RHABDOMYOSARCOMA OF THE UPPER RESPIRATORY TRACT

By Thomas Sim

SYNOPSIS

Rhabdomyosarcomas are rare and highly malignant tumours which arise from rhabdomyoblasts. It was only recently that their classification into 4 histo-pathological types has been agreed upon i.e. embryonal, botyroid, alveolar and pleomorphic. The first three types occur mainly in children and young adults, while the fourth (pleomorphic) type occurs in older adults.

Three cases of rhabdomyosarcoma seen recently in the Ear, Nose and Throat Department, Outram Road General Hospital are described. Two patients aged 2 and 3 years had embryonal rhabdomyosarcoma in the nasopharynx with local extension into the nose and retropharyngeal space. The third patient aged 31 years had an alveolar rhabdomyosarcoma in the right nasal passage. All three cases had local cervical metastases, while one also had hilar secondaries. Despite radical surgery combined with radiotherapy or chemotherapy, the prognosis is very poor. All the three patients were dead within six months of the initial presentation.

INTRODUCTION

Rhabdomyosarcomas are highly malignant tumours of rhabdomyoblasts with a microscopic pattern similar to that of embryonic striated muscle cells. They are very uncommon tumours which have only recently been classified and their histo-pathological features worked out and agreed upon. In 1946 Stout delineated their histo-pathological characteristics making it possible to classify certain tumours in this category with assurance despite an inability to demonstrate cross-striations.

Further work by Stobbe and Dargeon (1950), Horn and Enterline (1955) and Riopelle and Theriault (1956) has enabled rhabdomyosarcomas to be classified into four distinct types:—

(*a*) Embryonal
(*b*) Botyroid
(*c*) Alveolar
(*d*) Pleomorphic

While rhabdomyosarcomas are still uncommon tumours, it is now realised that they occur much more frequently than was realised in the past. This is because pathologists are now aware that cross or longitudinal striations are often found only after prolonged and painstaking search and are not essential to the diagnosis.

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Three cases of rhabdomyosarcoma seen recently in the Ear, Nose and Throat Department, Outram Road General Hospital are herewith described to illustrate their features and behaviour pattern.

CASE REPORTS

Case 1

The patient (S.K.H.) was a three year old female Chinese who was referred to the department with a two weeks' history of cough and fever associated with progressive swelling of the right side of the neck. On examination she was febrile and anaemic. The right eye was proptosed. The right nasal passage was filled with a fleshy mass and the soft palate was depressed on the right side by a tumour in the nasopharynx. There was an enlarged hard and fixed lymph node, 5 cms. by 10 cms. in the right upper cervical region (Fig. 1).

X-ray of the chest showed increased nodular markings in both peri-hilar regions and the right hilum was enlarged. X-ray of the nasopharynx showed a soft tissue tumour occupying the entire nasopharynx (Fig. 2).

An examination under general anaesthetic was carried out revealing a friable, haemorrhagic and necrotic tumour mass in the right side of the nasopharynx extending into the right nasal fossa. Biopsies were taken from both regions.

Histology of the sections was reported as "a highly cellular anaplastic sarcoma, with positive stain for myofibrils. Appearances are those of embryonal rhabdomyosarcoma" (Fig. 3).

