CASE PRESENTATION

Behavioural Patterns of Adenocystic Carcinoma of Lacrimal Glands

By Hassan Al Arrayed* and Vaidya P.L.*

* Consultant of Ophthalmology
Salmaniya Medical Centre
Bahrain

SUMMARY

ADENOCYSTIC carcinoma or a cylindroma is a highly malignant tumour of the salivary glands. Very rarely it arises from the lacrimal gland.

Here we report two cases of adenocystic carcinoma of the lacrimal gland seen in the last 5 years in Bahrain and treated at Salmaniya Medical Centre with emphasis on the clinical course in relation to the histopathological pattern.

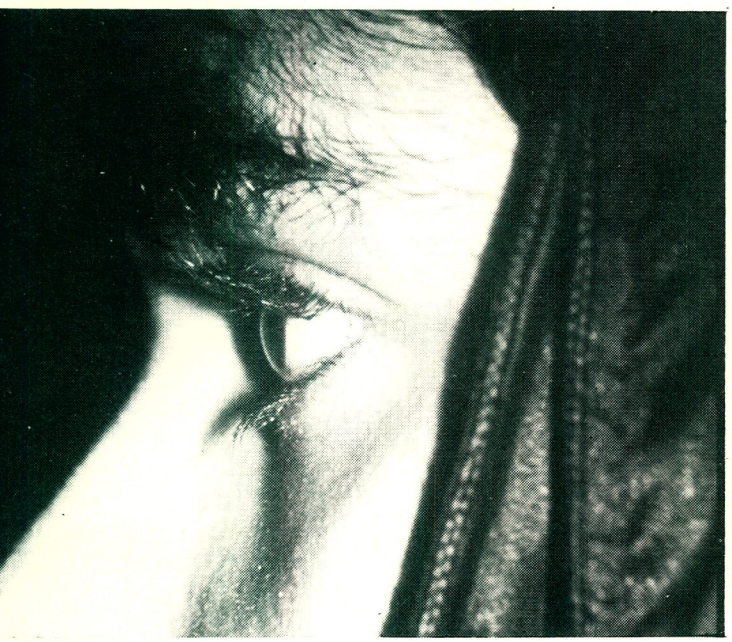
CASE REPORTS

Case No. 1 (F.S.A.)

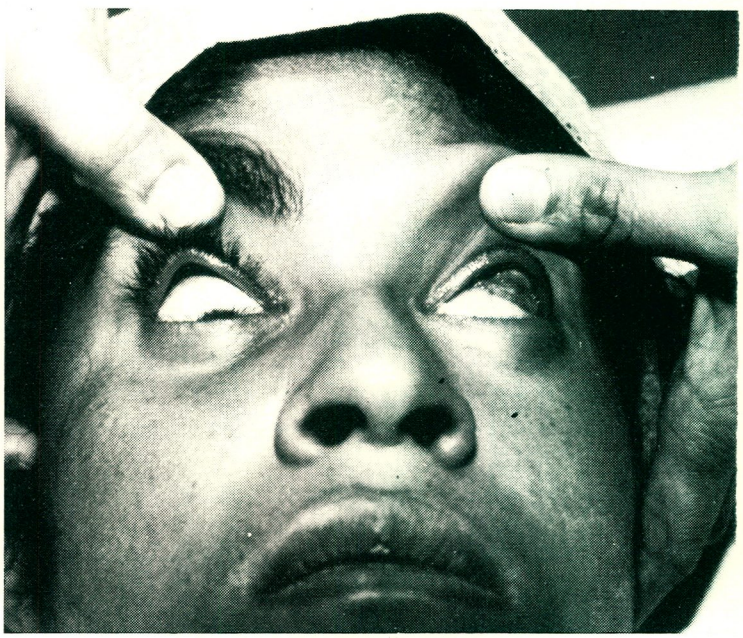
F.S.A., a 16 year old Bahraini female presented to the eye clinic on 24.6.79 with slight discomfort and redness in the left eye of 2 days duration. Visual acuity was 6/6 in each eye. Examination of the left eye revealed a slight proptosis with inward deviation of the eye ball (Fig. 1). On palpation, a firm mass in the upper outer quadrant of the orbit was left.

