CASE PRESENTATION

Orbital Rhabdomyosarcoma In a Bahraini Child Report of a Case and Review of the Literature

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ABSTRACT

A case of orbital rhabdomyosarcoma (RMS) in a Bahraini male child was detected in the early stage and confirmed by biopsy. The initial treatment was chemotherapy and radiotherapy, followed by exenteration for the recurrence. The patient survived for three years and three months and eventually died. The details of histopathology, chemotherapy, radiotherapy and review of literature are presented in this article.

In children rhabdomyosarcoma of the orbit is the most common primary mesenchymal malignancy of the orbit. It is a rare tumour and accounts for only 9% of the orbital tumours in children[1]. Here we report the first case in our 15 years retrospective studies of orbital tumours in Bahrain.

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CASE REPORT

A.M.Y. an 8 year old boy first attended the eye clinic on 9th February '81 for a painless swelling under the left upper lid of two week's duration. Clinical examination revealed a swelling in the superotemporal quadrant of the left orbit with downward displacement of the eyeball (Fig. 1). The ocular movements were not restricted. The visual acuity was 6/6 in each eye. The swelling was cystic to palpation and had the size of a marble. There were no signs of inflammation.

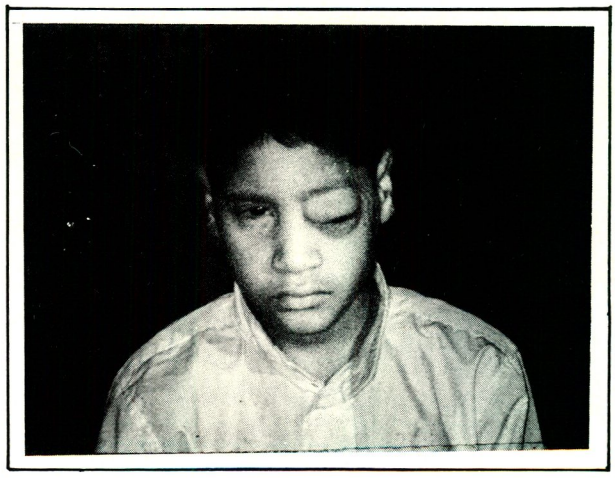

Fig. 1 : *Swelling in the orbit with downward displacement of the eyeball.*

Excision biopsy was attempted within the same week but the mass being situated posteriorly in the orbit could not be excised completely. It was a cystic mass measuring about 17 x 20 mm, was dark bluish and fragile. Histopathology reported a RMS of embryonal type showing diffusely infiltrating myxoid tissue composed of spindle, oval, round and bizarre giant cells with pleomorphic large hyperchromatic nuclei (Fig. 2). The cytoplasm was frequently drawn out in long processes at places like ribbons (Fig. 3) which with PTAH (phosphotungstic acid haematoxylin) stain, occasionally revealed ill-defined cross-striations.