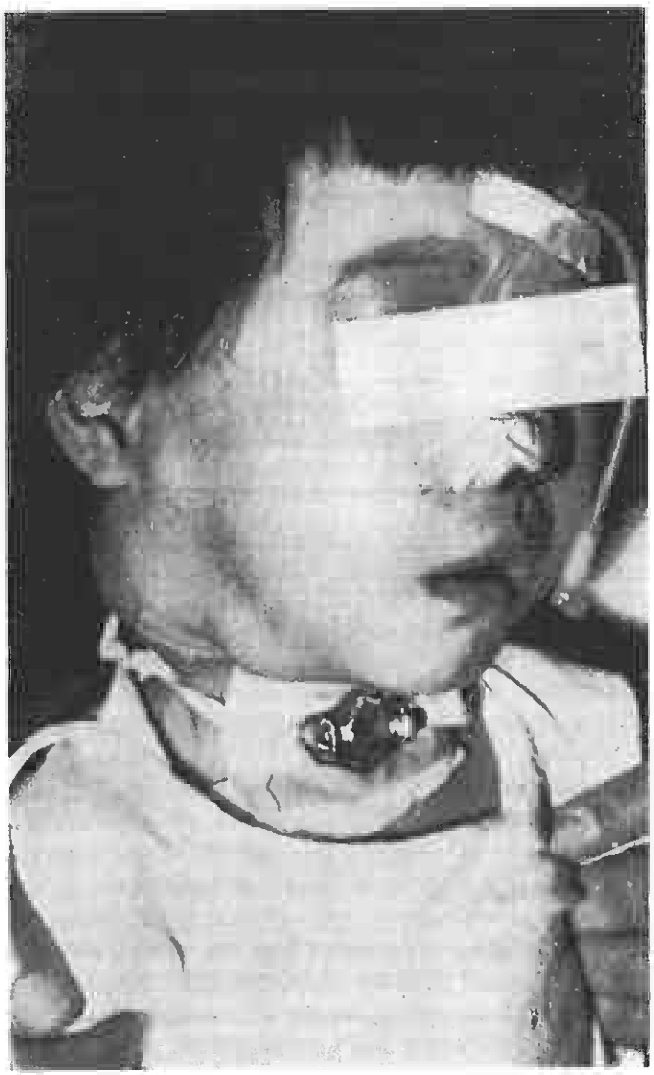

Fig. 1. Case 1 (Patient S.K.H.) with right proptosis and cervical metastases. Post-operative photograph also showing tracheostomy.

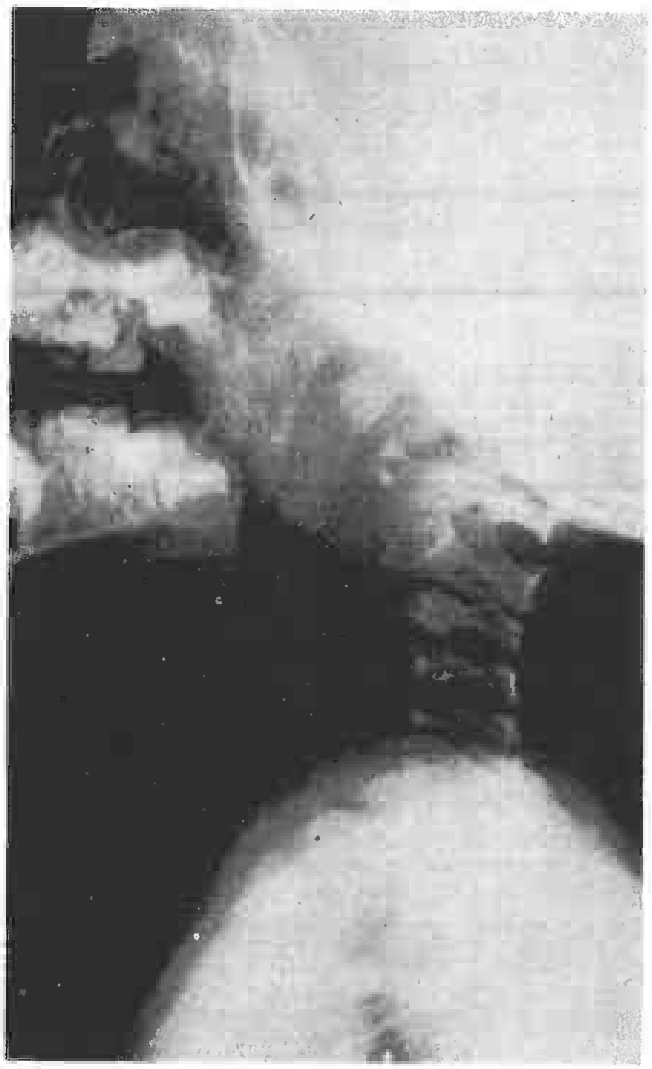

Fig. 2. Lateral X-ray of the neck of Case 1 showing soft tissue tumour occupying nasopharyngeal space.

Diagnosis: Embryonal rhabdomyosarcoma of the nasopharynx and right nasal fossa with lymph node metastases in the right upper cervical and hilar regions.

Post-operatively she developed acute respiratory distress and an emergency tracheostomy was performed.

She was started on Telecobalt irradiation but her general condition deteriorated and she died three weeks after commencement of the radiotherapy.

Case 2

The patient (T.W.) was a two year old male Chinese who was referred to the department with a three months' history of cough associated with breathlessness. On further questioning, it was learnt that he had been previously treated by an orthopaedic surgeon for "torticollis" first noticed when he was four months old. The mother found that the child always held his head in a fixed forward position and refused to move it. He also lost weight over the previous six months and was noticed to be generally weak and lethargic. No abnormality was found by the orthopaedic surgeon.

