartilaginous Subcutis: Choristoma and Heterotopy

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Choristoma indicates the Greek "X^{quaros}" implying separated. It describes an accumulation of normal histological tissue of an organ or a body constituent distant from the typical location. Heterotopic cartilage located beneath the skin, in the vicinity of head and neck represents a cartilaginous choristoma. Choristomas are usually distinctive and elucidate the heterotopy of thyroid gland, bone, glial tissue, salivary gland etc. Choristomas are inherently diverse and benign and grow imperceptibly. Excision is rendered for cosmetic purposes and for malignant transformation. External cartilaginous choristomas in the head and neck are now designated as chondrocutaneous branchial remnants or accessory pinna/tragus, with the assumption that the indicated lesions are of the branchial arch residue. Analogous heterotopic cartilage in the oral cavity, tonsils and pharynx is named Cartilaginous Choristoma. Notwithstanding, the biological expression and histopathology of the lesions with heterotopic cartilage covered with skin or mucosa are identical. The masses are extremely slow growing. Patients clinically present when the tissue aggregates become prominent due to gradual evolution. Cartilage is observed on palpation and gross exam. Concomitant malformations and a conclusive family history are not exhibited.

Salivary gland choristoma
Central
Gingival
Cartilaginous choristoma
Osseous choristoma
Lingual thyroid choristoma
Lingual sebaceous choristoma
Glial choristoma
Gastric mucosal choristoma
Cystic
Solid

Figure 1: Types of Choristomas [1].

Histomorphology of chondrocutaneous branchial remnants or accessory pinna/tragus or cartilaginous or chondroid choristoma is comparable and comprehensive, with subcutaneous mature cartilage, a circumferential connective tissue with unremarkable superficial stratified squamous epithelium. Anatomically interchangeable lesions and locations evoke comparable autonyms, in contrast to the taxonomic criterion. Chondrocartilagenous branchial remnants are extrinsic head and neck deformities and the existing phrase (1997) for heterotopic, subcutaneous cervical cartilage. Intrinsic malformations are alluded to as cartilaginous or chondroid choristomas The elemental histopathology is that of the existence of cartilage in the subcutaneous tissue or lower dermis and a superimposition with the upper dermis and the epidermal stratified squamous epithelium. Accessory pinna/tragus is a term which is used preferably for pre-auricular disfigurements. The cartilage fragments represent heterotopy, the circumscribed connective tissue and the superimposed epithelium are compatible and inherent to the site of the lesion.

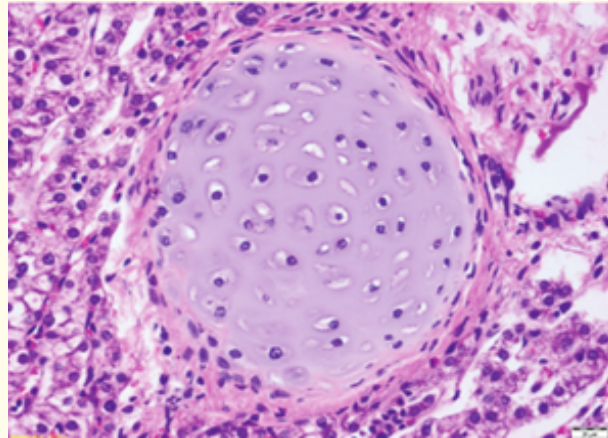


Figure 2: Cartilaginous Choristoma with healthy cartilage [2].

Chondrocutaneous as a terminology is indicative of cartilage and skin exhibiting as a single entity and being of the similar ancestry. The epidermis derives from the ectoderm while the neural crest develops the dermis for the face, neck, bone and craniofacial cartilage. Pathogenetic principle of a cartilaginous choristoma is

