

**H&N Neuroendocrine Neoplasms:
Classification and Diagnostic
Considerations**

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Broad Classification:

A. Epithelial:

- Neuroendocrine carcinoma (Typical and atypical carcinoid and PDC)
- Medullary thyroid carcinoma
- Parathyroid tumors

B. Non-Epithelial:

- Neuroblastoma
- Paraganglioma

Neuroendocrine Carcinoma

- Incidence:
 - <1% of H&N tumors

- Etiology:
 - Unknown!!
 - Radiation
 - Smoking

Histogenesis

- Neuro-endocrine cell
- Undifferentiated / basal / stem cell

Locations:

- Larynx
- Salivary glands (major and minor)
- Sino-nasal region
- Middle ear and endolymphatic sac

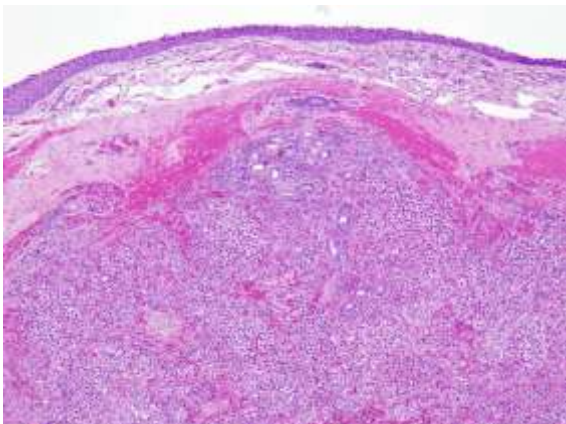
Larynx

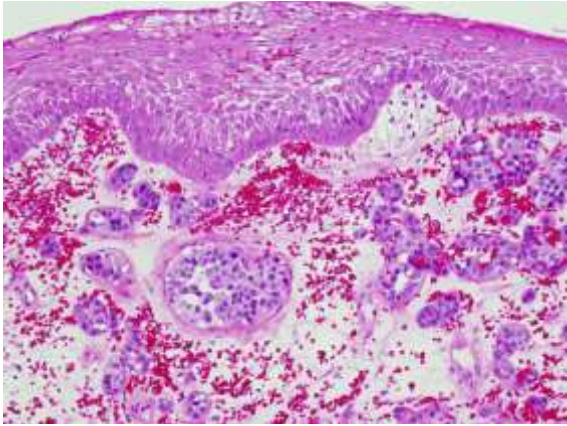


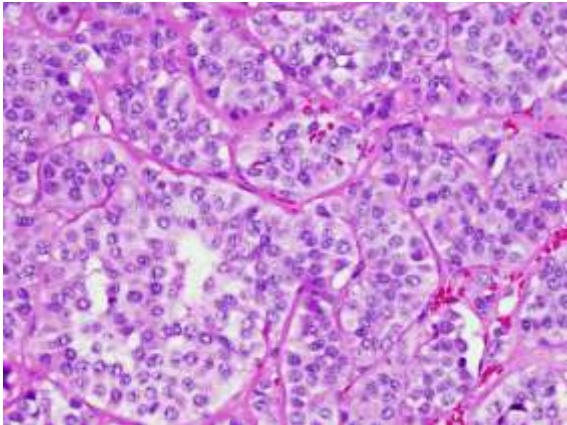
Histologic Classification:

- Well differentiated (typical carcinoid)
- Moderately differentiated (atypical carcinoid)
- Poorly differentiated (small, large cell)

