

Intra-Cranial Metastatic Lacrimal Gland Tumour

N A Wahab*, U Ramasamy****, J George*, M Madhavan**, A R Arif*, J Abdullah*

*Department of Neurosciences, **Department of Pathology, ***Department of Radiology, ****Department of Surgery, School of Medical Sciences, Universiti Sains Malaysia, Kubang Kerian, Kelantan

Summary

We report a case of an adult who presented with progressive swelling in the right eye with suggestive of intra-cranial lesion on imaging. Histopathological examination revealed a lacrimal gland malignant mixed tumour.

Key Words: Malignant Lacrimal Gland Tumour, Intra-cranial metastasis, Mixed Gland Tumour

Introduction

The term mixed cell tumour was first used to describe lacrimal gland lesion as early as 1874. The term at first encompassed both benign and malignant lesions but gradually evolved to refer specifically to pleomorphic adenoma. The lesion was originally called mixed tumour, because it consisted of two distinct elements; the epithelial lining of the ducts and mesenchymal appearance. Epithelial tumours of the lacrimal gland are rare. Among the lacrimal gland tumours, benign mixed tumour (pleomorphic adenoma) is found most commonly. The remaining tumours are malignant which are rare.

Case report

A 44-year-old man was admitted with progressive generalised weakness and intermittent frontal headache for one month, associated with right progressive orbital protrusion. There was no history of trauma, fever and vomiting. His short term memory was gradually abolished, associated with urinary incontinence. At ten years old, he claimed to have undergone orbital surgery for a growth removal. Unfortunately, it recurred after one year and he defaulted follow up. He had unequal distribution of weakness of his limbs, with brisk reflexes, up going plantar response and no sensory

deficits. Ophthalmologic examination revealed right proptosis, with the globe pushed forward and downward with upward movement restriction. Fundoscopic examination showed swollen and infiltrative optic disc with congested vessels. His haematological and biochemical investigations were within normal.

A CT scan brain with contrast demonstrated an aggressive tumour mass encroaching into the right frontal sinus, lateral and superior orbital wall, with involvement of right fronto-parietal region of the brain. Massive cerebral edema with mass effect was seen (Fig 1).

An emergency debulking surgery was done. The tumour was found to be infiltrating into the right cavity of the orbit, the orbital bone, underlying dura and Sylvian fissure of the brain. Histopathological examinations revealed cuboidal to polygonal epithelial cells arranged in islands and tubules in hyalinised stroma as that of a mixed tumour. On higher magnification (Fig 2), the epithelial cells appeared pleomorphic and contain hyperchromatic nuclei and showed atypical mitotic figures. There is evidence of brain parenchyma tumour infiltration. Hence, a diagnosis of malignant mixed tumour was made.

However, post-operatively the patient developed septicaemia. The source of infection is most probably

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Corresponding Author: Nasser Abdul Wahab, Department of Neurosciences, School of Medical Sciences, Universiti Sains Malaysia, Kubang Kerian, Kelantan

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from the exposed frontal sinus which was infiltrated by the tumour. A routine septic work-out failed to isolate any specific infective organism. In addition, the patient also developed acute renal failure. Despite the intravenous antibiotics, the patient pass-away on the second week post operatively.

Discussion

The lacrimal gland is considered to be a minor salivary gland that shares histologic features with the major salivary glands. Hence, in many respects, lacrimal gland tumors resemble those that arise in the major salivary glands. Among malignant tumours originating in salivary glands and lacrimal glands, adenoid cystic carcinoma is seen most frequently¹. Malignant mixed tumour arising in lacrimal gland is rarely described. These lesions showed both areas of benign mixed tumour and malignancy. The malignant components are most often adenocarcinoma. In this particular case, it shows mixed pattern as that of benign mixed tumour.

However, the epithelial component is malignant in nature.

In a benign mixed tumour, all of the cells should have benign cytologic features. These tumours grow by expansion and do not show evidence of true invasion. When invasion into adjacent tissue is identified, the lesion is fully malignant and can metastasize. Our case showed both radiological and histopathological evidence of infiltration.

