

Recent enlargement of a parotid tumor: Self assessment questions

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Abstract

CLINICAL CASE

A 45-year-old male came with a recent history of a sudden increase in size of a swelling present behind and below his left ear for ten years. This swelling of around 3x3cm was neither painful nor had it caused any discomfort for the past ten years. Suddenly, for the last six months, it had started to grow from its original size to its present size. Now, occasionally he has severe pain which is like a pin prick sensation. He also complains of slight deviation of his mouth to the opposite side. On examination, he had a 15x15cm firm swelling in his left parotid region (Figure 1) with superficial necrosis. It had a bosselated surface, non-tender and was fixed to the underlying structure. He had a facial nerve palsy on the left side. Neck glands were not palpable.

Figure 1

Figure 1: Large bosselated mass over the left parotid area pushing the left ear anteriorly



WHAT ARE THE DIFFERENTIAL DIAGNOSES ON THE BASIS OF THE CLINICAL

PRESENTATION?

Rapid growth in a long-standing painless parotid tumour can be due to infection, cystic degeneration, haemorrhage or malignant transformation in a benign parotid tumour; secondaries from cutaneous malignancy of head and neck by lymphatic spread to intraparotid lymph nodes; metastasis from lungs, kidneys, breasts via blood, lymphoma or anaplastic carcinoma in benign lymphoepithelial lesions (Sjogren's syndrome, Mikulicz' disease) and Kimura's disease⁽¹⁾. The latter is a chronic eosinophilic inflammatory disorder involving subcutaneous tissues, lymph nodes of head and neck or parotid gland and it is characterized by lymphadenopathy, peripheral eosinophilia and elevated IgE. It is mostly benign but there are reports of its malignant transformation as well.

On clinical presentation, features like facial weakness, pain and paresthesia hint towards invasion by a malignant tumour. However, we can altogether not rule out a benign lesion as in large benign tumours, the nerve is often stretched around the mass thereby accounting for paresthesia, paresis and the tense parotid fascia leads to pain⁽²⁾ necessitating further investigations.

HOW TO CONFIRM THE DIAGNOSIS?

Fine Needle Aspiration Cytology (FNAC) is a non-controversial first line investigation and various authors recommend its liberal use^(4,5). It has a sensitivity of 85-99% and specificity of 96-100%. In the case of pleomorphic adenoma – the commonest benign parotid tumour – histopathologic examination shows the presence of epithelial cells, myoepithelial cells, mucoid material, cartilage and lymphoid tissue; hence it is also called mixed tumour. Malignant mixed tumour, on the other hand, is characterized by perivascular and perineural invasion, significant cellular

atypia and mitosis⁽²⁾. Magnetic Resonance Imaging (MRI) is also an essential investigation modality. It delineates the location, nature, extent and spread of the tumour and the anatomical relation of the tumour with the facial nerve and the internal architecture of the gland. Benign lesions appear as heterogeneous well-circumscribed masses of low signal on T₁-weighted images, with increasing signal on T₂-weighted sequences. More aggressive tumours have low T₁- and T₂-characteristics while high signal is observed on both T₁- and T₂-weighted images if there has been recent haemorrhage within the tumour. On the whole, MRI is superior to computed tomography (CT scan) excluding the fact that dystrophic calcification cannot be visualized on MRI⁽⁶⁾. Also, where facilities are available, newer modalities like gadolinium-enhanced MRI can be used to assess extraglandular infiltration and electroneuronography can be used to evaluate facial nerve involvement preoperatively⁽⁶⁾. Thus, correlation of clinical histopathological and radiological findings clinches the diagnosis and helps to decide on further management.

WHAT IS ITS MANAGEMENT ?

Surgery is the mainstay of the management of parotid tumours. Based upon the histological grade of the tumour and TNM staging, the type of surgery is decided. The rationale is to do superficial parotidectomy if the tumour is localized in the superficial lobe; total parotidectomy with sparing of cranial nerve VII if it is in the deep lobe or both lobes are affected and radical parotidectomy where the facial nerve is also sacrificed if it is macroscopically infiltrated by the tumour⁽³⁾. This neural involvement can be confirmed by frozen section histology done intraoperatively. Subsequent reconstruction should be done during the same sitting either by cable graft (using greater auricular nerve, sural nerve, IVth cranial nerve) or nerve transfer (using hypoglossal nerve). In the event that this is not done, an attempt to at least reconstruct zygomatic and buccal branches for eyelid closure and oral competence is advocated. Also, if there is suspicion of malignancy, sampling of upper deep cervical lymph nodes with frozen section is done followed by radical block dissection⁽³⁾ of neck if nodes are positive. In addition, if there is blatant infiltration of overlying skin, mandible, and these structures are also removed, then reconstruction is achieved by chest flap or free forearm flap. Adjuvant radiotherapy with total doses of 6000-6500 cGy over 6-7 weeks is indicated in cases of high grade tumour, positive margins, deep lobe tumours, nerve involvement, nodal metastasis, spillage, recurrence and doubtful adequacy of