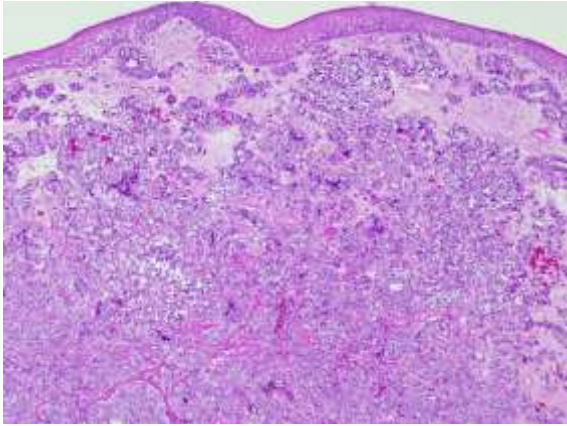
Typical Carcinoid

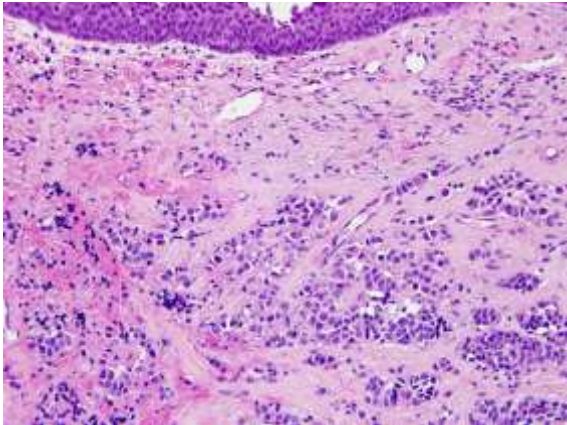


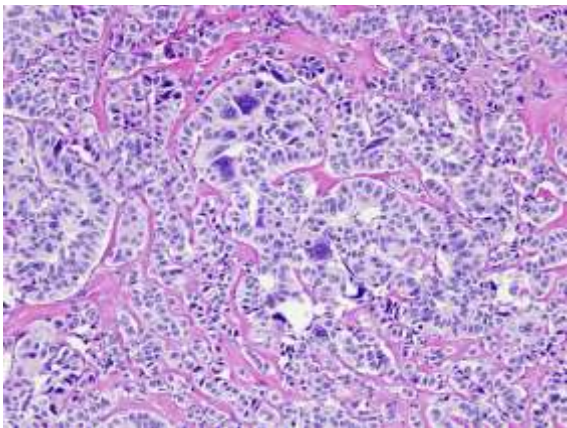


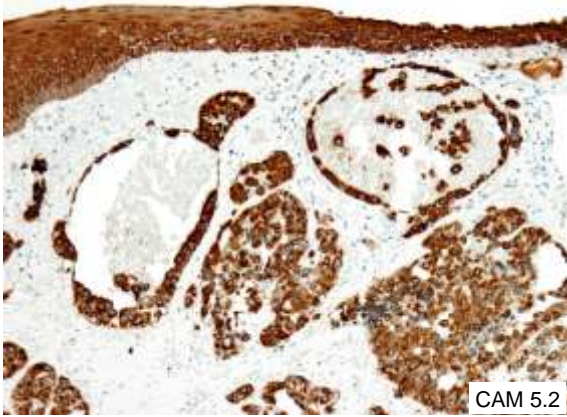


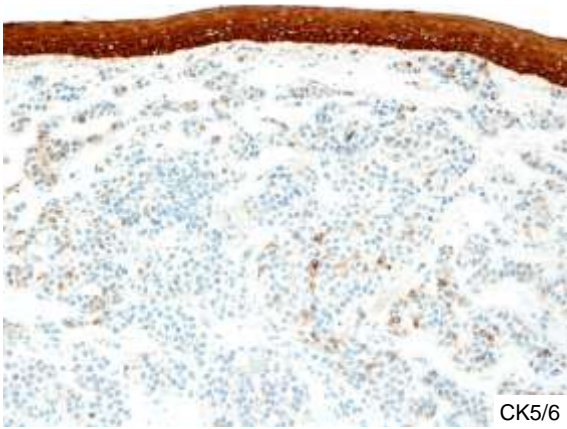
Atypical Carcinoid

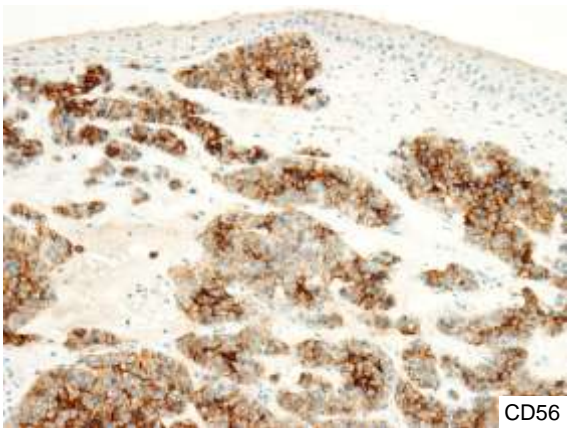




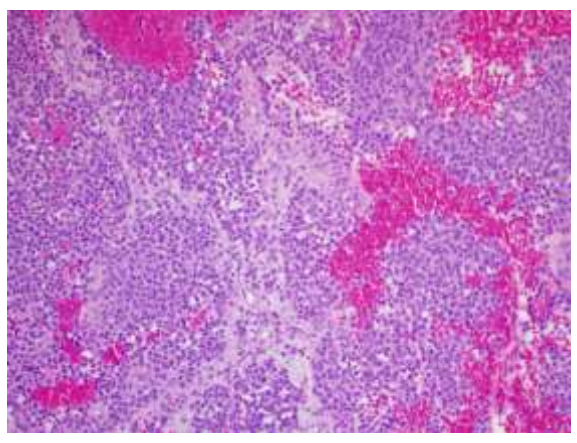


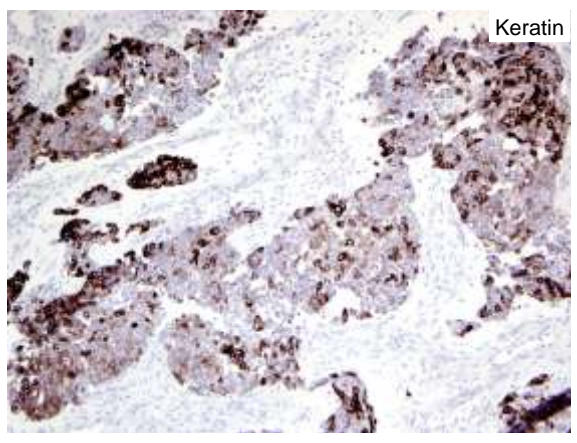