- 1) The Atlan., *et al.* "left behind theory" of cervical chondrocutaneous branchial remnants is applicable and it postulates that the cellular remains of the first and second pharyngeal arches which supplement the auricular hill 1 to 6 and trail in the neck during embryonic migration. Hence heterotopic cervical cartilage is the outcome of the cartilaginous differentiation. The 1st and 2nd pharyngeal arches configure the ear, tongue, maxilla, mandible, palatine tonsil and pharynx. A perfect anatomic and embryologic proposition must analyze the location, incidence and natural history of the lesion and their counterparts. Atlan's assumption is partial and incomplete as the cervical lesions require differentiation from the identical, morphological defects. The terminologies "displaced laterally, medially or overshoot" and instructing the embryonic cellular migration should replace the ambiguous expression "behind". Hence extrinsic and intrinsic chondrocutaneous branchial remnants are designated cartilaginous/chondroid choristomas. However distinct terminologies and locations formally exhibit similar histology. Choristoma indicates the non-specific existence of heterotopic tissue.
- 2) With cartilaginous heterotopy, the cartilage is the dynamic prerequisite while the connective tissue circumscription with superimposed stratified squamous epithelium is the inactive component. This perspective can conform with the fact that cartilaginous heterotopy in head and neck cannot be distinguished on a histological basis from the chondrocutaneous branchial remnant and the accessory/pinna or tragus except the oro-pharyngeal mucosa which envelopes the lesion where appropriate. Thus heterotopic cartilage and cartilaginous or osseous choristoma presenting in distant locations and with different classifications are essentially identical. Cartilaginous choristoma, besides the subcutaneous tissue, is encountered in the head and neck, nasopharynx, eye, tongue, middle ear etc.
- 3) The aberrant differentiation or proliferation of immature, pluri-potent cell rests in the head and neck is necessary for the cartilaginous choristoma to progress. Thus the "left behind" premise is not a comprehensive assumption. The existence of supernumerary nipples in the sole of the feet, thigh, scapula etc. can similar to this concept. This derivation interprets the existence of cartilaginous choristomas in accessory situations. The anomalous differentiation and resulting heterotopy exclusively arise from immature pluri-

potent cells. However the catalyst engendering the differentiation and heterotopy is obscure and debatable. Most instances are unilateral and lack a specific family history. Non-familial or incidental cases can interpret the presence of osseous choristomas in corresponding zones based on the fact that the mesodermal bone dislocates the cartilage even in the embryonal stage. The inherent choristomas are detected at an older age. The extraneous deformities are congenital and the evolution and loci constitute the discordance. Subcutaneous cartilaginous choristoma are extrinsic and superficial thus clinically detectable. Internal, asymptomatic cartilaginous choristomas are discovered when the lesion has evolved and is of massive proportions. The cartilaginous choristomas are indolent and slow growing. Characteristically, the non-neoplastic, inactive lesions elucidate the cartilaginous heterotopy which is dependent upon the critical growth factors (The growth factors are exhausted at the aberrant points of growth/evolution). The actively expeditious, benign lesions are attributed to the microenvironment which is conducive to growth. Malignant transformation has not been detected. Anomalous differentiation occurs primarily in the head and neck as the three embryonic cell layers and their derivatives are contiguous at this site. Unilateral, extrinsic lesions are found to coexist with the Goldenhar-Gorlin syndrome or the oculo-auriculo-vertebral (OAV) syndrome (hemifacial microsomia, Facio-auriculo- vertebral spectrum). Each syndrome has an exclusive range of severity that is directed by degree of persistence in the patient. Certain vital foetuses attain maturity with unilateral facial and vertebral mal-conformations. Severe bilateral monstrosities are incompatible with survival. Bilateral disfigurements which persist generally predispose to benign malformations. Subcutaneous cartilage is also delineated in the hamartoma and cartilaginous metaplasia. Hamartoma is a benign architecture and aggregate of disorganized native tissue. Cartilaginous metaplasia evolves subsequent to trauma or neoplastic transformation. Biologic behaviour excludes both hamartoma and cartilaginous metaplasia from the congenital category. Chondro-cartilaginous choristomas have a predilection for the male sex. The entire range of Choristomas delineates an identical pathogenesis and histology. Pluripotent cells usually differentiate in a conformable manner based on the procurement of the growth factors. The location of the lesion decides the proportion of growth, which could be considerable. Bilateral symmetrical external lesions are either innocuous, solitary aberrations or fatal when occurring in collaboration with other congenital/life threatening deformities.