On examination in our department, a tumour mass was found occupying the entire nasopharynx and extending down into the oropharynx and hypopharynx. An X-ray of the chest showed no lung lesion, while a lateral X-ray of the neck outlined the tumour mass (Figs. 4 and 5). The routine blood and urine examinations revealed no abnormalities except for an ESR of 26 mm. per hour.

An examination was done under general anaesthetic. A very firm, sessile tumour extending from the base of the skull to obliterate the entire nasopharyngeal space and extending down to the hypopharynx was found. The tumour was mainly sub-mucosal. A biopsy was taken from the mass.

The histology showed "a tumour of extremely anaplastic cells. Some of the cells have abundant

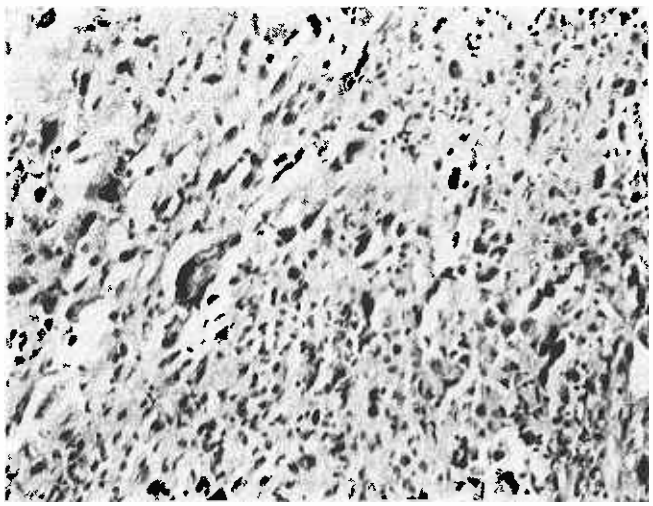

Fig. 3. Section shows anaplastic tumour cells lying in a fibrous stroma. Nuclei are bizarre and some cells have eosinophilic cytoplasm (H & E × 150).

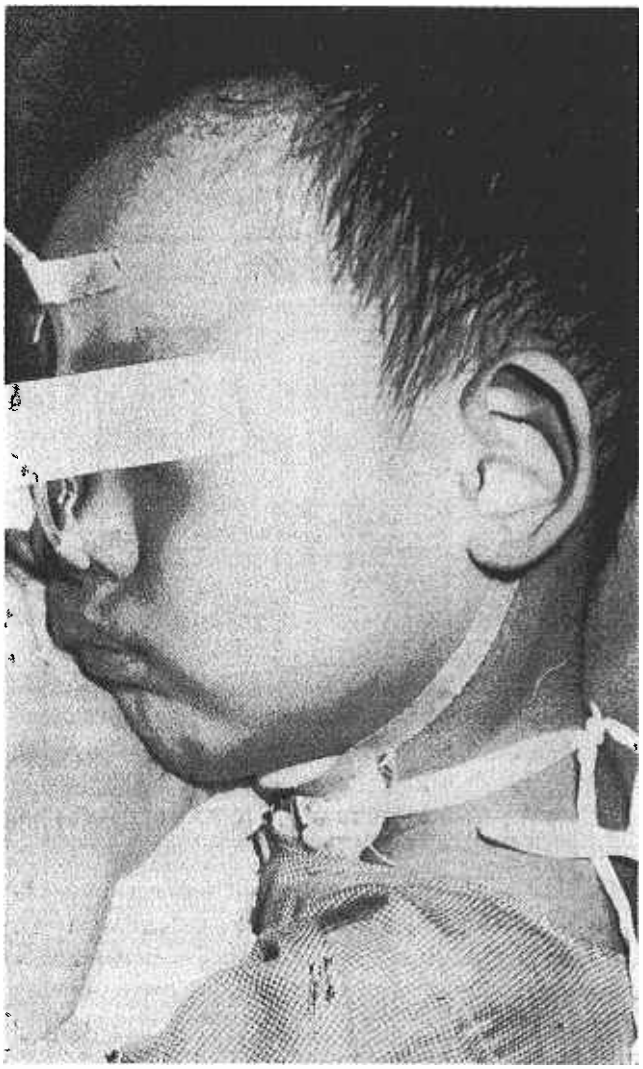

Fig. 4. Case 2 (Patient T.W.). Post-operative photograph showing left cervical metastasis. A tracheostomy has been done.

cytoplasm giving a positive stain for muscle sarcoplasm."

Diagnosis: Embryonal rhabdomyosarcoma of the nasopharynx.