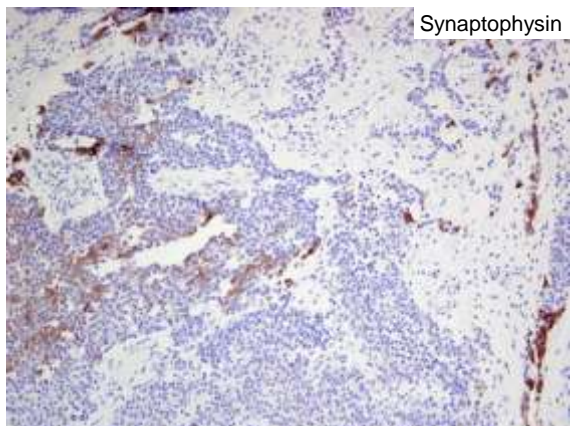




Poorly Differentiated







Immuno Markers:

Epithelial:

	<u>Differentiated</u>	<u>Undifferentiated</u>
• Pan cytokeratin (AE1/AE3)	++	+
• EMA	+	+/-
• CK 5/6	++	+

Neuroendocrine:

	<u>Differentiated</u>	<u>Undifferentiated</u>
• Chromogranin	+++	++
• Synaptophysin	+++	++
• Calcitonin	++	-
• CD 56	++	+

Management of Laryngeal NET:

