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MALIGNANT PERIPHERAL NERVE SHEATH TUMOUR IN A 12 YEARS GIRL:
A CASE REPORT

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ABSTRACT… Malignant peripheral nerve sheath tumours (MPNSTs) are rare, aggressive soft tissue sarcomas associated with poor prognosis, that most commonly affect patients aged 20 to 50 years, but have also been reported in children. The tumour is usually found in lower extremities and only 10% to 20% of all lesions occur in head and neck region thus making it a rare entity. Central involvement, particularly in the jawbones is quite unusual. There is little reported in literature on these tumors in Africa. Here we report a rare case of intraosseous MPNSTS occurring in the mandible in a 12-years old girl. The biological behavior and diagnostic challenges of this rare malignancy are discussed.

Key words: malignant peripheral nerve sheath tumor, biological behavior, diagnostic challenges.

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INTRODUCTION

Malignant peripheral nerve sheath tumour also known as malignant neurilemmoma, malignant schwannomas, neurofibrosarcomas or neurogenic sarcomas is a form of cancer of the connective tissues surrounding nerves. Given its origin and behavior it is classified as a sarcoma.[1,2] Accounts for 5% of all soft tissue sarcomas, with about half of the cases are diagnosed in people with neurofibromatosis; the lifetime risk for an MPNST in patients with neurofibromatosis type I is 8% to 13% (2). MPNST with rhabdomyoblastomatosis component are called malignant triton tumours. According to Enzinger and Weiss 1993, the term MPNST is preferred for these tumours because they may recapitulate the appearance of any cell of the Schwann cell and also the perineural fibroblast.[3] Most commonly affected are young adults in 20-30 years. Presents as enlarging mass that sometimes exhibits rapid growth with associated pain or nerve deficit. This tumour is usually found in lower extremities and retroperitoneal and is rare in head and neck area.[4,9] In head and neck, the mandible, lips and buccal mucosa are the most common site. Radiologically this tumour reveals widening of mandibular canal or mental foramen, with or without destruction of surrounding bone.[1,2] The intraosseous localization of MPNST is very rare in literature[1]: Dahlin and Krishanan[11] reported 10 cases, Wirth and Bray[12] 31 cases, Bullock et al.[13] 18 cases, and De la Monte et al.[14] presented 60 histologically documented cases. The diagnosis of MPNST has also been complicated by unclear criteria for determining the malignancy of a tumour originating in the nerve.[15] Since Harkin et al. (1978) first reported plexiform schwannoma[16]; several authors have reported plexiform schwannoma unassociated with Von-RecklingHausen's disease, which usually show benign histological features (Lee et al. 2001); (Woodruff et al. 1983); (Barbosa and Hansen 1984); and (Guarino 1993).[6,17–20] Here we report a case of a rare intra-osseous malignant peripheral nerve sheath tumour of mandible in an 12-year-old female patient. The biological behavior and diagnostic challenges of this rare tumour are discussed.

CASE REPORT

A 12- years- old girl presented to our clinic in February 2013 complaining of a huge swelling on the left side of the face which had been present since 2003 **(fig.1)**. It started as small nodular swelling on the left cheek which was firm, painless and gradually increased with time. In its course it

attained the size of her fist and she was operated in 2005. The swelling recurred 3years later and again was painless and firm. Examination revealed a huge swelling on the left side of the face about 37x40cm, oval with lobulations extending from level of left lateral orbital rim to lower border of the mandible superoinferiorly, and from sympheseal region to postauricular area anteroposteriorly. The overlying skin was slightly shiny and stretched, not attached to the swelling without increased local temperature. There were dilated vessels, firm to hard and some areas of softness in consistence. It involved the left nose and left eye, which were pushed medially and upward respectively. Pinna of the left ear was stretched and pushed laterally. There was reduced TMJ movement. The hard and soft palate were normal, teeth 36-43 were displaced to the right. The swelling extended from sympheseal region to left retromolar region. Lingual and buccal bone expansion pronounced in buccal aspect, bony hard in consistency. The tongue was free and mobile. Radiographic examination revealed a giant expansile bone destructive lesion involving the entire mandible with trabeculations.The appearance was that of mixed density (radiopaque/radiolucence) **(Fig-2)**.

An incisional biopsy was performed under local anesthesia. The histological analysis showed fibrous tissue stroma with collagens and many plump of spindle cells and fibroblasts. There were few scattered abnormal mitotic figures areas of hypercellular and hypocellular with nuclear pleomorphism and palisading. Based on these clinical, radiologic, and histopathologic features, a diagnosis of malignant schwannoma-lower jaw was made.