Excision of the tumour was carried out a week after the biopsy. The tumour was found to be attached to the pre-vertebral fascia with no definite borders. A Ryle's tube was inserted for feeding purposes and an elective tracheostomy was also done. He was referred for post-operative radiotherapy, but this was deemed not possible because of his poor general condition, the extensive nature of the growth and because of the close proximity of the spinal cord.

His general condition gradually worsened, the tumour mass recurred together with cervical metastasis and he also developed a left brachial plexus paralysis. He died of chest infection and malignant cachexia about four months after the operation.

Case 3

The patient (Y.K.C.) is a 31 year old male Chinese adult, who complained of right-sided nasal obstruction and epistaxis for three months. He also noticed a swelling on the left side of the neck. On examination the right nasal passage was found to be filled with a granular and fleshy tumour. There was some purulent discharge on the right side of the nasopharynx. There were bilateral enlarged submandibular lymph nodes (Figs. 6 and 7).

An examination of the nose was carried out under general anaesthesia and excision of the right submandibular lymph node was also done. A biopsy was taken from the tumour.

The histology report was as follows: "the tumour is composed of alveolated areas of cells which are mostly small with some eosinophilic cytoplasm. Some of the cells are multi-nucleated and large with abundant cytoplasm. The PTAH stain has brought out longitudinal myofibrils as well as cross-striations. The picture is that of alveolar rhabdomyosarcoma" (Figs. 8, 9 and 10).

X-rays of the sinuses showed opacity of the right ethmoid and antrum with ill-defined bony destruction (Fig. 11). X-ray of the chest was clear.

He was given a full course of Telecobalt irradiation. He was readmitted three months after completion of radiotherapy complaining of weakness and breathlessness. He was found to be severely anaemic and there was a left pleural effusion. The haematological picture showed an aplastic anaemia (Hb = 3·2 gm.%, Total white = 2,300 per c.mm. and Platelet count = 5,000 per c.mm.). He was transfused with packed cells but passed away during the transfusion.

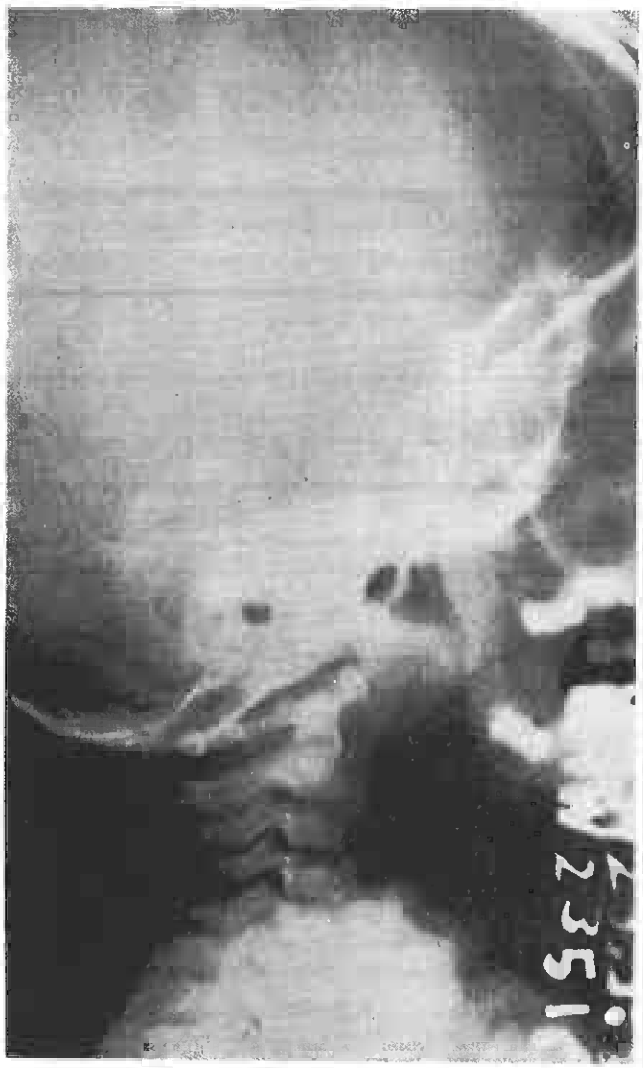

Fig. 5. Case 2. Lateral X-ray of the neck showing tumour mass in the nasopharynx.