- Differentiated: surgery
- Poorly differentiated: chemo + XRT

Clinical Characteristics of MD-NEC at MD Anderson

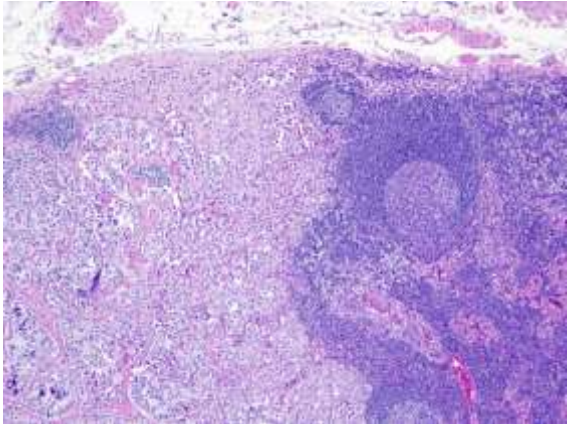
Case	Age ¹ Sex	Smoking history	T Stage	N Stage	M Stage	Initial Treatment	Recurrent	Status	Stressor Present	OS Months	OS Years	Survival 3 Years	Survival 5 Years
1	41M	No	T3	N3	1	C	No	DOC	NA	14.0	No	No	
2	44F	Yes	T1	N2	0	SFT	Yes	DOC	55.1	58.4	Yes	Yes	
3	26F	Yes	T1	N2	0	S	Yes	DOC	95.1	83.0	Yes	Yes	
4	52F	Yes	T1	N2	0	C = RHT	No	REC	49.8	33.0	Yes	NA	
5	39MF	Yes	T1	N2	0	C = RHT	Yes	REC	34.6	47.3	Yes	NA	
6	41F	No	T2	N2	0	S = RHT	No	REC	41.8	49.9	Yes	NA	
7	54M	Yes	T2	N2	0	C = SFT	Yes	DOC	30.8	48.0	Yes	Yes	
8	72MF	Yes	T1	N2	0	C = RHT	No	DWD	18.8	30.0	Yes	Yes	
9	44MF	Yes	T2	N2	0	SFT	Yes	DOC	3.0	15.7	No	NA	
10	42F	No	T2	N2	0	S	Yes	DOC	42.7	48.1	Yes	Yes	
11	51F	Yes	T2	N2	0	S = RHT	Yes	REC	22.3	31.3	Yes	NA	

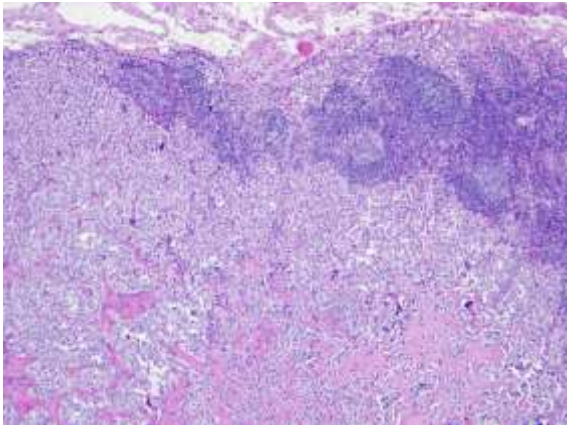
Differential Diagnosis:

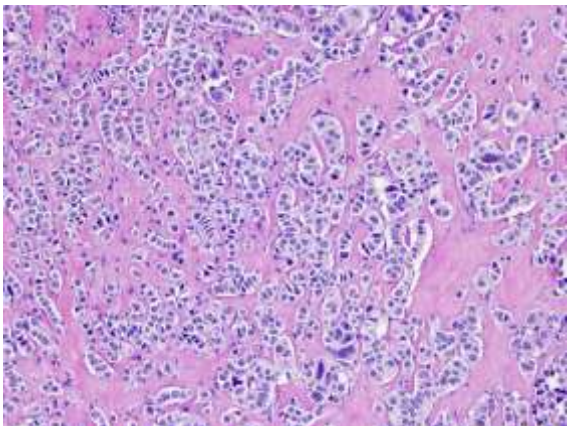
- A. Differentiated:
 - Medullary thyroid carcinoma
 - Paraganglioma
 - Basaloid salivary adenocarcinoma
 - RCC
- B. Undifferentiated:
 - Small round cell tumors
 - Undifferentiated carcinoma
 - Basaloid squamous carcinoma
 - Merkel Cell Carcinoma

Case Presentation:

- 42 year old female
- With left neck mass (4.0 cm)
- Serum calcitonin: high
- Excision of neck mass