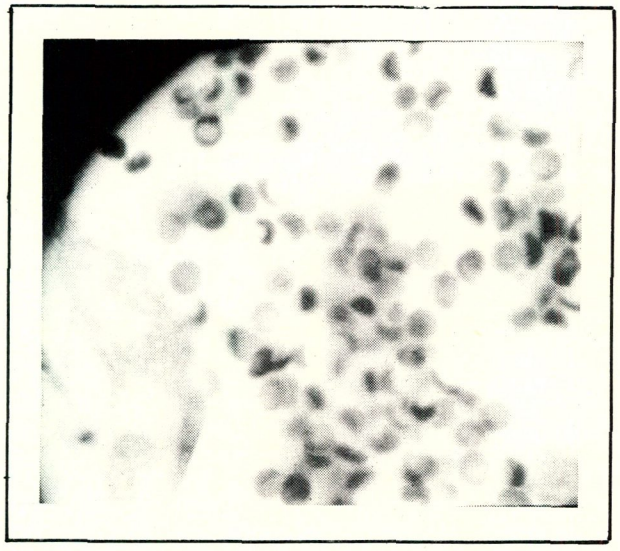

Fig. 2 : *Malignant cells with pleomorphic large hyperchromatic nuclei.*

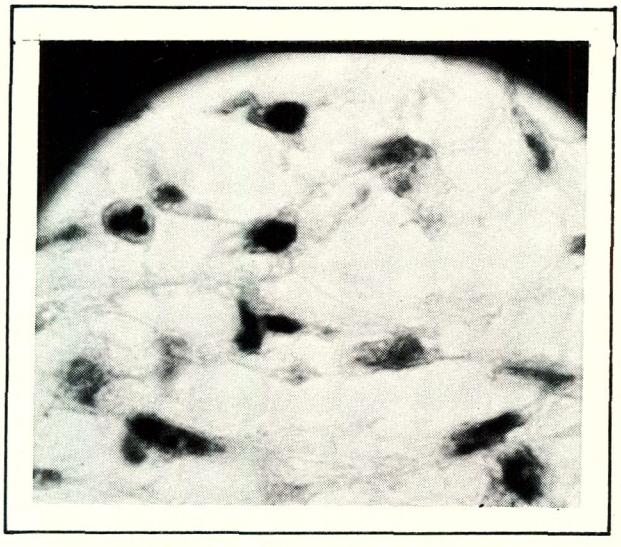

Fig. 3 : *Ribbon like processes of cytoplasm with cross striations*

Considering the seriousness of the disease and because of the incomplete removal of the tumour mass, the patient was referred to a specialised cancer centre for radiotherapy and chemotherapy in Kuwait as these are not available at Salmaniya Medical Centre.

Further investigations, including CT Scan, confirmed the presence of a residual tumour mass in the orbit. Radiotherapy was initiated on 5th April'81 giving total 50 Greys (5000 rads) over a five week period in two-wedged fields — anterior and lateral, followed by chemotherapy courses one month later. The VAC (Vincristine — Actinomycin Cyclophosphamide) regime was started on 7th May '81, every three weeks for 18 months and was monitored by platelet and white cell counts. The patient remained free from local recurrence for 20 months when he presented again with a mass in the left orbit and with a mobile enlarged lymphnode in the left axilla.

He was sent again to Kuwait where exenteration of the orbit was performed on 16th Oct. '82 and a pedicle graft was fashioned to cover the bare area (Fig. 4). The histopathology once again corroborated the diagnosis. However, seven months later (May '83) he was found to have multiple nodular swellings

