

Case #1 A 50 y/o male presents with a nasal mass, which is excised. Representative images of the lesion are shown. Immunohistochemistry of the lesions shows positivity for CD34. Based on these findings, what is the most likely diagnosis?

C. Solitary fibrous tumor

Answer: C. **Solitary Fibrous Tumor (SFT)**. SFTs were originally thought to be mesothelial in origin, but are more likely to belong to the category of myofibroblastic tumors. Histologically, they have a bland uniform “patternless pattern” of bands of collagen with foci of hemangiopericytic growth (storiform pattern with staghorn vessels). SFT and hemangiopericytomas are thought by many to represent two ends of a spectrum (both are CD34 positive). SFTs are CD34 + and SMA =, which helps differentiate them from fibromatosis (CD34 =, SMA +). (Sternberg, 4th Ed., p. 163-64)

Case #2 A mass was found in adjacent to the parotid gland. An excision was performed and representative sections are shown. Based on the findings, what is the best diagnosis?

A. Neurofibromatosis type 1

Answer: A. This case illustrates a **Plexiform Neurofibroma** which is almost always associated with neurofibromatosis type 1 (NF1). Morphologically the lesion will have an appearance of a neural lesion (Schwann cells inside a nerve sheath) with the additional finding of tortuous structures. This is somewhat similar to a “bag of worms” appearance characteristic of other lesions.

Case #3 A 40 y/o male presents with a parotid mass. Based on the histologic images, the best diagnosis is:

B. Pleomorphic adenoma

Answer: B. **Pleomorphic Adenoma (PA)** PAs are the most common neoplasm of the salivary gland (54-76% of all tumors) and are benign. They are composed of epithelial glands, myoepithelial cells, and mesenchymal stroma. One of the most important things to remember is the large amount of morphologic heterogeneity that can occur with these tumors. Any one of the components may predominate, which may confuse the diagnosis. Many people believe that a myoepithelioma represents an extreme spectrum in histologic variability of pleomorphic adenomas. If carcinoma is present within the PA, then the term *carcinoma ex* is used. (Thompson, p. 295-300)

Case #4 A 40 y/o female presents with a nasal obstruction. A mass is removed from the lateral wall, and representative histologic images are shown. Based on the findings, the best diagnosis is:

- C. Cylindrical cell papilloma

Answer: C. **Oncocytic Papilloma / Cylindrical cell Schneiderian Papilloma (CCP)** CCPs have an exophytic papillary architecture with multiple cell layers of eosinophilic/oncocytic cells with well defined cell borders. Intraepithelial microcysts are common in these tumors (present in this case). Seromucus glands can also be found in the submucosa, which is not seen in inverted papillomas (not present in this case). It is also common to find cilia present focally on the surface. CCPs have an excellent prognosis, rarely have malignant transformation, and have equal gender distribution. Endophytic/inverted and exophytic Schneiderian papillomas are more common in men (4:1), and the endophytic papilloma will have malignant transformation in approximately 10% of cases (Thompson, p. 124-131)

Case #5 A 30 y/o male is found to have a buccal lesion. An excision was performed and representative images are shown. The best diagnosis is:

- B. Irritation fibroma

Answer: B. **Irritation / Traumatic Fibroma** This is a general term of a lesion that often present on the buccal mucosa of the oral cavity. Its name is descriptive of the etiology and appearance. The lesion usually has a polypoid (dome shaped) appearance with dermal fibrosis and is reactive (not neoplastic) in nature. The mucosal surface can be variably thickened and keratotic.

Case #6 A 50 y/o female presents with a parotid mass. Based on representative images of this lesion shown on the website, which of the following is the best diagnosis?

- D. Acinic cell carcinoma

Answer: D. **Acinic Cell Carcinoma.** Acinic cell carcinoma (AcCC) can have many different morphology patterns (solid, microcystic, papillary-cystic, & follicular) with different cells types (non-specific glandular, intercalated ductal, vacuolated, and clear). This can cause a great diversity in appearances under the microscope. Fortunately, most cases have a serous appearance, and the zymogen granules can be highlighted with a PAS stain. It is important to keep AcCC in the back of one's mind where confronted with a difficult salivary gland lesion. (Thompson, p. 332-339)

Question #1 All of the following are true with regards to cholesteatomas, EXCEPT:

- E. Is a neoplastic lesion

Answer: E. Cholesteatomas are not true neoplasms but can act in a destructive and aggressive fashion. This destructive nature is mediated primarily by collagenase, which is osteodestructive. These lesions occur in the middle ear, are more common in men, and often present in the third or fourth decade of life. (Sternberg, 4th Ed., p. 1046-47)

Question #2 All of the following are true about acinic cell carcinoma EXCEPT:

- A. Most common malignant salivary gland tumor in children

Answer: A. Acinic cell carcinoma is the second most common malignant salivary gland tumor in children. Mucoepidermoid carcinoma is the most common malignant salivary gland tumor in children. (Thompson 332-339)

Question #3 Which of the following subtypes of nasopharyngeal carcinoma has the best prognosis?

- A. Non-keratinizing nasopharyngeal carcinoma

Answer: A. Non-keratinizing (undifferentiated) nasopharyngeal carcinoma is the least differentiated of the nasopharyngeal carcinomas, but has a better prognosis because it is very sensitive to radiation therapy.

Question #4 A 41 y/o male presents with a nasal obstruction and septal perforation. Multiple biopsies are taken which shows predominately necrosis. Serologic studies are positive for a high c-ANCA titer. Based on these findings, what is the most likely diagnosis?

- D. Wegner's granulomatosis

Answer: D. Wegner's granulomatosis is a vasculitis that may result in ischemic necrosis. Pulmonary and renal involvement is most common, but there are frequent biopsies from the nasal cavity because it can easily cause symptoms and it is very assessable to biopsy. It is often difficult to identify the vasculitis component in a small biopsy. The positive c-ANCA is very suggestive of Wegner's granulomatosis and should be mentioned in the differential diagnosis of unexplained necrotic nasal lesions. With that being said, NK/T cell lymphoma can sneak up with a similar clinical presentation (minus the positive c-ANCA titer). NK/T cell lymphoma is usually angiocentric and virtually 100% positive for EBER (EBV in situ hybridization). (Thompson, p. 108-111, 191-194)

References:

Sternberg's Diagnostic Surgical Pathology. Mills, SE, *et al.* Fourth Edition. 2004.

Head and Neck Pathology. Thompson, LD, *et al.* First Edition. 2006.

Notes for question set:¹

¹ PathMD strives for the highest quality and accuracy. However, the *PathMD: Board Review Letter* is for review purposes and not meant for clinical decision making. It should not be used in place of review of primary reference texts and the current medical literature. If inaccuracies are identified, please notify us so that a correction may be published. (info@PathMD.com)