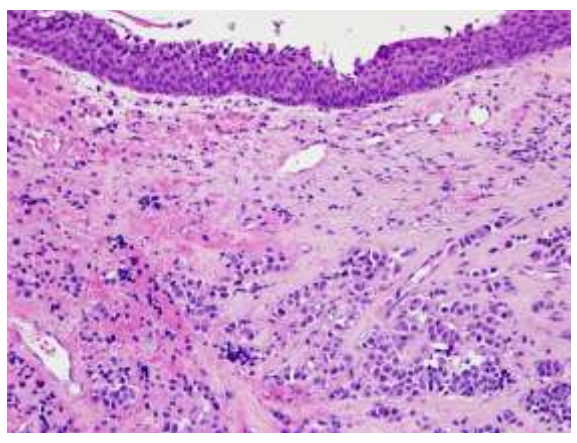


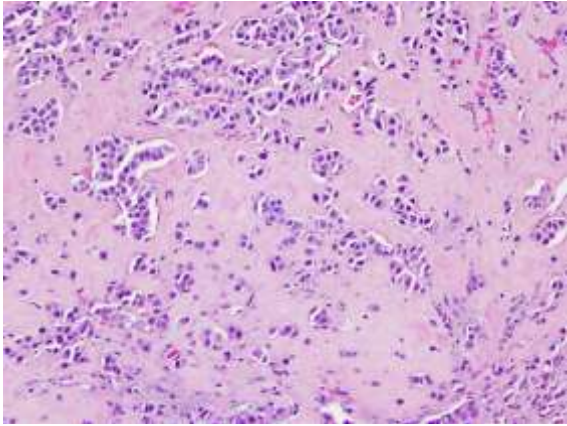
Diagnosis:

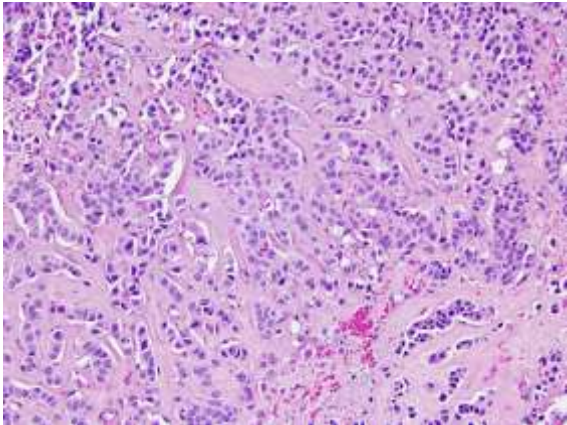
- Neuroendocrine carcinoma favor metastatic MTC

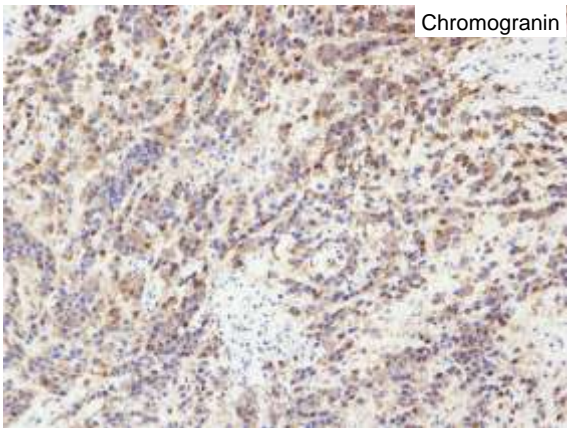
Follow-up:

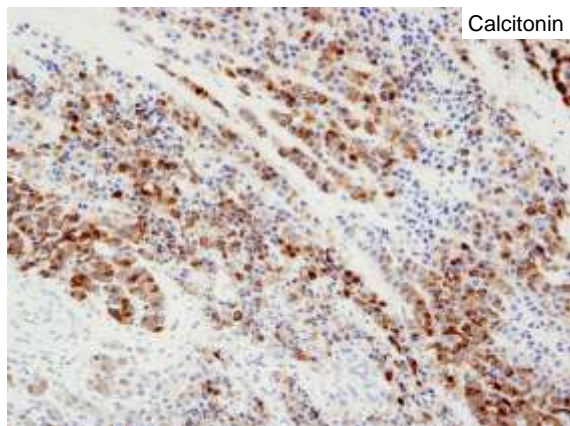
- Thyroid examination normal
- Hx of polypoid laryngeal nodule 5 years











Comparative IHC of Laryngeal Primary and Lymph Node Metastasis:

