LACRIMAL GLAND CARCINOMA

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SUMMARY

Three consecutive cases of lacrimal gland carcinoma seen at the University Hospital during a twelve-year period are presented. One case was unusual, presenting at the age of sixteen years and developing metastases to the lungs. Adenoid cystic carcinoma is the commonest type of lacrimal malignancy and its clinico-pathological features and surgical management are discussed.

INTRODUCTION

Carcinoma of the lacrimal gland is a relatively rare form of malignancy. Chandran and Bhandari [1] found only three such cases at the University Hospital, Kuala Lumpur, during the period 1968-81, where 46 orbital biopsies were performed. These tumours are so uncommon that no single institution can collect and study a substantial number of cases. There have been no previous reports of lacrimal gland carcinoma from this region.

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The aim of this paper is to present 3 cases of lacrimal gland carcinoma, which includes a case with the unusual complication of metastases to the lungs.

CASE 1

P.N. was a 24 year old Indian female seen in 1976 with a complaint of proptosis of left eye with periorbital pain and deteriorating vision for 5 months.

On examination visual acuity was 6/5 in the right eye and 6/36 in the left eye. She had left proptosis (7mm) downwards, and nasally. Movements were limited superiorly and laterally with diplopia in these positions. A lacrimal tumour was palpable.

Investigations: Blood: N.A.D. V.D.R.L. — Negative. Thyroid function tests were normal. X-ray skull showed abnormal depression of left lacrimal fossa.

A lateral orbitotomy revealed mass in superior-lateral part of orbit extending to floor of orbit. It was not encapsulated, firm in consistency and attached to the lateral rectus. It cuts with a gritty feeling. Eyeball with tumour was excised.

Histopathology: Shows sheets and trabeculae of closely packed cells with large nuclei exhibiting frequent mitoses and scanty cytoplasm. Cystic and glandular spaces are present in areas giving it a cribriform appearance. The groups of tumour cells are separated by a hyaline stroma (Fig. 1). The margin of resection is involved by the tumour.

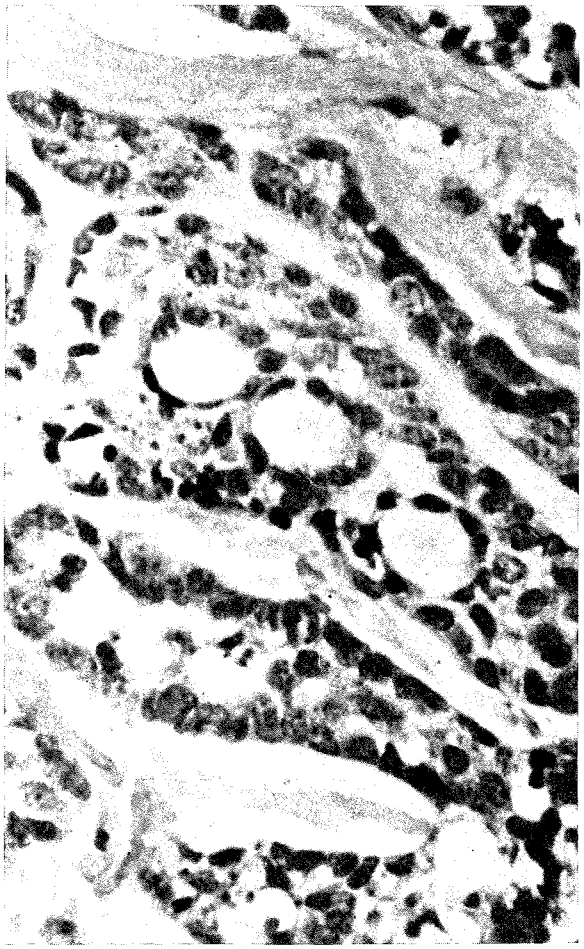

Fig. 1 Cystic and glandular spaces are present in areas giving it a cribriform appearance.

Diagnosis : Adenoid cystic carcinoma.

On follow up the patient defaulted and came only when there was recurrence of proptosis, with pain on left side of the face and inability to open mouth completely. As it was radioresistant, palliative extenteration was done. There was some residual mass.

CASE 2

C.O.Y. was a 16 year old female Chinese seen in 1971 complaining of a gradually enlarging lump in the right eyelid for 2 years associated with epiphora and occasional dull pain.

On examination visual acuity was 6/9 in the right eye and 6/6 in the left eye. The right eyeball was displaced downward and nasally. There was a hard mass in the upper outer quadrant not attached to skin but appears attached to bone. No restriction of movement, bruit or pulsation. Fundus: N.A.D.

Investigation: X-ray skull — There is a loss of translucency in the right orbit otherwise N.A.D. Chest X-ray N.A.D., K.T. negative.

Right anterior orbitotomy was done and a hard conical tumour measuring 3 x 3 cm extending from orbital margin to the apex of orbit was seen. The roof and lateral margin of the orbit was thickened. Tumour was not removed because of its extensive nature.

Histopathology: Shows groups of tumour cells with hyperchromatic nuclei, exhibiting occasional mitoses, separated by a hyaline stroma. In areas glandular and cystic formations, containing mucin was present. Diagnosis: Adenoid cystic carcinoma.

The patient was given a full course of radiotherapy and discharged. She was admitted again in May 1974 for right corneal ulcer and she developed haemoptysis.

Chest X-ray showed extensive metastases in both lungs. X-ray skull showed partial destruction of the roof and lateral wall on right side. There was a soft tissue mass in the frontal sinus with destruction of bony margins. The frontal bone showed some patchy destruction. The patient was referred to Penang General Hospital.

CASE 3

S.S was a 32 year old Malay woman seen in 1977, with a complaint of swelling in the right temple for one year and increasing rapidly in size for the past 2 months.