X-ray of the orbit confirmed the presence of mass which was indenting the left lacrimal fossa. Provisionally the tumour was diagnosed as a lacrimal gland tumour, most probably a benign mixed tumour (Fig. 2). Excisional biopsy was performed on 28.6.79. The whole mass came almost in toto. The

1. Case 1 : Pre op propotosed eye ball.
2. Case 1 : On the operating table pre op showing the fullness of the outer upper fornix Left Eye.

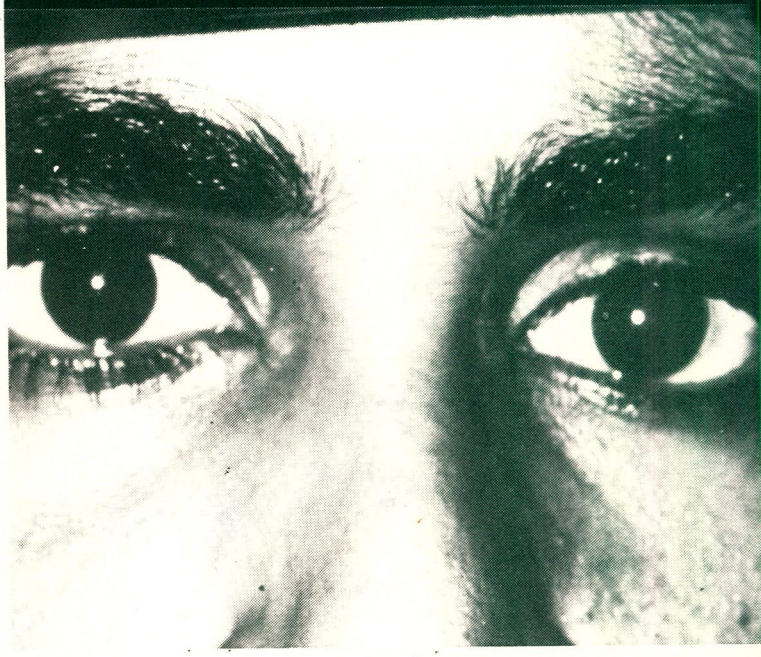


1



2

3



4

3. Case 1 : Post op the eye ball regressed and no more propotosed.

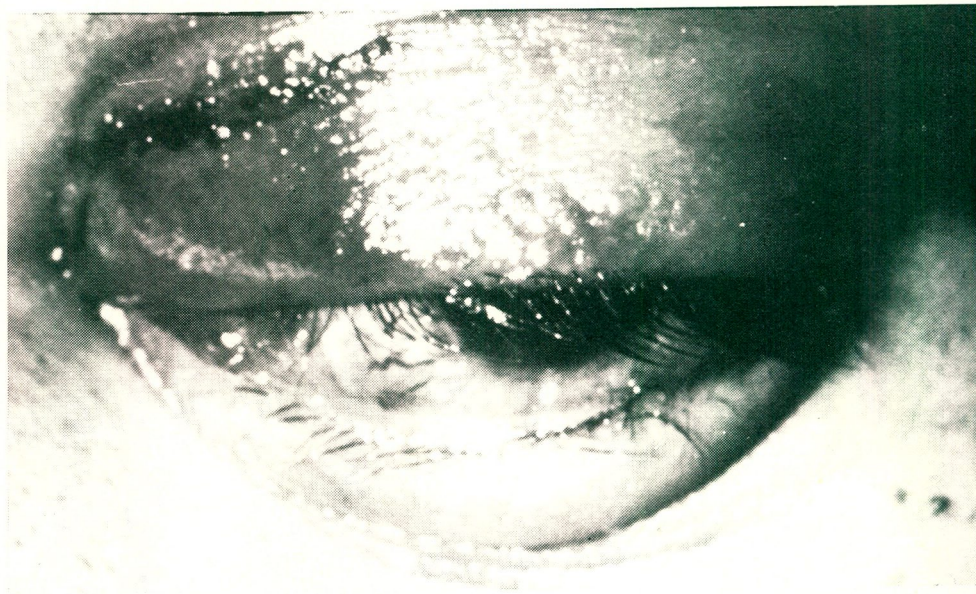
4. Case 1 : Pre op showing proptosis of left eye.