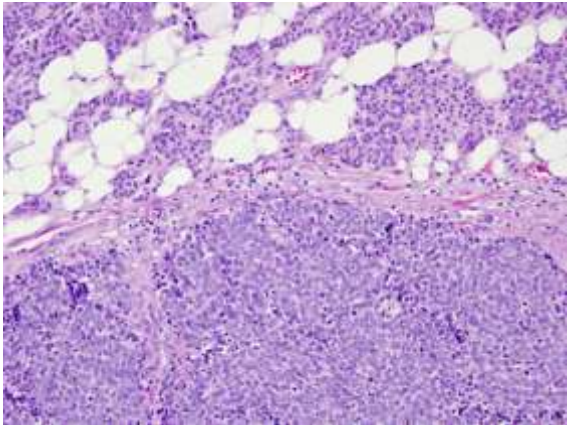
Marker	Primary	L-N
Somatostatin	-	-
Serotonin	++	+
ACTH	-	-
NSE	++	++
Chromogranin	+++	++
CEA	++	+
Calcitonin	++	++

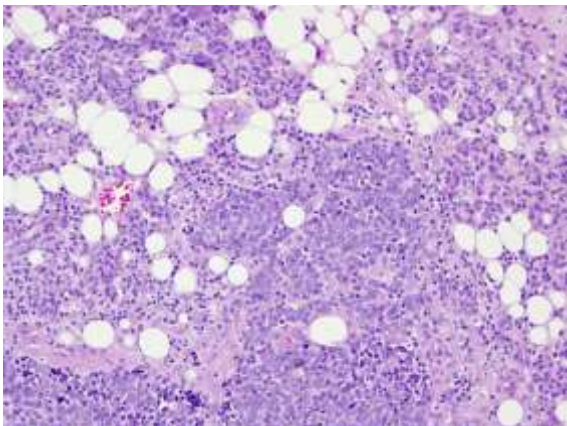
Diagnosis:

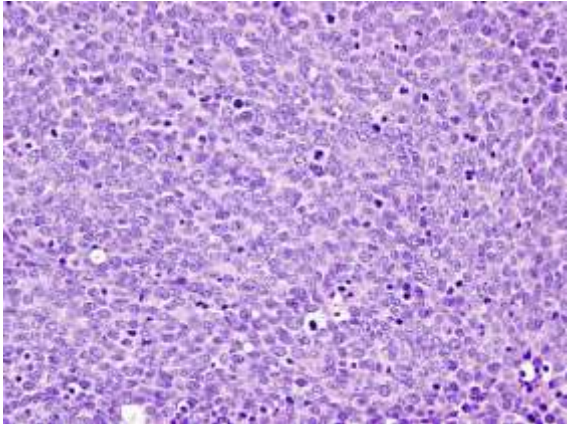
- MTC-like carcinoid tumor of the Larynx

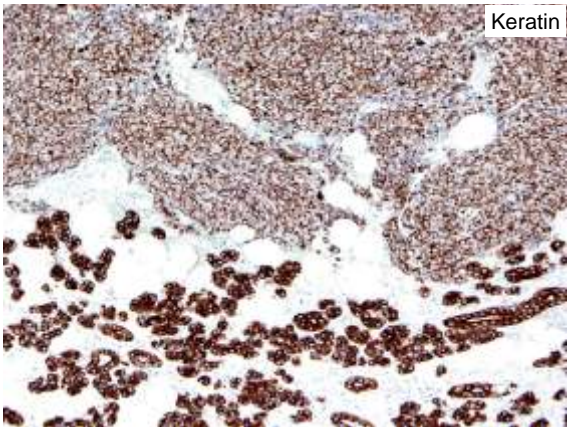
Salivary Glands

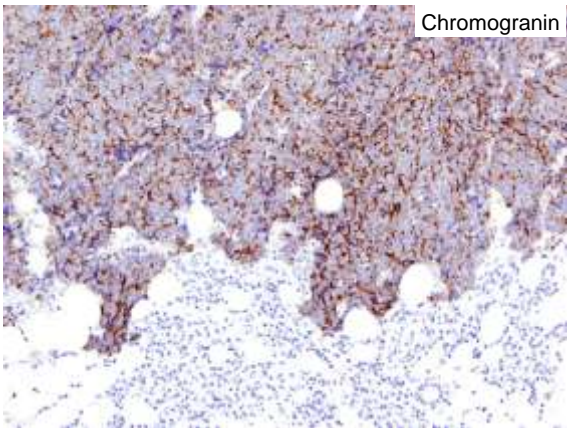
- Primary: rare
- Differentiated tumors: rare
- Secondary: more common