On examination visual acuity 2/60 in the right eye and 6/9 in the left eye. There was right proptosis 11mm forwards and downwards. A hard bilobed mass in the right upper temporal quadrant fixed to bone was palpable. There was chemosis of the conjunctiva. The fundus appeared normal.

Investigation: Chest was N.A.D. Skull X-ray showed that right optic foramen increased in size and defective temporally. Blood T.P.H.A. was non-reactive.

An excision biopsy was done by the anterior approach.

Histopathology: The tumour is composed of cords and sheets of cells with occasional arrangement in the form of incomplete acini and glandular structures. (Fig. 2). The cells are

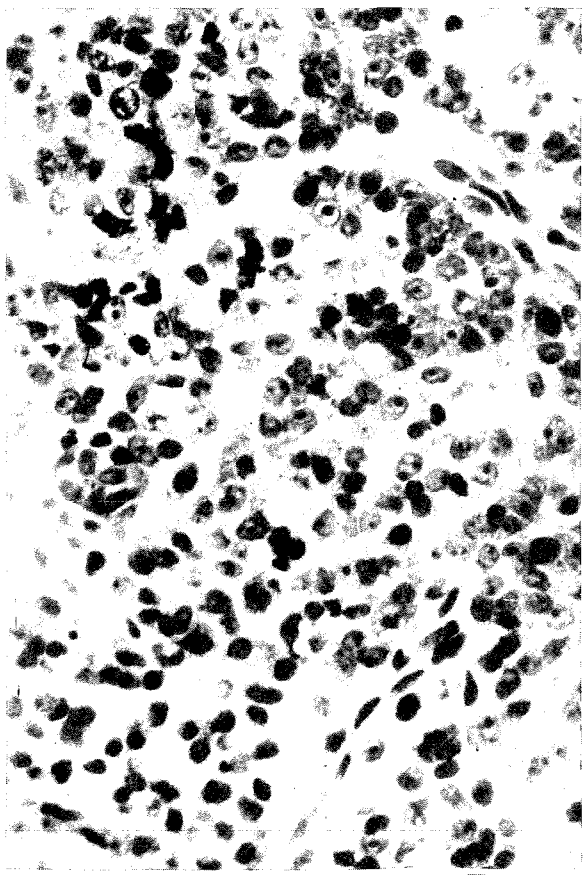

Fig. 2 The tumour is composed of cords and sheets with occasional arrangement in form of incomplete acini and glandular structures.

pleomorphic with large vesicular nucleoli. Mitoses are occasionally seen. Diagnosis: Adenocarcinoma, moderately differentiated. The patient refused further treatment and no follow up information is available as she defaulted.

DISCUSSION

Adenoid cystic carcinoma (ACC) accounts for 2 out of our 3 carcinomas of the lacrimal gland. The third is a moderately differentiated, adenocarcinoma. ACC is the most common single type of malignancy of the lacrimal gland. Of the 26 cases of lacrimal gland tumours of epithelial origin studied by Forrest, [2] 11 were of this type. It derives its name from a distinctive histological pattern, which consists of sheets, or trabeculae of deeply staining polygonal cells, with areas showing cystic spaces of varying sizes, lined by cells that are several layers in thickness. ACC are not confined to the

Fig. 3 Lacrimal tumour with forward and downward displacement of the eye and drooping of upper eyelid.

lacrimal gland but also encountered, though less frequently, in the salivary glands.

Zimmerman and associates [3] in a study of 29 ACC found the median age to be 37.5 years. In a later study by Henderson [4] of 9 cases, the median age was 44 years, the ages ranging from 27 to 65 years. It will be noted that our second patient was only 16 years of age. Ley and Wolter [5] reported 2 cases in 12 year old girls.

In addition to ACC there are a number of miscellaneous malignant tumours such as adenocarcinomas of varying degrees of differentiation squamous cell carcinoma, and malignant mixed tumours.

Most of the clinical signs of lacrimal gland carcinomas are similar to other lacrimal tumours; there is a palpable orbital mass in the superior temporal quadrant, downwards and nasal displacement of the eye, swelling, drooping of the adjacent upper eyelid (Fig. 3) and increased lacrimation. All our patients in this report, had a palpable tumour with downward displacement of the eye. In the two cases with ACC, pain was a presenting feature. Pain and tenderness are caused by infiltration of orbital nerves by these rapidly growing unencapsulated tumours. Infiltration is also the cause of bone erosion seen in the X-ray films. In an earlier stage bone erosion may be absent as in Henderson's [4] 9 cases which showed no

radiological changes when first seen. Following local spread to periorbital tissues and the bony orbit, the carcinoma may extend to the temporal region. Intracranial extension through the optic foramen is not uncommon. Metastases to the lungs seen in Case 2, three years after onset, is unusual, although generally regarded as a possibility.

As ACC is a highly malignant tumour, radical operation offers the only chance of cure. However, in the Mayo Clinic study 8 out of 9 cases died despite extenteration alone, or combined with bone removal and irradiation. Extenteration implies that all soft tissue has been removed from the orbit but it is difficult to ensure that tumour tissue is not present at the excised margins. In two of our cases, the surgical margins were involved and complete clearance was not feasible. In ACC the carcinoma with the attached periorbital tissue and adjacent bone, should be excised immediately after biopsy and frozen section confirmation, at a single operation. [6] A more rapid spread can be expected, if only a preliminary biopsy is performed and followed by an excision at a later date. Opinion is divided as to the effectiveness of irradiation in lacrimal gland malignancies. It is advocated post-operatively, by some, in cases of high grade malignancy especially if there is doubt about complete removal of the tumour.

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