

METASTATIC MALIGNANT SCHWANNOMA OF ORBIT: A CASE REPORT

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SUMMARY

A case of metastatic malignant schwannoma of the right orbit is presented here. Although the patient had disseminated disease, it was the eye problems which were striking and most symptomatic.

The orbital lesion was at first thought to be the primary tumour until a history of an excised right wrist mass was probed into and the histology traced.

This report is written as malignant schwannoma is an uncommon tumour. The difficulty in diagnosing malignant schwannoma is mentioned. The importance of diagnosis of an orbital metastatic lesion is also pointed out as this is often misdiagnosed.

INTRODUCTION

Schwannoma or neurilemmoma arises from schwann cells which have their embryonal origin in the neural crests. The tumour can occur any-

where. Malignant transformation is rare and such a change is more apt to occur in neurofibromatosis (10 – 15%).¹

Malignant change is indicated clinically by accelerated growth in a previously quiescent tumour, multiple recurrences after excision and the appearance of distant metastasis. Microscopically, malignancy is indicated by increasing cellular anaplasia, increased mitotic activity, infiltration into adjacent tissues and the progressive loss of those distinctive features of cell form and arrangement which indicate a nerve origin.

This is the only reported case of metastatic lesion in the University Hospital, Kuala Lumpur since 1968.

CASE HISTORY

A 35-year-old Chinese male had a swelling on the right wrist for more than 10 years. As the swelling grew bigger, he had it excised in late 1984 at the Kuala Lipis District Hospital. There was local recurrence of tumour after a few months. The patient did not have any follow-up at the Kuala Lipis District Hospital. The histology of the tumour was traced and it was reported to be synovial sarcoma.

On 17 July 1985, the patient was referred to the Universiti Kebangsaan Malaysia's (UKM) Eye Department for diplopia, blurring of vision and mild discomfort in the right eye of about one

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month duration. There was a 3mm proptosis and downward displacement of the right eye. The visual acuity was R6/36 and L6/6. Movements were limited in all directions. The conjunctiva was chemotic with subconjunctival haemorrhages and the fundus showed papilloedema. The right wrist scar and recurrent swelling were noted. The epitrochlear, cervical and axillary lymph nodes were palpable.

The CT scan of the right orbit showed a soft tissue mass in the supero-lateral quadrant of the orbit with no intracranial extensions. The chest X-ray was noted to be clear (when reviewed later, this showed early pulmonary metastasis). The lymph node biopsy was reported as leiomyosarcoma but later this as well as the orbital biopsy were reported as inconclusive. The patient deferred from UKM before a repeat right orbital biopsy could be done to ascertain the correct diagnosis.

On 10 October 1985, the patient came to the University Hospital with gross proptosis, diminished vision and pain in the right eye. Physical examination showed the patient to be well-built and fairly well-nourished. There were no *cafe au lait* spots and no subcutaneous nodules anywhere. The epitrochlear, both cervical, right axillary and both inguinal lymph nodes were palpable. The right wrist showed an indurated scar and a tender, infected mass about 2 x 3 cm in size on the ventral aspect. The chest was normal to percussion and auscultation. There was no enlargement of the liver or spleen.

The right eye was grossly proptosed and the movements markedly restricted especially on elevation and dextroversion (Fig. 1). The left eye was normal. The visual acuity was R6/60 and L6/6. Diplopia was present in all directions. A mass, soft and cystic, could be felt in the upper and lower outer quadrants of the right orbit extending over the lateral wall of the right orbit.

There was ptosis of the right upper lid which was also oedematous. The conjunctiva was injected and the chemotic and the optic disc showed papilloedema.