Differential Diagnosis:

- Merkel cell Ca
- Melanoma
- Metastatic, NEC
- RMS

Managment

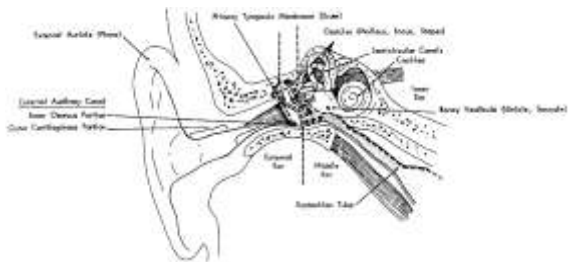
Primary:

- Surgery ±
- Chemotherapy/XRT

Secondary:

- Chemotherapy/XRT

Middle Ear and Mastoid Neoplasms:



Tumor types:

- Adenomatous tumors
- Paraganglioma
- Neuromas/ acoustic
- Meningioma (intratemporal)
- Schwannoma, Labyrinthine
- Inverted papilloma
- Hemangiopericytoma
- Fibromatosis
- Metastasis

**Middle Ear
Adenomatous Tumors**

Histogenesis:

- Surface epithelium
- Stem / basal progenitor

(no evidence for neuroendocrine cells)

Clinical Features:

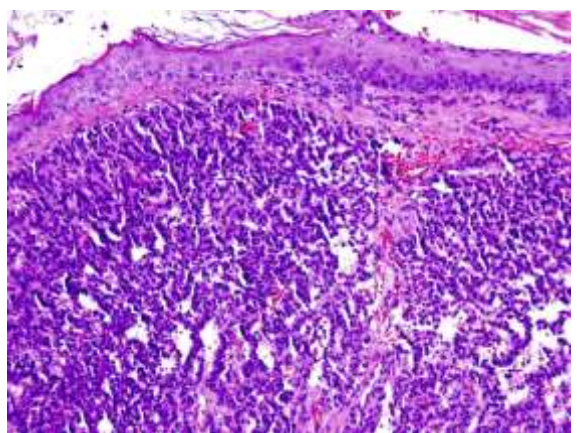
- 2nd to ninth decades
- No sex predilection

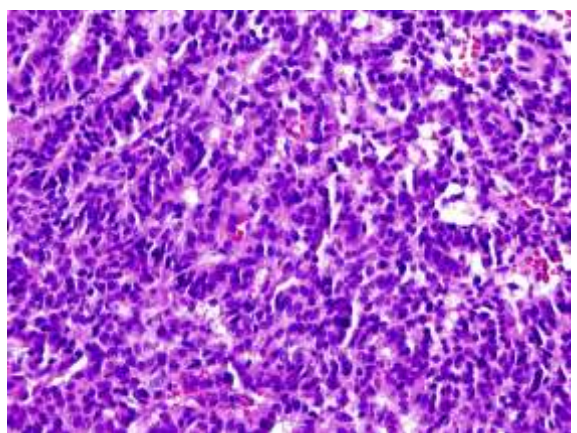
Symptoms:

- Decreased hearing
- Ear fullness
- Tinnitus

Signs:

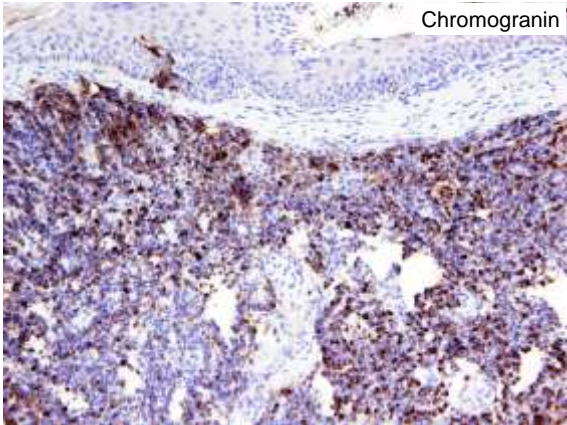
- Mass, middle ear cleft
- Discharge

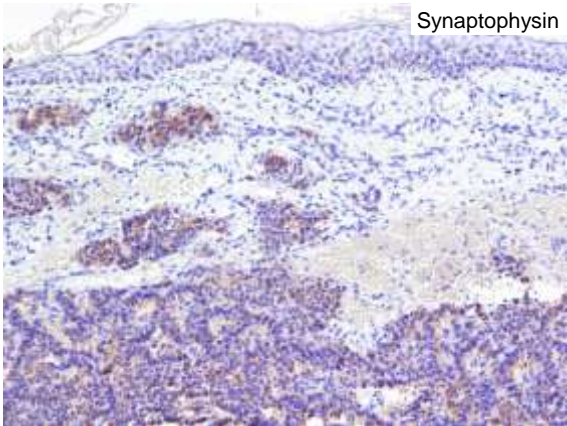




Markers:

- Mucin: negative
- Endocrine markers: positive





Differential Diagnosis:

- Ceruminous carcinoma: lateral 2/3 of the external ear canal
- Salivary choristoma

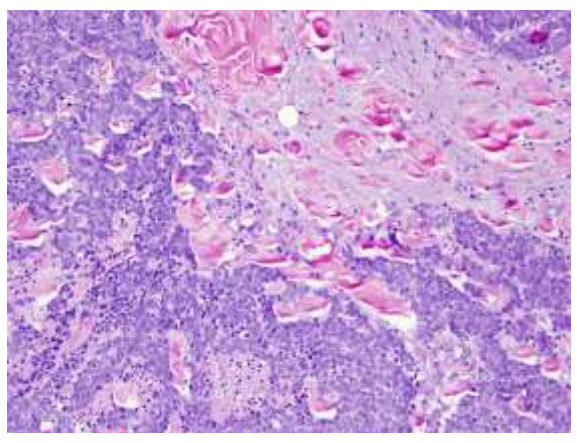
Behavior:

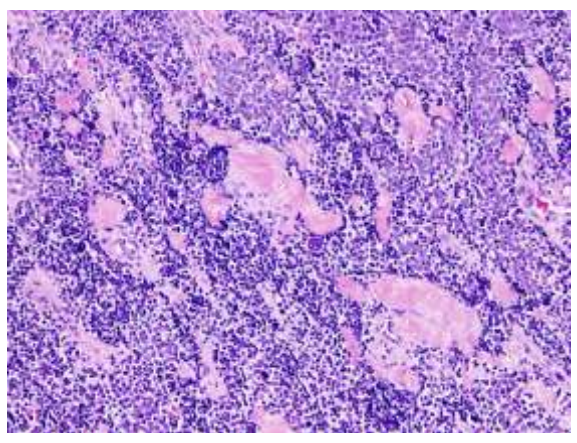
- Non invasive: adenoma / carcinoid
- Bone destruction: adenocarcinoma/NEC

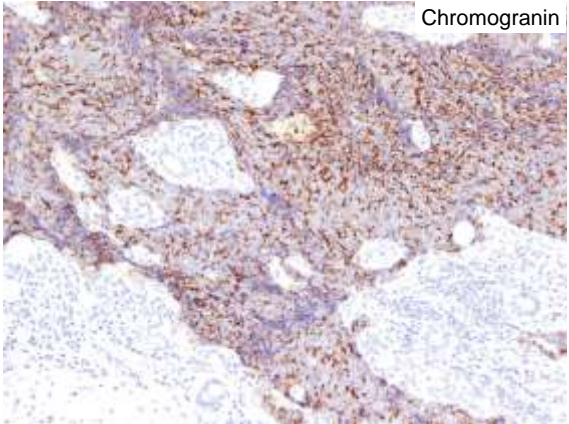
Merkel Cell Carcinoma

Origin:

- Metastasis: common
- Primary: ? rare







Management:

- Chemo + XRT

Conclusions:

H&N NET

- Rare
- Heterogeneous
- Exclusion of mimics (D.D)

Thank you