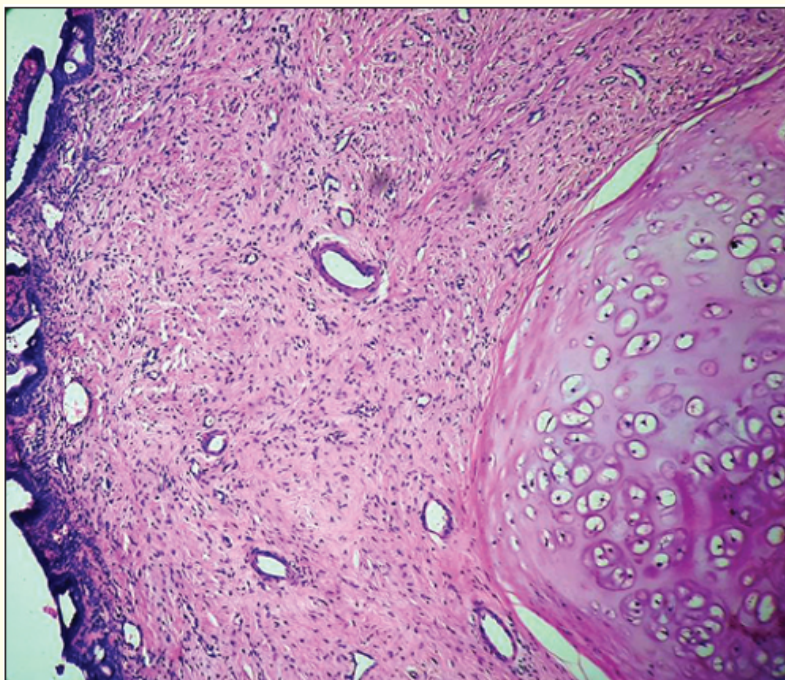


Figure 3: *Conjunctival Choristoma with normal cartilage [3].*

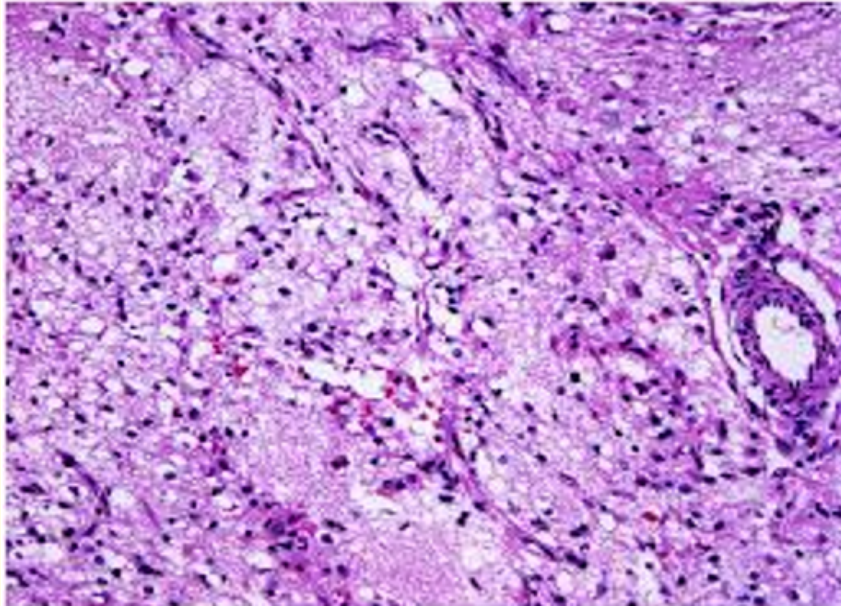


Figure 4: Neuroglial choristoma with neurons and neuroglia [4].