surgery⁽²⁾. Chemotherapy has a role mostly in unresectable tumours and cytotoxic drugs used are cisplatin, cyclophosphamide and adriamycin⁽²⁾.

DISCUSSION

Pleomorphic adenoma is the commonest benign tumour of the salivary gland (50-70% of benign tumours). Because it is slow-growing, the patient can tolerate the swelling for many years before seeking medical advice. However, after a period of 15-20 years, it is estimated that 6% of pleomorphic adenomas will undergo malignant transformation⁽³⁾. It is predominantly the epithelial component of the mixed tumour which undergoes malignant transformation. Other studies say that the risk for malignant transformation is 1.5% within the first 5 years but increases to 9.5% once the benign tumour has been present for more than 15 years⁽⁷⁾. The propensity for malignant transformation is greater in patients below 50 years with pain and non-inferior pole lesion⁽⁸⁾. The malignant pleomorphic adenoma, also called carcinoma ex pleomorphic adenoma, is an aggressive tumour with high incidence of metastases and carries a bad prognosis. The 5-year survival rate varies from 50-70% but the 20-year survival approaches zero⁽³⁾. Thus, its successful management depends upon early diagnosis and a radical parotidectomy with immediate soft tissue and nerve reconstruction. However, owing to a disappointingly low sensitivity of FNAC for carcinoma ex pleomorphic adenoma, the diagnosis is delayed and inadequate resection is done⁽⁹⁾. In this regard, tenascin expression during tumour morphogenesis as revealed by immunohistochemical studies may help ⁽¹⁰⁾.

Concerning the type of surgery selected in parotid tumours, it is obvious that in order to achieve the least recurrence and morbidity, the tumour as well as minimal amount of normal parenchyma must be resected. Thus, limited resection, partial superficial parotidectomy and functional superficial⁽¹¹⁾ parotidectomy are done where expertise is available. In partial superficial parotidectomy, a 1cm margin of normal parotid parenchyma is resected together with the tumour while in functional superficial parotidectomy, efforts are made to preserve the function of the residual gland^(11, 12). Benefits offered by limited resection are: smaller incision, less post-operative facial distortion, short operating time and lower incidence of Frey's syndrome because less of the parotid gland is removed⁽⁶⁾. In the case of a deep lobe tumour, some even advocate sparing the superficial lobe as it gives better cosmetic results and also there is less likelihood of gustatory sweating because of the interposition of tissue

between the skin and the cut ends of the secretomotor fibres⁽¹³⁾. Post-operative complications are: infection, nerve injuries, Frey's syndrome, facial palsy, salivary fistula, haematoma, haemorrhage, neuroma, keloid scar, greater auricular nerve schwannoma, seroma and recurrence⁽¹⁴⁾. The latter is due to the fact that pleomorphic adenomas often form finger-like projections into the pseudocapsule formed by the compressed peripheral parenchyma. These pseudopods, when left behind after tumour resection, give rise to recurrence. However, because local recurrence is not an immediate threat to life and is benign and slow growing and therefore often detected by the patient himself, clinic attendance for follow-up is unnecessary and more emphasis is laid on patient education and self examination⁽¹⁵⁾.

Radiation as adjuvant therapy is used mostly in cases where resection of the neoplasia is inadequate. Radiation portals should include the site of surgery, base of skull with mastoid process, stylomastoid foramen and foramen ovale. However, radiotherapy has drawbacks like xerostomia, otitis media, hair loss, skin changes, dental caries and increased incidence of second malignancies in children. Consequently, a low total dose of radiation is given in low-grade tumours and a high total dose in high-grade tumours.

In summary, carcinoma ex pleomorphic adenoma is an aggressive tumour developing in long-standing pleomorphic adenoma. It is diagnosed based on FNAC and MRI and is treated by superficial parotidectomy or partial superficial parotidectomy and adjuvant radiotherapy.

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