Malignant mixed tumour of the lacrimal gland usually present clinically with proptosis and displacement of the globe, as the lacrimal gland is located supero-temporally above the globe within a small fossa behind the orbital rim. Upper eyelid fullness may be found associated with a palpable mass in the lacrimal fossa. Possible clues of malignancy include presence of pain, bone destruction on radiographic studies, and rapid enlargement. Benign mixed tumour of lacrimal gland evolves slowly over years, whereas malignant tumours usually in about six months with shorter symptoms. Any acceleration of symptoms over a short period is most likely to have a malignant mixed tumour. These are to be differentiated from inflammatory lesions of the lacrimal gland, with reduction in the volume of lesion following treatment. The results of computed tomography obtained before treatment should be compared to the results after therapy.

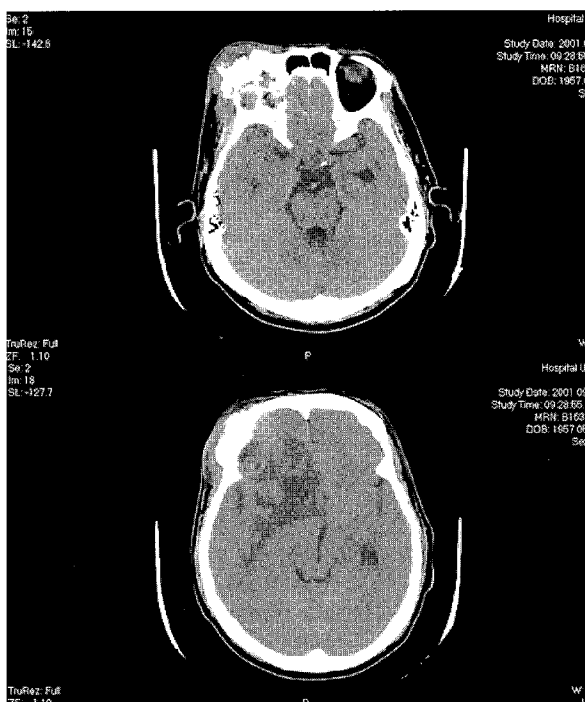


Fig 1: A CT scan brain with contrast demonstrated an aggressive tumour mass encroaching into the right frontal sinus, lateral and superior orbital wall, with involvement of right frontoparietal region of the brain



Fig 2: Epithelial cells infiltrating the brain parenchyma shows the cytological features of malignancy. (H&E, 100X)

It was traditionally believed that a small circumscribed lesion is more likely to be benign and should be managed by excisional biopsy. In contrast, a larger or poorly circumscribed tumor is more likely to be malignant and should have incisional biopsy followed by orbital exenteration if malignancy is confirmed histopathologically. However, there is an emerging philosophy that an attempt should be made to do an excisional biopsy on all cases if possible and that the diagnosis of a benign or malignant epithelial tumor of the lacrimal gland should be made histopathologically after the suspected tumor is removed completely by excisional biopsy². The clinician should decide on the basis of the history and the radiographic findings whether a patient more likely has benign or malignant tumour.

The recommended treatment for malignant epithelial tumours includes orbital exenteration (removal of the lids, globe, orbital soft tissues, and periosteum of the orbital walls). When a malignant tumour is suspected, an incisional biopsy can be performed through the lid to establish the diagnosis before proceeding with

orbital exenteration, including the biopsy site. A lateral orbitotomy should be used for performing a biopsy on a possibly malignant tumour, because tumour cells may be implanted in the periosteum, bone or temporalis fossa. En bloc one stage resections of all the lacrimal gland tumours, including the bony wall of the lacrimal gland fossa, are performed to minimize early dissemination via bone.

In recent years, immunohistochemistry has been helpful in establishing the diagnosis of a number of lacrimal gland tumors that were previously not known to occur in the lacrimal gland³. Immunohistochemical studies have also been reported to provide clues to the histogenesis of benign and malignant mixed tumors of the lacrimal gland⁴.

Once a carcinoma evolves in a mixed tumour of the lacrimal gland, the prognosis appears to be poor. Such patients may present in various forms of metastasis; pulmonary, cervical lymph node, local extension into the anterior cranial fossa, and even with intracranial metastasis.

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