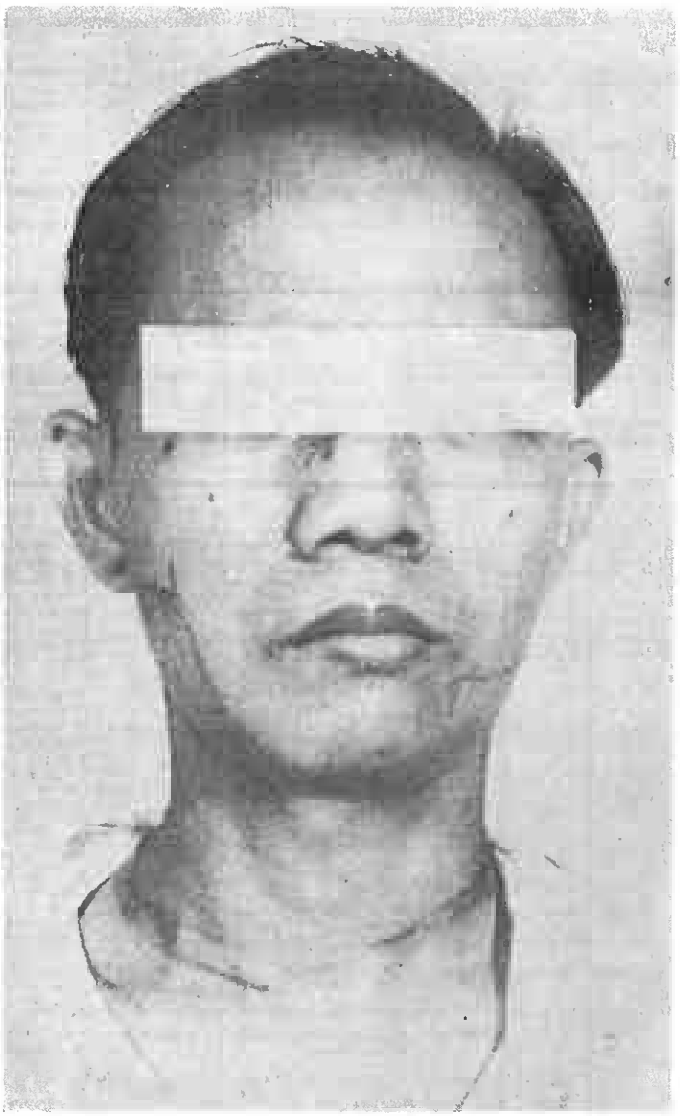

Fig. 6. Case 3 (Patient Y.K.C.). Photograph showing bulge on the right nasal region and right submandibular metastasis. Note the irradiation marks.

DISCUSSION

As has been suggested by Horn and Enterline (1958) and now accepted by most authorities, rhabdomyosarcomas can be classified into four distinct types:—

(a) *Embryonal Rhabdomyosarcoma*

This was first described by Stobbe and Dargeon in 1950 as a series of cases not fitting Stout's original description, but which are also undoubted examples of rhabdomyosarcoma. They are the commonest group to be found in the head and neck and occur mainly in children. The tumour consists of spindle-shaped cells arranged in interlacing and parallel bundles and at times resemble syncytial masses. Cross-striations are difficult to find and are not essential to the diagnosis.

(b) *Botyroid Rhabdomyosarcoma*

In 1955, Horn *et al* first reported cases of polypoid or botyroid rhabdomyosarcomas, which are tumours morphologically identical with the better known botyroid sarcomas of the uro-genital tract. It is now agreed that they are a morphological variant of the embryonal type, assuming the grape-like or polypoid pattern of growth when they take their origin just beneath the mucous membrane of a hollow viscus or body cavity. The cambium layer of Nicolson, a neoplastic cellular zone beneath the epithelium, provides ease of recognition. Striations are again rare.

(c) *Pleomorphic Rhabdomyosarcoma*

This is the type originally described by Stout in 1946. It is cellular and as its name implies, pleomorphic. "Strap" cells, cells with nuclei in tandem and rhabdomyoblasts with longitudinal striations are usually found.