5. Case 2 : Showing sever proptosis and oedema. Also showing the healing wound done for the Incisional biopsy.

proptosis almost disappeared (Fig. 3) and the eye recovered very well (Fig. 4). The histopathology report was one of Adeno Cystic Carcinoma of the lacrimal gland. A second opinion from the department of Ophthalmology, IOWA University confirmed the histopathological diagnosis.

Following the excisional biopsy, patient did not receive any adjunctive post operative radiation therapy. Periodic follow-up did not show any evidence of local recurrence or distal metastasis.

The patient's father was reluctant for the exenteration until Dec. 1979. Exenteration was done on 10th Dec. 1979. In the areas of bony indentations an attempt was made to excise the lacrimal fossa floor particularly. The orbital walls were covered with a split thickness skin graft from the thigh of the patient. Thereafter she was followed up regularly.



5

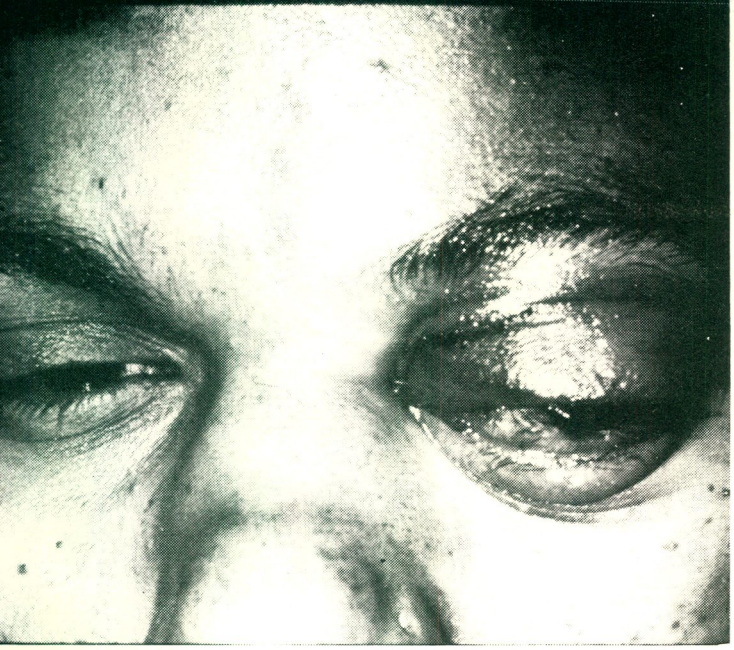
One year after the excision of the mass she was found to have a firm nodular swelling on the outer part of the orbital margin, which gradually increased in size and became tender. It was suspected to be a local recurrence of the adeno cystic carcinoma.

Excision of the mass was done on 24.3.81 which was confirmed histopathologically as adeno cystic carcinoma of the lacrimal glands. She still complains of pain in the socket. She is regularly being followed up by the same person and could observe no evidence of local recurrence or direct metastasis.

Case No. 2 (Z.A.R.)

A 26 year old Bahraini female (Z.A.R.) attended the eye clinic on 6.3.80 for the first time complaining of pain and protrusion of the left eye ball, and left sided headache of one year duration.