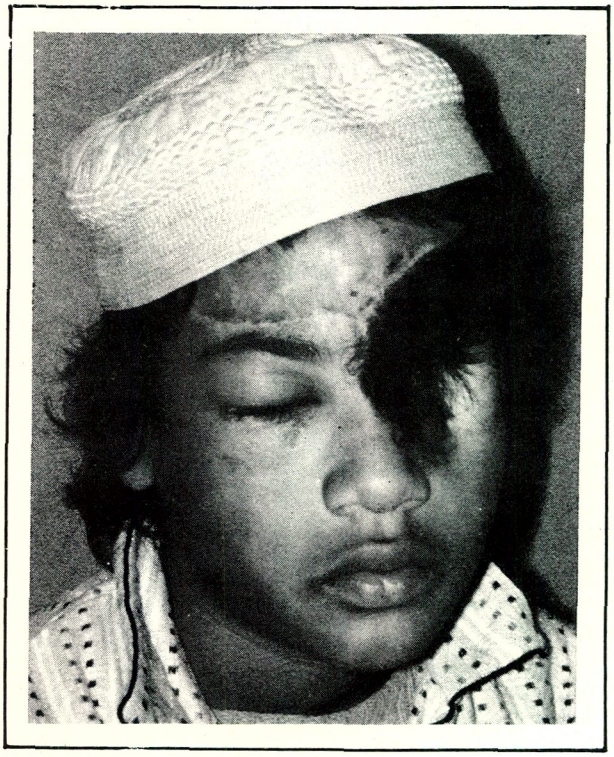

Fig. 4 : *Pedicle graft to cover bare area.*

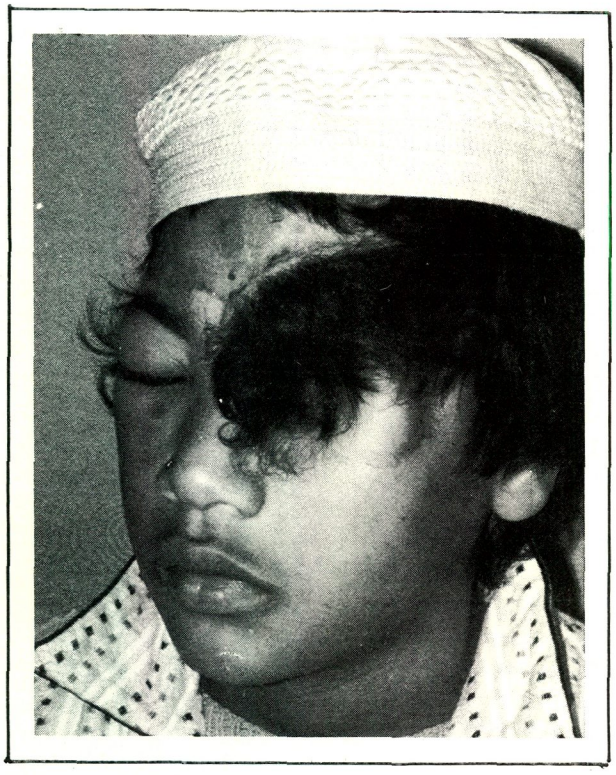

Fig. 5 : *Recurrent tumour masses under the graft with spread to the other side of the face.*

under the graft (Fig. 5). The recurrent masses were excised and regrafting was done which kept him free from further recurrence for four months. Then he showed up again in Sept. '83 with a huge recurrence for which nothing further could be done as the prognosis was very poor. Two months later a bone marrow biopsy was performed which revealed no malignant infiltration. The chemotherapy was continued at monthy intervals. On 6th Jan. '84 he had profuse bleeding from the tumour mass following a trivial trauma. His general and mental health deteriorated fast. Chemotherapy was given, but the tumour went on increasing enormously spreading to the other side of the face and encroaching upon the nose. It kept oozing foul-smelling blood and the patient became severely anaemic necessitating a blood transfusion.

An attempt to excise the mass was made on 4th May '84, but the patient remained in coma following the surgery and died two weeks later.

DISCUSSION

Reviews and literature dating a decade back ascribed to RMS a "high degree of malignancy with poor prognosis"[2]. With increasing knowledge on the biologic behaviour of the malignancy, the therapeutic approach to the disease has changed and greatly increased the disease-free interval for the patient and resulted in a significant increase in cure rate.[3]

The most frequently used modalities at the present time are surgical excision, radiotherapy and chemotherapy[5] — each separately or in a combination of any two or even all three.

Balestrazzi et al.[5] from their wide experience concluded the protocol for this lethal tumour to be : immediate biopsy, immediate irradiation with sufficient high doses, exenteration of the orbit only for a recurrence, followed by irradiation and/or perfusion and/or systemic chemotherapy.

Sagerman et al.[1] from the Columbia group demonstrated the efficacy of primary radiation therapy, showing 71% of 45 patients alive after a mean follow up period of two years.

Manual Lederman and Kenneth Wyber reported a series of 29 cases[6]. They followed a protocol of primary irradiation as an initial treatment retaining the eye ball, and in cases of local recurrences, they used implantation of radio-active material. Out of 29 cases only 12 patients survived for varying periods of time. Pinkel and Pickren[7]. demonstrated successful results of chemotherapy with surgery and radiation. Pratt et al.[8] suggested the use of combination chemotherapy. Ghavimi et al.[9] in reviewing a series of 29 patients adapted a protocol of surgical excision of the primary lesion, if possible, radiotherapy in patients with gross or microscopic residual disease followed by chemotherapy for two years. 24 of 29 patients were alive with no evidence of disease for 4-42 months after diagnosis.

A unilateral proptosis rapidly developing in children usually gets the ophthalmologist alarmed, especially in view of the possibility of RMS. The majority of authors on the subject have reported a slight predilection of this tumour among male children with no hereditary incident[4,5,10]. The average age of onset is reported to be about 6 years with the majority less than 10 years. It is found to be extremely rare after 25 years of age[11], although Ellenbogen et al. have reported a case of orbital RMS in a newborn infant[12] and Kassel and colleagues[13] described a RMS intimately related to the extrinsic ocular muscles in a 78 year old man.

The untreated neoplasm is fatal[4]. The embryonal and alveolar types are the most aggressive

variety and since they are predominant in childhood, the disorder seems more alarming and progressive in this age group. In fatal cases the average period of survival is about 18 months after treatment. Before the advent of contemporary combined chemotherapy and radiotherapy the two year survival rate for children with RMS orbit was 40%[14]. If local recurrence or distant metastasis occur, it usually appears within the first two years of treatment[14]. Ellsworth[15] noted two late recurrences, 3 and 7 years after radiotherapy amont the 45 children in the Columbia study. The lungs are the most common site for metastasis. Our patient had palpable lymphode in the axilla on the same side as the first sign of remote metastasis.