(d) *Alveolar Rhabdomyosarcoma*

This peculiar but distinctive type having an alveolar pattern and sometimes simulating carcinoma, has been recently identified by Riopelle

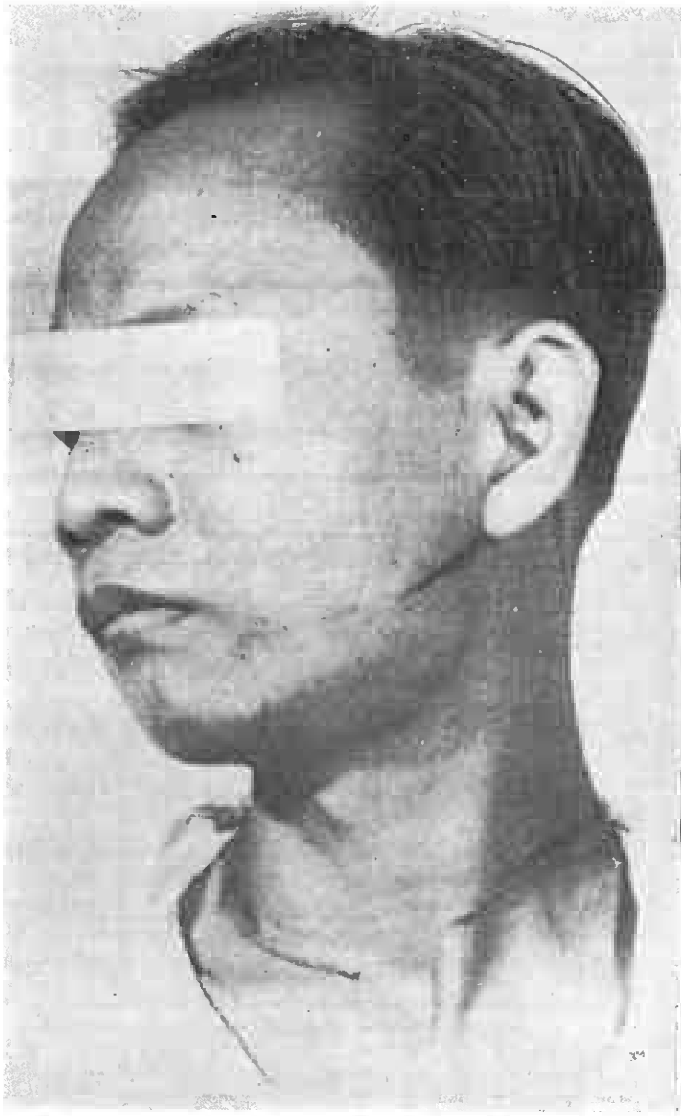

Fig. 7. Case 3. Lateral view showing the left submandibular metastasis.

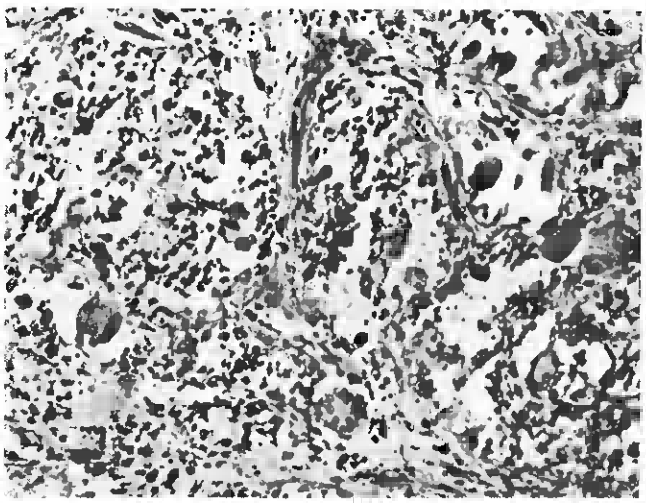

Fig. 8. Case 3. Histological section shows rhabdomyoblasts of varying sizes enclosed by fibrous trabecullae giving as alveolar pattern. Note the tumour giant cells with abundant eosinophilic cytoplasm (H & E × 150).