Vision in the left eye was counting fingers at 5 meter and 6/18 in the right eye. She had myopic refractive error in both eyes. Tomography of the orbit showed a diffuse swelling of the supra orbital bone with stipling. Proptosis started to increase rapidly with increasing tenderness of the supra-orbital region (Fig. 5). Con-



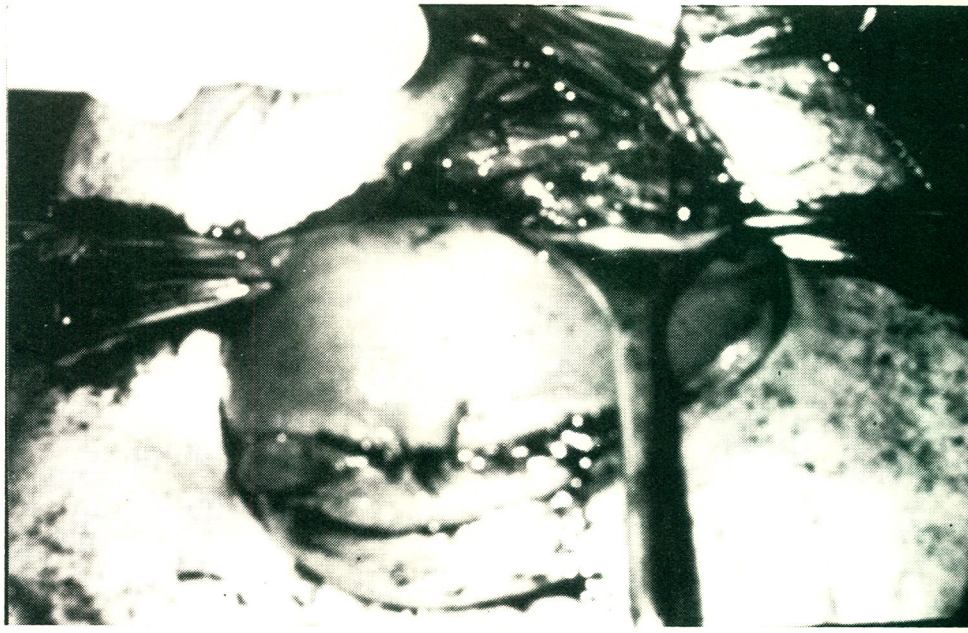
6

7

6. Case 2 : Showing the whole left side is oedematores and irregularities of bony surfaces of the skull. The eye ball is pushed downward.

7. Case 2 : During the attempt of excentration opening what could be orbital septum at the superior orbital margin.

8. Case 2 : Going deeper. The bones of the skull were completely eroded and necrotic. The brain tissue was almost showing. At this stage it was felt excentration is very dangerous for the patient.



8

sidering it an inflammatory swelling (pseudotumour), orbitotomy was not done and at the same time she was put on systemic antibiotic and steroids. Inflammatory signs reduced slightly and the patient left the hospital against medical advice on 19.3.80. On 22.3.80 the patient was readmitted with increased pain, proptosis and headache (Fig. 6). She had a palpable mass on the medical upper quadrant of the orbit. Incision biopsy was done on 26.3.80 which revealed adeno cystic carcinoma of the lacrimal gland.

Following the incisional biopsy, the tumour grew rapidly with restriction of all eye movements and sever loss of vision of the affected eye. Exenteration was attempted on 13.4.80 (Fig. 7), but could not be achieved due to the massive infilteration of the tumour into the surrounding soft tissues and the bones of the skull (Fig. 8). She was sent to Kuwait for radiotherapy on 1.5.80.

Apparently she was relieved of the symptoms following the radiotherapy. However the vision was totally lost. After 7 months, on 10.1.81 she came with severe pain, swelling of right forehead and protrusion of the right eye ball with complete loss of vision of 2 weeks duration. X-ray of skull showed marked bony metastasis of the right orbit and skull bones which was an evidence of advanced spread of the tumour with very grave prognosis. During her stay in hospital, tumour spread very rapidly with sequelae of malignancy.