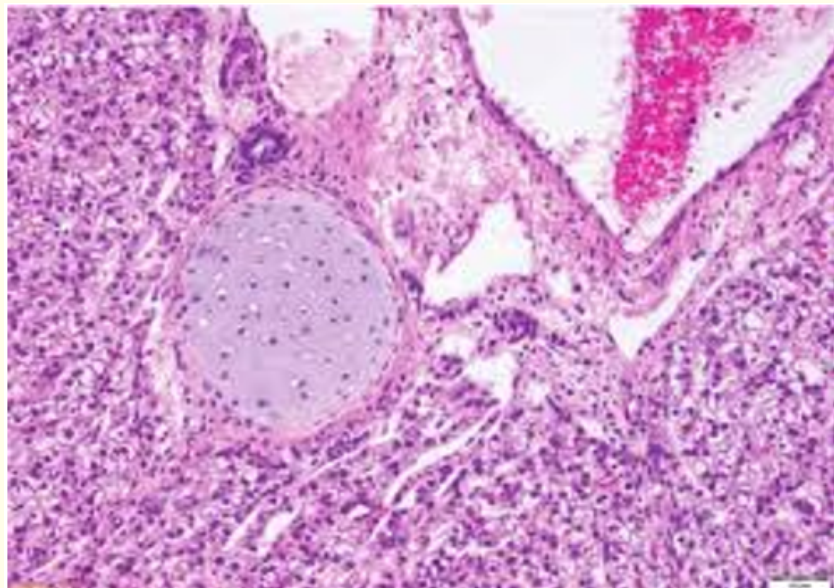


Figure 5: Cartilaginous Choristoma [2].

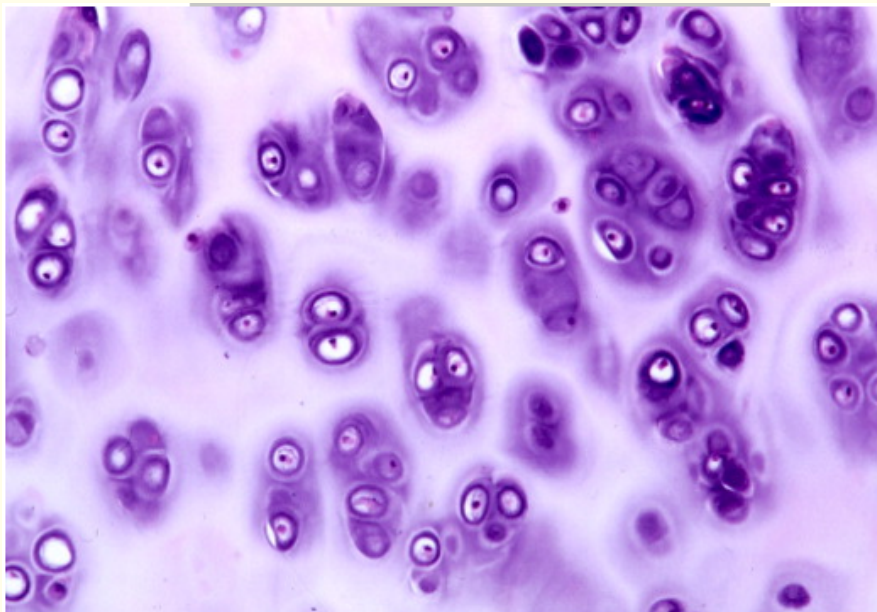


Figure 6: Cartilaginous Choristoma-aspiration cytology [5].

Soft tissues of the head and neck may manifest rare osseous choristomas [6]. Microscopy demonstrates calcified nodules enclosed by fibrous and adipose tissue. The calcified nodule consists of cartilage with the typical lacunae containing chondrocytes. Mature cartilage is central while the immature chondroid foci are located at the periphery. The precise aetiology and pathogenesis of the specific cartilaginous arrangement in peculiar sites, particularly in the oral cavity, is obscure. The propounded theories of ectopic chondroma are i) multipotent cells surviving in an undifferentiated mesenchyme. ii) the presence of ectopic embryonic cell rests iii) the existence of metaplasia. The benign aberrations necessitate a surgical excision. Ruckert [7] presented the first anomaly in a tonsil of a neonate, the component of a persisting branchial arch and the foremost intraoral cartilaginous choristoma [8,9].

Bibliography

1. Image 1 Courtesy: Dental Research Journal.
2. Image 2 and Image 5 Courtesy: Research gate.
3. Image 3 Courtesy Indian Journal of Ophthalmology.
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