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Pleomorphic Adenoma of Parotid Gland- A Review

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Abstract:

Pleomorphic adenoma of the parotid is the most common tumor of salivary gland origin, accounting for 60 to 70 percent of all benign salivary gland tumors. This lesion usually presents as a slow-growing painless mass inferior to the pinna of the ear.The diagnosis is based on clinical presentation, magnetic resonance imaging or computed tomography, and fine-needle aspiration biopsy.The treatment is wide excision in which the entire capsule is removed but the facial nerve is spared. Proper diagnosis and treatment are necessary to prevent the complications of tumor recurrence and malignant transformation. Carcinoma ex pleomorphic adenoma arises in longstanding tumors and has a five-year recurrence rate of 75 percent.

Key words:

Myoepithelial Carcinoma, Salivary Duct Carcinoma, Adenoid Cystic Carcinoma, Adenoid, Cystic Carcinoma, Sarco Mastoid Carcinoma.

Introduction:

Pleomorphic adenomas of the salivary glands, also known as benign mixed tumours (BMT), are the most common salivary gland tumours. It is the most common benign salivary gland tumor composed predominantly by the proliferation of the myoepithelial cells and a wide spectrum of the epithelial and the mesenchymal tissue component surrounded by a distinctive capsule.[1]

Epidemiology

Pleomorphic adenomas account for 70-80% of benign salivary gland tumours and are especially common in the parotid gland.[1,2] Patients are typically middle-aged and the incidence is slightly higher in females than males (2:1).[1]The oncogenic simian virus (SV40) may play a role in onset or progression and prior head and neck irradiation is a risk factor for the development of these tumours.[3,4].

Causes:

The causes of pleomorphic adenomas are still unknown and the risk factors have not been fully ascertained yet. In addition to age, risk factors may be related to smoking habits, alcohol abuse, a diet rich in cholesterol and previous radiation therapy treatments in the face and neck area.[1]

Clinical presentation

Pleomorphic adenomas accounts for 60% of all parotid gland tumors,50% of submandibular tumors and 25% of sublingual tumors. It is encountered in patients of all ages. It is a slow growing tumor. It is soft or slightly firm on palpation and on larger gland it is freely movable. In parotid glands the tumor is spherical and arises in the superficial lobe as an obvious mass. In minor gland there is soft to slightly firm swelling without any ulceration.

Pleomorphic adenomas are also commonly found in the lacrimal glands where they account for approximately 50% of lacrimal gland tumours.[5]

Histopathology

As the name suggests, pleomorphic adenomas are composed of a mixture of variable histology. They contain both epithelial and myoepithelial (mesenchymal) tissues, with mixed histology. They appear encapsulated and well-circumscribed however the pseudocapsule is delicate and incomplete with microscopic extensions reaching beyond it, accounting for the high risk of recurrence when these tumours are enucleated.[5,6]

The gross appearance depends upon the relative proportion of epithelial elements and a stromal component which may range from myxoid to cartilage. Tumours with a prominent cartilaginous matrix have a bluish-grey opalescent appearance.[7]

Three histological types have been described: myxoid (hypocellular): most common, highest rate of recurrence, cellular and classic.

Radiographic features

On all modalities, these tumours typically appear as rounded masses with well-defined, "bosselated" or "polylobulated" borders (many small undulations, not truly lobulated). They are most commonly located within the parotid gland, particularly the superficial lobe. When they arise from the deep lobe of the parotid they can appear entirely extra parotid, seen in the prestyloid parapharyngeal space, without a fat plane between it and the parotid, and widen the stylomandibular tunnel. Pleomorphic adenomas can also arise from salivary rest cells in the parapharyngeal space itself without connection to the parotid gland.[7]

Ultrasound

They are typically hypoechoic and may show posterior acoustic enhancement. Ultrasound is also useful in guiding a biopsy (both FNAC and core biopsies) but needs to be carried out with care to avoid facial nerve damage.[8,9]

CT

When small, they have homogeneous attenuation and prominent enhancement. When larger, they can be heterogeneous with less prominent enhancement, foci of necrosis, and possible delayed enhancement. Small regions of calcification are common.[1,10]

MRI

The signal characteristics are homogeneous when the tumour is small. Larger tumours may be heterogeneous.

T1: usually of low intensity

T2: characteristically of very high intensity (especially myxoid type)[6]

often have a rim of decreased signal intensity on T2-weighted images representing the surrounding fibrous capsule

T1 C+ (Gd): usually demonstrates homogeneous enhancement.

Treatment and prognosis

Surgical excision is curative, however, as the tumour is poorly encapsulated (despite imaging suggesting otherwise) there is a significant rate of recurrence in the tumour bed. Exact rates of recurrence vary widely depending on series and surgical technique (1-50%).[1]Historically these tumours were removed by enucleation, resulting in recurrence rates of 20-45% .[6] To minimise this occurrence, no open surgical biopsy should be performed. Rather, a partial (superficial) or total parotidectomy ensures a wide margin. The facial nerve should be spared.[11,12] Using this approach, the recurrence rate has reduced dramatically to 1-4% Percutaneous ultrasound biopsy (both FNAC and core biopsy) can be performed safely and is associated with very low tumour seeding rates and without facial nerve injury provided meticulous technique is used 8,9. When in the minor salivary glands, a 5 mm margin should be obtained. These tumours do not invade into periosteum, thus bone need not be resected 4. When tumour bed recurrences occur, they can be extremely difficult to control, with management options including monitoring only, surgery, or radiotherapy.

Complications

There is a small risk of malignant transformation into a carcinoma ex-pleomorphic adenoma which is proportional to the time the lesion is in situ (1.5% in first 5 years, 9.5% after 15 years), thus excision is recommended in essentially all cases. Additional risk factors for malignancy include advanced age, large size, radiation therapy, and recurrent tumours.[2,6]In addition to carcinoma ex-pleomorphic adenoma, true malignant mixed tumours of the salivary glands usually arise from pre-existing pleomorphic adenomas.[1,3]Metastasising pleomorphic adenoma is the third type of malignant mixed tumour of salivary glands and is the rarest. It presents with metastases to lung, bone, and soft tissues despite having 'benign' histology.[1]

Conclusion

Pleomorphic adenoma is a common benign salivary gland neoplasm characterized by neoplastic proliferation of parenchymatous glandular cells along with myoepithelial components, having a malignant potentiality. Pleomorphic adenomas are more common in parotid gland. Even though they are rare in submandibular gland, they should be considered as differential diagnosis in case of submandibular mass lesions. Pleomorphic adenomas are treated by complete surgical excision. But, because of recurrence potential of this tumors, long-term follow up is necessary.[13,14]

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