In the hospital the patient was confused. On 17.1.81 patient fell from her bed and had severe epistaxis from the fragile tumour extensing in the sinuses.

She died the next day early morning.

DISCUSSION

Clinical Features :

The early signs and symptoms of

this highly malignant tumour are very similar to those of mixed benign tumours affecting the salivary or lacrimal glands. When the tumour arises from the lacrimal gland, the patient usually presents with minimal watering or discomfort in the eyes. There is forward and downward displacement of the eye ball by a palpable firm mass in the superior temporal quadrant of the orbit. Upper eye lid fullness may be present. Usually there is no pain or tenderness, but it is frequently present in recurrences which invade the periorbital and adjecent nerve fibres.

The pain is referred to the temporal side of the eye brow or even to the forehead. Limitations of eye movements will be present in upward and outward gaze[1] as the tumour infilterates the extraocular muscles as seen in Case No. 1.

Adeno cystic carcinoma is a slowly growing, relentlessly progressive malignant epithelial tumour. It usually arises from the salivary glands (about 20% of all the tumours of the salivary glands are adeno cystic carcinoma). It arises less frequently from the lacrimal gland[2].

Radiography studies would show increased bone density with or without bony erosion. X-ray studies are of a limited differential diagnostic value[3]. Richard and Dallow[3] studies has mentioned some information about orbital soft tissue lesion which may could also be gained from these studies; but often the findings are not definite in these cases and he indicated that one of the most valuable techniques for defining many soft tissue abnormalities of the orbit is ultrasonography which can differentiate many soft tissue abnormalities of the orbit. A mode and B mode scan can help in the location, configuration and outlining of the extent of the tumour, also the tissue type may be categorised broadly by ultrasonic criteria into cystic, solid, angiomatous or infilterating type[3]. However this mode of diagnostic technique was still not available in Salmaniya Medical Centre.

Clinicopathological course :

It is the most malignant among all lacrimal gland tumours. The tumour is slow growing and invades the surrounding soft tissues and bones. Extension occuring mainly through the perineural tissue, but could also occurs along tissue plains and via the blood vessels.[4] It has a very high local recurrence rate after surgical removal of the primary tumour with a late involvement of regional lymphnodes and distant metastasis. Chemotherapy is ineffective except adriamycin which brings about partial remission[2].

The peak incidence of adeno cystic carcinoma is in the fourth decade. It is commonly seen between the age of 30 — 60 years, although much younger patients may rarely be affected. Female predilection has been observed in some studies. (both our patients were female). Hendersons series[1] reported a 12 year old female patient with adeno cystic carcinoma. Font and Gamel[3] who reported the largest series in the literature gave an age range of 12 — 76 years. Dagher et el reported on the youngest patient 9 years old with a malignant epithelial tumour of lacrimal gland[5]. In our study Case No. 1 was 16 years old female.

Regional lymphnondes metastasis are late, but it has been observed in 20 — 30% of all the patients with A.C.C. and distant metastasis in about 40%.

There is often a very long interval between the establishment of the diagnosis of the primary tumour and the appearance of the metastasis.