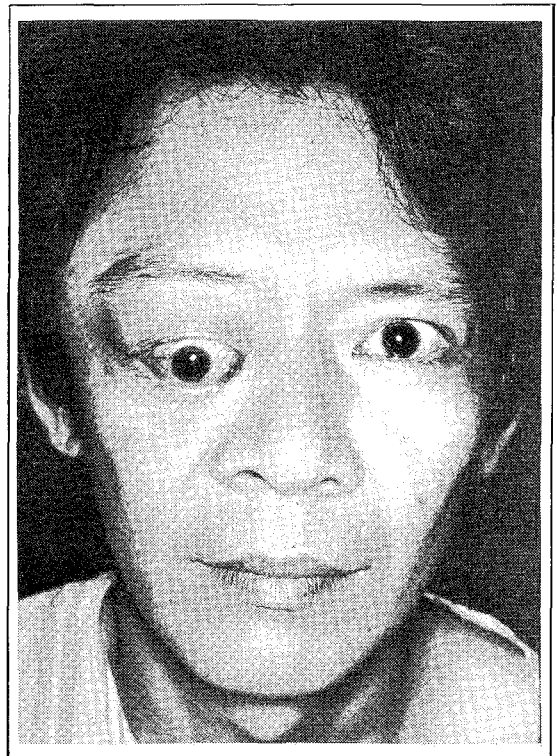


Fig. 1 The patient with gross proptosis—right eye.

X-ray of the right orbit showed an increase in opacity in the orbit with erosive widening of lateral and inferior walls. The CT scan showed the same mass extending into the anterior cranial fossa (Fig. 2). The chest X-ray showed a nodular mass in the right hilum and multiple shadows in both lung fields. The right wrist X-ray revealed a soft tissue swelling with calcification. The bone scan showed metastases in the skull and vertebrae. Biopsies of the right orbital mass, right axillary lymph node and the right wrist mass were done; all showed the same pathology — that of malignant schwannoma.

A diagnosis of metastatic malignant schwannoma of the right orbit was made. The primary was the right wrist mass.

Histopathology

The microscopic findings of the orbital tumour and the metastasis in the axillary lymph node were