Nonetheless, there is a close similarity in regard to age and sex between our study and many of the major studies on RMS published in literature on the subject. Our patient was a male child in whom the malady was detected at the age of 8 years, he had no history of similar problems in the family.

RMS, usually first diagnosed in a young healthy individual, may be preceded by a history of trauma[4]. According to the study of E. Ballestrazzi et al. from the University of Rome, 15% of RMS arise in the orbit, clinically present as a small swelling in the lid, usually situated in the supero-internal quadrant of the orbit which grows rapidly causing a painless, irreducible unilateral proptosis[5]. Ptosis and epiphora may be present in the initial stages[10]. The tumour can develop anywhere in the orbit but it is most commonly situated behind or above the globe. In the case of our patient the swelling was situated in the supero-temporal quadrant and was observed for two weeks. There was a downward displacement of the eyeball with no restriction to the movement of the affected eye.

Routine X-ray studies in all these cases are seldom diagnostic or at the most they may show an evidence of a nonspecific increase in soft tissue density[4]. In our case, X-rays of the orbit did not reveal any diagnostic clue.

The diagnosis of RMS should be suspected in all children with a rapidly developing exophthalmos, because RMS is a histologic diagnosis. Therefore, early excision biopsy is mandatory[14]. The excision biopsy may not be feasible in all cases as it depends upon the accessibility of the tumour mass. A posteriorly extending mass may not be excised completely as happened in our case.

In the past 10 years numerous reports have been published using a combination of surgery with radiotherapy, multidrug chemotherapy or both. Thus disease-free survival has greatly increased which represents an important advance in treatment.

In our study the patient underwent primary surgical excision which left a residual tumour mass. He received high doses of radiotherapy immediately followed by chemotherapy for 18 months. Later an exenteration was performed and chemotherapy (VAC regime) was initiated. The patient survived for 39 months from the time of diagnosis.

REFERENCES

1. Harley RD. Paediatric Ophthalmology. 2nd ed. Philadelphia : W B Saunders, 1983; 1252-1256.

2. Jones IS, Reese AB, Krout J. Orbital RMS : An analysis of 62 cases. Trans Am Ophth Soc 1965; 223.

3. Bizer LS. Rhabdomyosarcoma. Am J Surg 1980; 140 :687 - 691.

4. Henderson JW. Orbital tumours. Philadelphia : W B Saunders, 1973; 270 - 284.

5. Balestrazzi E, Scorcia G, Moreno F, Araia F. University of Rome. Proceedings of the 2nd meeting of the International Society for Paediatric Ophthalmology, 1979 : 83 - 91.

6. Lederman M, Wyber K. Proceedings of the Royal Society of Medicine, 1976; 895 - 903.

7. Pinkel D, Pickren J. Rhabdomyosarcoma in children. J Am Med Assoc 1961; 175 : 4 : 293 - 298.

8. Pratt CB, James DH Jr, Holton CP, Pinkel D. Combination therapy including vincristine (NSC 67574) for malignant solid tumour in children. Cancer chemotherapy report 52, 1968; 489 - 495.

9. Ghavimi F, Exelby RP, D'Angio GJ, et al. Multidisciplinary treatment of embryonal RMS in children. Cancer 1975; 35 : 677 - 686.

10. Gardner A, Klintworth G. Pathobiology of ocular disease: A dynamic approach. New York : Dekker, Marcel Inc., 1982; 757 - 763.

11. Reese AB. Tumours of the eye. 3rd ed. New York: Harper and Rowe, 1976; 538 - 539.

12. Ellenbogen E, Lasky MA. Rhabdomyosarcoma of the orbit in the newborn. Am J Ophthalmol 1975; 80 : 6 : 1024 - 1027.

13. Kassel SM, Copenhaver R, Areen VM. Orbital RMS. Am J Ophthalmol 1965; 60 : 811 - 818.

14. Nelson LB. Paediatric Ophthalmology. Philadelphia : W B Saunders, 1984; 213.

15. Ellsworth RM. Rhabdomyosarcoma of the orbit. In : New Orleans Academy of Ophthalmology symposium on surgery of the orbit and adnexa. St. Louis : C V Mosby Co., 1973; 41 - 47.