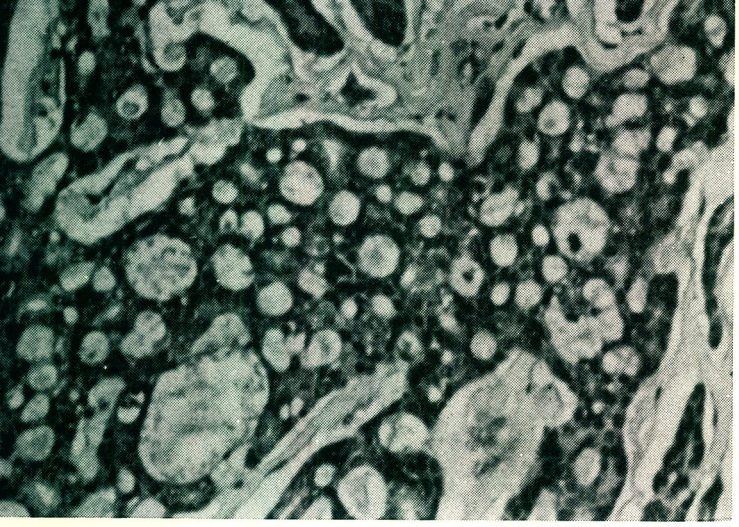
HISTOPATHOLOGY

The majority of these tumours arise denevo, but the few that arise from benign mixed tumour have a better prognosis. Macroscopically it may be a capsulated or an incompletely encapsulated tumour. Usually it is a greyish firm mass with a nodular surface. It consists of cords or sheets of small uniform epithelial cells occuring in a glandular or cystic pattern of different sizes containing mucin (Fig. 9).

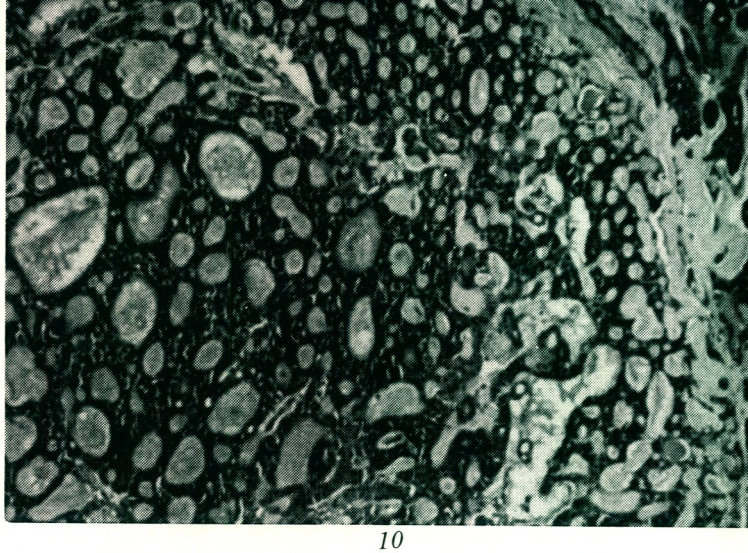
Occasionally one finds areas of solid cellular growth with a basolid or anaplastic appearance and areas of coagulation necrosis (Fig. 10).

A more rapid and fulminating clinical course with early recurrences, metastasis and death within 2 — 3 years occures with this histologic pattern as in Case No. 2.

In the studies of Eby et al in 1972 and Eneroth et al, they concluded that A.C.C. which demonstrate a solid pattern had a worst prognosis and more aggressive clinical behaviour than with the tubular pattern[6]. Tubular pattern was the most differentiated from of A.C.C. with best prognosis[7] as in Case 1. In Case No. 1, the tumour consisted of cords and small columns of basal like cells surrounded by cystic spaces producing a cribriform pattern (Fig. 8). The cystic spaces were filled with mucin. This was a well differentiated tumour having the best prognosis, but the patient came back with local recurrence after one year and that was removed successfully. So far she has no evidence of distant metastasis inspite of the delay in exentrating the orbit and having no post operative radiation therapy.



9



10

9. Case 1 : Low power histopathology picture of adenocystic carcinoma of Lacrimal gland of case one.
10. Case 1 : High power histopathology picture of adenocystic carcinoma of Lacrimal gland.

While in Case No. 2, the tumour was composed of cells set in abundant fibrotic stroma with aciner foundation and occasional cysts. Cytoplasm was illdefined and nuclei were hyperchromatic. Diagnosis and surgical interference were rather delayed as the patient presented late. This pathological pattern would indicate a very bad prognosis. She had a very rapid spread of the tumour with local and distant metastasis. Ultimately she died in less than one year time.