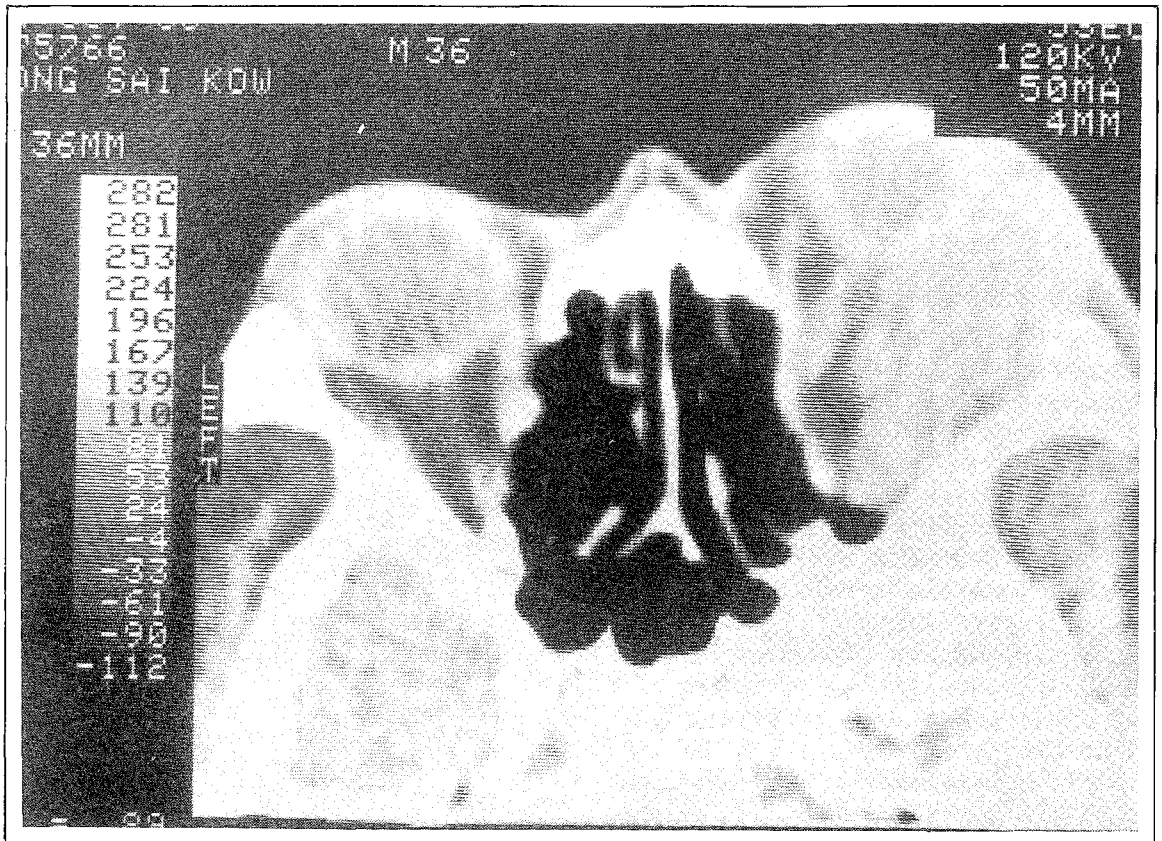


Fig. 2 C.T. Scan showing the right orbital mass.

essentially similar. The tumour cells were spindle or oval-shaped with hyperchromatic nuclei exhibiting frequent mitoses. In the orbital tumour nuclear pallisading and Antoni type A or B (cellular or acellular) areas were apparent. Microcystic formation and foamy histiocytes were seen scattered in areas. In the axillary metastases, the tumour virtually replaced the lymph node tissue and in addition to nuclear pallisading, whorl formation was readily discernible (Fig. 3).

Treatment

Palliative DXT was given to the right orbital mass. There was some reduction of tumour size and relief of orbital pain. Systemic antibiotic was given to control the infection of the right wrist. A course of chemotherapy was also tried in view of the patient's age. The patient's condition deteriorated. He developed pleural effusion

which was tapped off and on to relieve breathlessness. He died a few months later.

DISCUSSION

The diagnosis of malignant schwannoma on histological grounds may be difficult as the nerve tissue tumours cannot readily be separated from the various fibrosarcomas and leiomyosarcomas unless one can show that the tumour arises from a definite nerve tissue.² The difficulty is due to the fact that both schwann cells of nerve sheath and fibroblasts can form reticulin or connective tissue fibrils. The pallisading nuclei in malignant schwannoma is also characteristic of leiomyosarcoma, leiomyoma and fibroma. Under the electron microscope, the cellular characteristics of schwann cells include long spacing collagen, which is again not pathognomonic of nerve tissues. Hence the right wrist tumour of our patient was

