Malignant tumours

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Mucoepidermoid carcinoma Mucoepidermoid carcinomas are malignancies consisting of mucinous, intermediate and squamoid tumour cells in vari - able proportions. They are the most common salivary gland malignancies in children and young adults, with a peak inci - dence in the second decade of life. They are known to occur following radiation or chemotherapy in childhood. T hey occur - in both major and minor salivary glands, with the parotid being the most common site involved. The y generally present as soft to firm, painless masses with a gradual increase in size. The tumours are classified as low , intermediate or high grade based on histology . High-grade mucoepidermoid carcinomas tend to be locally aggressive with bone and/or skin involvement and nodal metastases. Distant metastases are seen mainly to the lungs. Complete surgical excision with wide margins is advo - - cated for mucoepidermoid carcinoma. Appropriate adjuvant radiotherapy is the treatment of choice for intermediate- to high-grade mucoe pidermoid carcinomas. On gross examination, the tumours are circumscribed or infiltrative and partially cystic. On histopathology , mucoepi - dermoid carcinomas have squamoid, mucin-producing and intermediate cells in variable proportions. There can be a - cystic and solid growth pattern. Low-grade mucoepider moid carcinomas are generally cystic, well circumscribed and rich in mucous cells. Intermediate-grade tumours are less circum - scribed and more solid, usually with a predominant intermedi - - ate cell component. High-grade mucoepidermoid carcinomas are usually solid, infiltrative and show nuclear atypia, mito - sis, necrosis, perineural invasion and lymphovascular emboli. Demonstra tion of at least focal intracellular mucin is essential for the diagnosis of high-grade mucoepidermoid carcinoma. Summary box 54.5 Mucoepidermoid carcinoma /uni25CF /uni25CF /uni25CF Adenoid cystic carcinoma - Adenoid cystic carcinoma is a slow-growing malignancy composed of both epithelial and myoepithelial cells and

Most common salivary gland malignancy Can occur in minor and major salivary glands Most common site: parotid

It has varying outcomes with good 5-year control but poor 10-year survival owing to the higher incidence of delayed distant metastases. They occur mainly in the fifth to sixth decades, with a slight female preponderance (1.5:1). Most of these malignancies occur in the major salivary glands, but they can also be seen in minor glands in the oral cavity , paranasal sinuses, tracheobronchial tree, etc. Most patients will present with slow-growing masses, with the presence of numbness, paraesthesia or pain. Facial and other neural palsies may be present depending on the site of the tumour. Nodal metastases are seen with high-grade lesions and asymptomatic distant metastases, especially lung metastases, are a frequent presentation. In addition, bone, liver and brain metastases are also seen. Radical surgical excision with or without adjuvant radiotherapy is the treatment of choice. Single-modality radio therapy is associated with inferior control outcomes. There is an emerging role of proton ion and carbon ion therapy , espe cially in unresectable/metastatic disease. Factors influencing surviv al include the tumour site, stage, nodal

disease, presence of perineural spread and grade of tumour. Grossly , adenoid cystic carcinoma presents as a poorly circumscribed, fir m, grey-white and solid mass. On histo pathology , it is an unencapsulated, infiltrative biphasic neoplasm with variable proportions of epithelial and myoepithelial cells and shows cribriform tubular and solid patterns. The cells show small, angulated, hyperchromatic nuclei with scant cytoplasm. The cribriform pattern is characterised by neoplastic cells arranged around small, sharply punched out cylindromatous spaces containing basophilic matrix. The tubular pattern shows bilayered tubules with a true lumen. The solid pattern is less common and shows sheets and nests of tumour cells without lumen formation. Perineural invasion is widely seen in adenoid cystic carcinoma. High-grade transformation can occur. Immunohistochemically , the ductal cells are positive for c-KIT , and myoepithelial cells are positive for p63 and SMA. Acinic cell carcinoma Acinic cell carcinoma is composed of neoplastic acinar cells. It is a low- to intermediate-grade tumour occurring mostly (90%) in the parotid gland. They typically present in the fifth decade and have a slight female predilection (1.5:1). They are generally slow-growing, painless, mobile, solitary tumours and rarely present with facial palsy . A small proportion may be high grade and may metastasise to cervical nodes and lung. Complete excision with an adequate margin is the recommended treatment. Recurrences can occur in cases of incomplete resection, deep lobe involvement and larger size tumours. On histopathology , the tumours show characteristic serous acinar cells and variable proportions of other cell types, including clear, vacuolated, intercalated duct-type oncocytic and hobnail features. They can have solid (most common), follicular or microcystic patterns with a prominent lymphoid infiltrate. Mitoses, necroses and nuclear pleomorphism are rare. Immunohistochemically , the neoplastic cells are positive for DOG1 and SOX10, whereas they are immunonegative for mammaglobin, di ff erentiating them from secretory carcinoma. Carcinoma ex pleomorphic adenoma (epithelial and/or myoepithelial) arises in association with primary or recurrent pleomorphic adenoma. The carcinoma component can be either purely epithelial or myoepithelial in presentation, with infiltration into the surrounding glandular and extraglandular tissue. It occurs mainly in the parotid gland, is more common in women and presents a decade later than pleomorphic adenoma (sixth decade). It often presents as a rapidly growing mass (within a longstanding swelling) associated with pain and facial palsy . Radical surgical excision with or without adjuvant radiotherapy is the treatment of choice. Local and distant metastases occur in 70% of cases with poor 5-year survival outcomes of 25–65%. On histopathology , the tumour shows variable propor - - tions of both pleomorphic adenoma and high-grade adeno - carcinoma such as salivary duct carcinoma or myoepithelial - carcinoma. It is subclassified as non-invasive or intracapsular (tumour is confined within pleomorphic adenoma), minimally invasive (tumour breaching the pleomorphic adenoma cap - sule) and widely invasive into adjacent salivar y gland and soft tissue. TP53 mutations and amplification of HER2 (in salivary - duct carcinoma) with a high degree of genetic instability and copy-number alterations are seen. Salivary duct carcinoma Also known as high-grade ductal carcinoma, salivary duct carcinoma is a high-grade adenocarcinoma, resembling high- grade mammary ductal carcinoma. The tumour may arise de novo or as a malignant component of carcinoma ex pleomor - phic adenoma. It is relatively less common and mostly arises in the parotid. It is commonly seen in elderly men in their sixth or seventh decade. Salivary duct carcinoma is an aggressive tumour presenting as a rapidly gro wing mass, often with facial palsy , pain and cervical lymphadenopathy . Complete excision with wide margins (total parotidectomy) with neck dissection is the treatment of choice. On histopathology , it resembles high-grade invasive ductal breast cancer with a large duct- like configuration with comedo necrosis and cribriform and Roman bridge-like features. Vascular and perineural invasion is seen. Salivary duct

carcinoma also shows androgen receptor and HER2 receptor positivity in a significant percentage of cases, making it a target for treatment, especially in the recur - rent/metastatic setting. It has a high predilection for local recurrence and regional and distant metastases and a poor overall survival. Malignant tumours

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