TREATMENT

The objectives in the treatment of adeno cystic carcinoma or orbital origin should be complete excision of the mass enbloc rather than biopsy.

As the adeno cystic carcinoma has an infilterative character, complete removal is not possible; but one should make an attmpt to remove the tumour as completely as possible to minimise the chances of local recurrences and remote metastasis. It has been shown on examination of exenterated specimen that it generally reveals the incomplete excision of the tumour and tumour cells have been left behind in the remaining orbital soft tissue or bone[4]. A more rapid spread can be anticipated if only a biopsy is performed (Case No. 2). Usually there is delay between the two operations as patient will hesitate to accept such a drastic surgery without additional consultation. (Case No. 1).

Effectiveness of post operative irradiations is doubtful. A total dosage of approximately 6 000 R (as measured in air) is administered in cases of bony invasion and incomplete surgical removal.

Irradiation slows down the rate of progression and may provide temporary relief of pain with or without bone resection.

In Mayo Clinic study[1], 8 out of 9 patients died after exenteration with or without bony excision. 9th patient was lost for follow up. In Zimmerman's series of 23 patients of Adeno Cystic Carcinoma, all had recurrences. 7 were still living at the time of the study. Average period of survival was 4 years[1].

CONCLUSION

We conclude from our study that the prognosis of adeno cystic carcinoma could be predicted from the histopathological pattern. Case No. 1 was encapsulated and showed a pattern of cords and small columns of basal like cells. Prognosis was good and she survived 2½ years. Case No. 2 died in less than one year because of her highly malignant invasive tumour.

The poor prognosis of this tumour, despite its relative accessibility to detection and radical surgery, emphasizes the need for early diagnosis and many additional diagnostic procedures that may contribute to a better understanding of the pathological behaviour of this often fatal disease. Besides that in a community like Bahrain, the patients will be very reluctant to accept radical surgery such as exenteration of the orbit and that is an additional risk factor which could be minimised by health education programme.

ACKNOWLEDGEMENT

We wish to thank our colleagues at the Department of Ophthalmology, Salmaniya Medical Centre and Prof. F. Blodi Chairman of the Dept. of Ophthalmology, IOWA University, U.S.A.

REFERENCES

1. Henderson : — 430 — 436 (Edition 1973); Orbital tumours by Henderson.

2. Vermeer R.J., Minedo H.M.; Partial remission of advanced adenoid cystic carcinoma obtained with adriamycin; Cancer 43 (5) : 1604 — 1606. May 1979.

Calvin L Miller et al.; Adenoid Cystic Carcinoma of the entrum and epiphora; Am. J. of Ophth. Vol. 83, No. 4, 582 — 586. April 1977.

3. Richard L I Dallow MD.; Contraversy in Ophthalmology. 326 — 327. 1977.

4. Font R.L., Gamel J.W.; Epithelial tumours of the lacrimal gland : Analysis of 265 cases in ocular and adeneval tumours; Birmingham, Ala., Aesculapins Publishing Co. 1978. 787 — 807.

5. Ghassau Dagher MD., Richard L Anderson. MD., Karl C. Ossoining. MD., John D. Baker MD.; Adenoid Cystic Carcinoma of the lacrimal gland in a child Arch. of Ophthalmology Vol. 98. 1098 — 1100. June 1980.

6. Eby. L.S., Johnson P.S. and Baker H.W.; Adenoid Cystic Carcinoma of head and neck. Cancer 29 — 1168. 1972.

7. Persin K.H.. et. al.; Adenoid cystic carcinoma arising in the salivary glands. Cancer 42 (1) : 265 — 282. July 1978.

H.E. Tariq Almoayed, the Minister of Information with Dr. Jaffer Al-Bareeq, President of the Bahrain Medical Society presenting the Research Award to Dr. Fayeq Al-Hilli on 10th December, 1981.