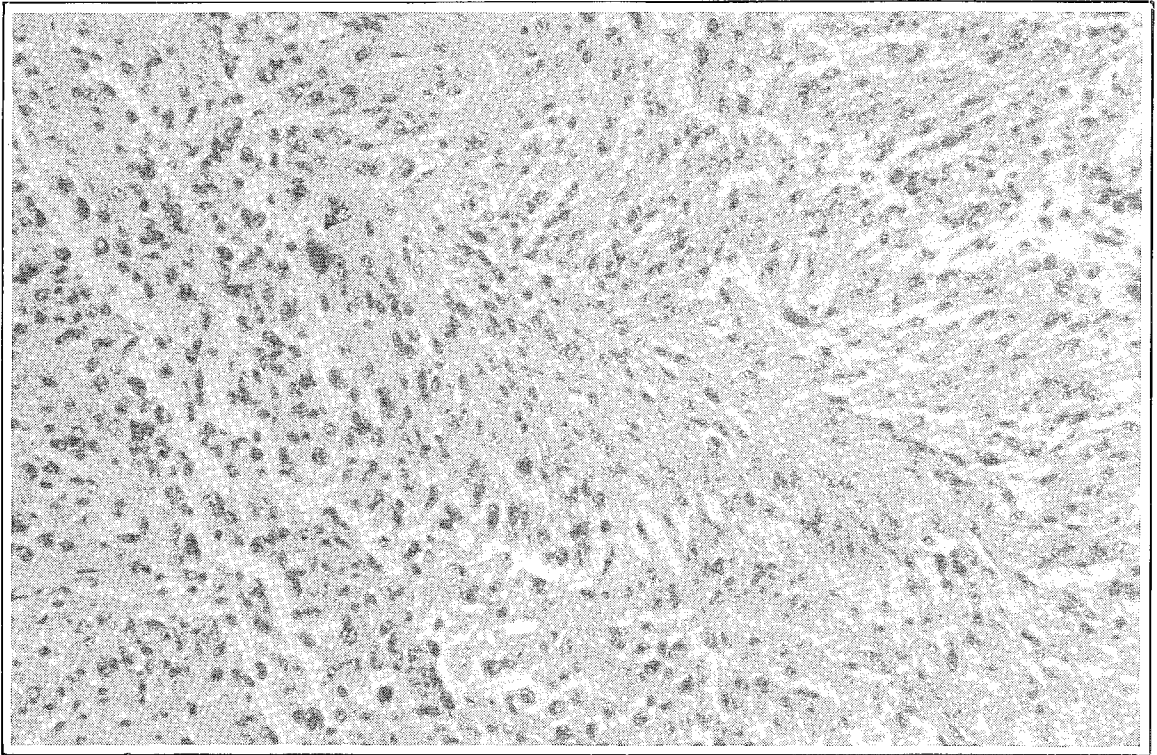


Fig. 3 A storiform pattern with nuclear palisading. Nuclei are oval or spindle-shaped and a mitotic figure is seen left of the centre.

diagnosed as synovial sarcoma and the lymph node biopsy as leiomyosarcoma.

Malignant schwannoma cells can also display unusual differentiation example features of rhabdomyosarcoma, liposarcoma and osteoid and cartilagenous metaplasia. This can add to the difficulty in diagnosis.

Malignant schwannomas are mostly slow growing. However, they differ markedly in their clinical course. They have been reported to recur locally, spread along the nerve involved and metastasise to lungs and lymph nodes.³ Adequate excision of these tumours is essential to prevent recurrences and metastasis. Orbital metastasis is rare, the majority of metastatic orbital tumours being carcinomas (7%) with the primary mostly from the lungs, breasts and the gastro-intestinal tract. The incidence of metastatic orbital tumours is reported as 5%, 2–3% and 9%.⁴ The true

assess as this would involve ocular examination of all patients with systemic malignancies. The incidence in such series would be higher. Most metastatic orbital tumours must go through the lungs, and the pulmonary metastasis accompany the orbital metastasis. Spread to the orbit is *via* blood as the orbit has no lymphatics.

The metastatic nature of orbital lesion is often misdiagnosed by ophthalmologists in spite of the availability of information of pre-existing tumour. Often this history has to be probed for. It is useful to remember certain features of metastatic orbital lesions. The orbital symptoms appear rather suddenly.⁴ The patient complains of diplopia, lid swelling and ptosis. Ophthalmoplegia and diplopia are more marked early in metastatic lesions than in primary. Conjunctival chemosis and lid swellings are also distinctive features. Thus atypical features of orbital tumours like early pain, ptosis, ophthalmoplegia and bone

destruction should alert the ophthalmologist to the possibility of the tumour being metastatic.

A careful systemic workup including a chest X-ray should be carried out and may lead to the discovery of the primary lesion. In 30% of Henderson's series, the orbital metastasis was the first sign of an unknown silent primary lesion elsewhere.⁴

Most patients with orbital metastasis have a short survival period. Conservative and palliative therapy is therefore recommended. Radiotherapy to the orbit can shrink tumour size and increase the patient's comfort.

Surgical excision is indicated in those patients with longer survival period while some may need exenteration to relieve intractable orbital pain.

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