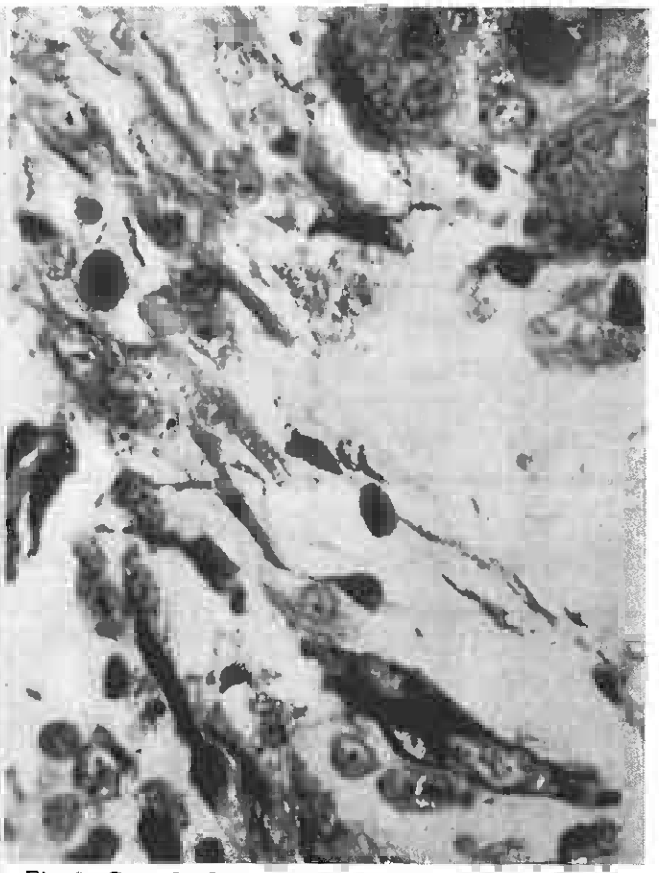

Fig. 9. Case 3. Section shows cross-striations in the cytoplasm of three rhabdomyoblasts (P.T.A.H. × 500).

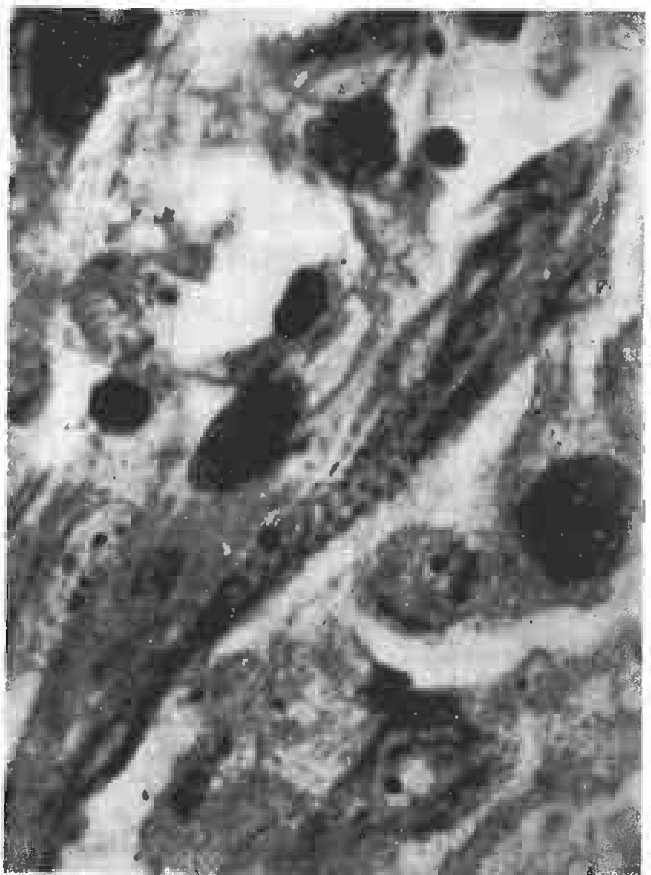

Fig. 10. Case 3. High power view of a rhabdomyoblast with cross-striations seen as continuous dark dots (P.T.A.H. × 1,100).

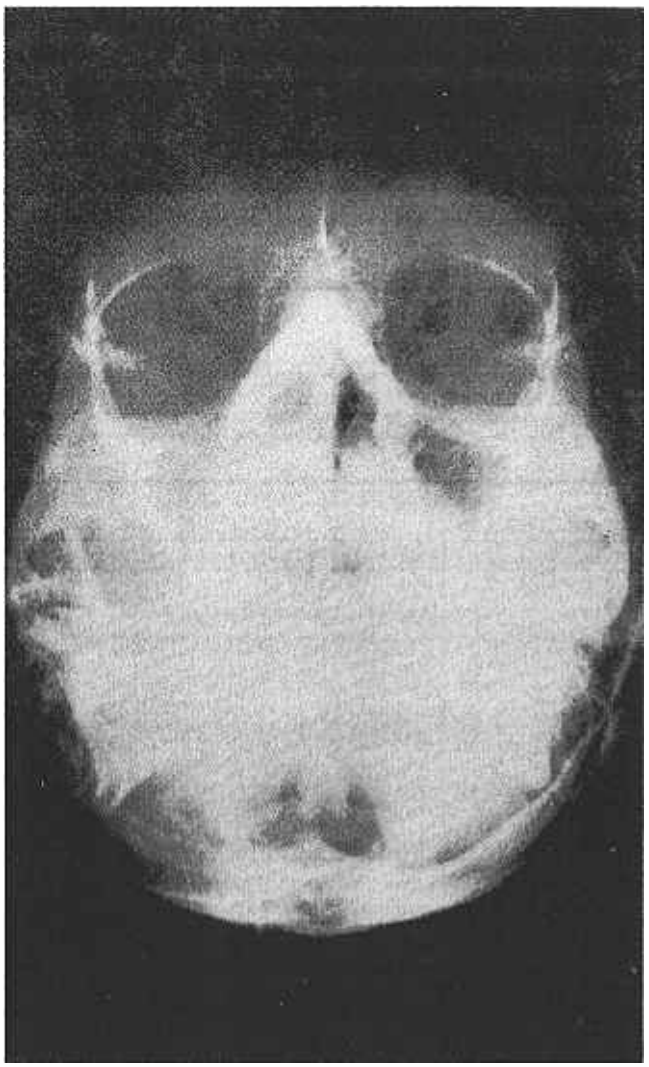

Fig. 11. Case 3. X-ray of the sinuses showing opacity of the right antrum with ill-defined bony destruction.

and Theriault in 1956. Microscopically the tumour is characterised by nests of cells separated by fibrous trabeculae. Some of the cells in the alveolar nests may be striated.

Of the three cases described, Cases 1 and 2 were of the embryonal type, while Case 3 was of the alveolar type. The age group of patients with rhabdomyosarcoma ranges from birth to the eighth decade, but there is a correlation between the tumour type and the age group. Thus the pleomorphic type occurs mainly in adults, the alveolar type usually among adolescents and young adults (Case 3 was 31 years), while the embryonal and botyroid varieties occur mainly in children and even infants (Cases 1 and 2 were aged 3 and 2 years respectively).

Pack and Eberhart (1952) first suggested separating rhabdomyosarcomas into those arising from the visceral tissues and those arising from the peripheral soft tissues, for it is found that the pleomorphic type arises largely from the peripheral tissues e.g. in the limbs, while the other three types arise mainly from the viscera. In the head and neck there are but few reports of rhabdomyosarcomas arising from the upper respiratory tract—Reitter (1921), Cooper (1934), McCuaig (1952), Allen (1960), Vieta *et al* (1962), Dito and Batsakis (1963), Shirasawa and Taguchi (1963), Masson and Soulle (1965) and Williams *et al* (1968). Williams *et al* (1968) found that they comprise only 3·3% of malignant tumours in this region. Cases 1 and 2 had tumours in the nasopharynx with local extension, while Case 3 had a nasal tumour.

Rhabdomyosarcomas have been reported mainly in the Caucasians and are rare in the Negroes and Mongoloids. However the three cases reported here were in Chinese (Mongoloids).

The clinical presentation of these tumours varies with the site of origin, the commonest being that of a painless mass. In our cases there were nasal symptoms e.g. epistaxis, nasal obstruction and discharge and throat symptoms e.g. breathlessness, dysphagia, sore throat, cough and expectoration. As they are highly malignant tumours, rhabdomyosarcomas have a marked propensity for local extension and metastatic spread, the latter both by haematogenous and by lymphogenous dissemination. The most common sites for distant metastases are the lungs and bones. Regional lymph nodes are frequently affected (33% incidence reported by Masson and Soulle (1965), 50% by Dito and Batsakis (1963); and all three cases in this report). Distant lymph node metastases e.g. in Case 1 have also been reported.

Patients with rhabdomyosarcoma have a poor prognosis, even with modern methods of treatment. Various combinations of surgery, radiotherapy and chemotherapy have been tried out by various centres. The five-year survival figures reported by various authors is in the region of 10-20%. Thus Dito and Batsakis (1963) had a 12·2% (6 cases out of 49) and Masson and Soulle (1965) had a 21·6% (19 cases out of 88) five-year survival rate. The three cases reported here all died within about six months of the diagnosis. The best chance for survival would appear to be an early and accurate diagnosis, followed by wide surgical excision of the primary tumour with regional block dissection of the lymphatic field. This may be followed by post-operative radiotherapy. Any recurrence should be treated by chemotherapy e.g. Actinomycin D, Cyclophosphamide or Vincaleukablastine with irradiation.

ACKNOWLEDGEMENTS

I wish to thank Dr. Andrew Chew, Medical Superintendant, Outram Road General Hospital for his kind permission to publish the cases.

I also wish to thank Dr. Tan Kheng Khoo, Dr. Ong Beng Hock and Mr. Tan Tee Cheok of the Pathology Department for their guidance and assistance in the production of the photographs and the description of the histological slides.

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