Under general anesthesia a tracheotomy was done. Bilateral submandibular incision with left preauricular and cervical extension was used to approach the tumor on the mandible. A very huge fibrous lesion with multiple lobules was exposed and removed in total by disarticulating the mandible on both sides. The tumour measured about 40x40 cm in its greatest dimension and weighed 2.5kg. Postoperative healing occurred without complications except subcutenous emphysema which appeared over anterior chest wall and upper limbs on day two postoperatively. The post-operative final appearance was acceptable **(Fig.3)**. The post-operative histological results confirm malignant schwannoma. One year later a patient reported again with small roundish lesion on the right side of the face which was located within the soft tissues. The lesion was surgically removed under general anesthesia. Unfortunately the patient lost her life from anesthetic complications in the recovering room.

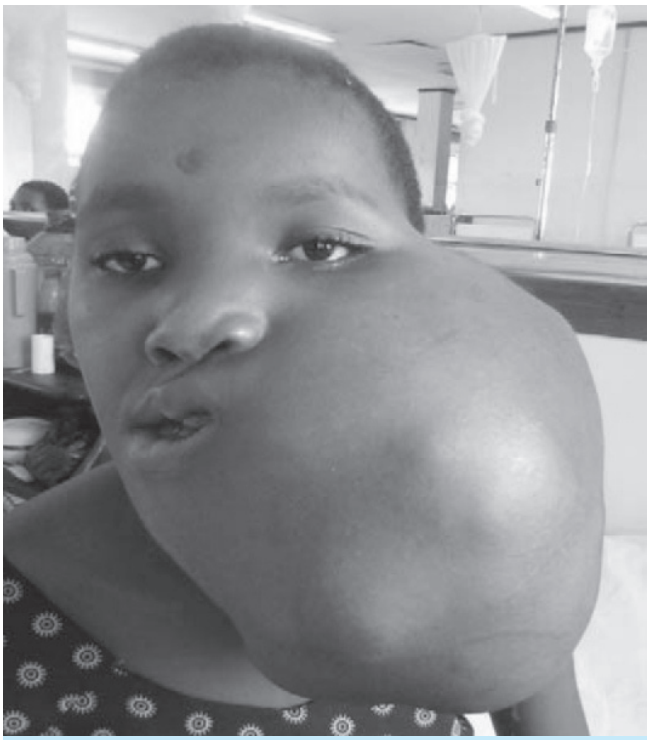

Fig-1. Patient before surgery

DISCUSSIONS

Neurogenic tumours are all probably derived from schwann cells. There are three varieties of benign tumours; neurilemmoma, neurofibroma and plexiform neurofibroma. However, there is only a single type of malignant neurogenic tumourrecognised-malignant schwannoma also known as malignant peripheral nerve sheath tumour (MPNST). The nomenclature surrounding malignant schwannoma has long been a subject of discussion.

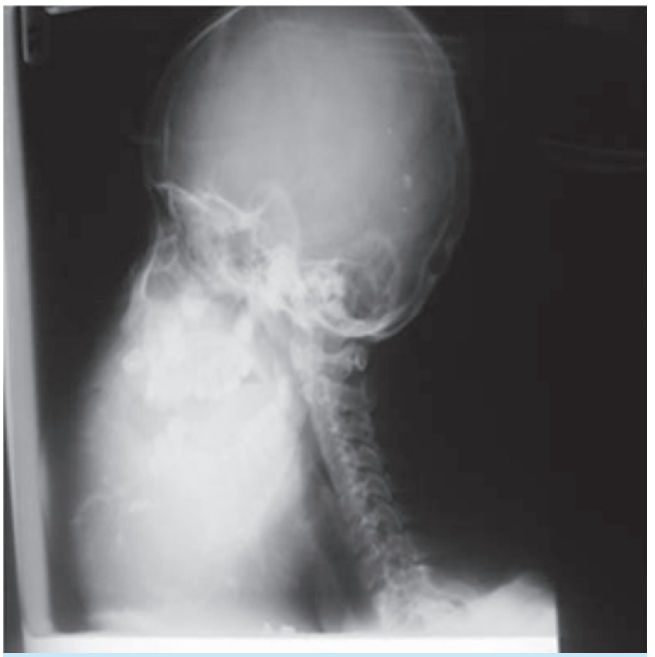

Fig-2. Radiographic appearance of the tumour. Mixed density (radiopaque/radiolucence)

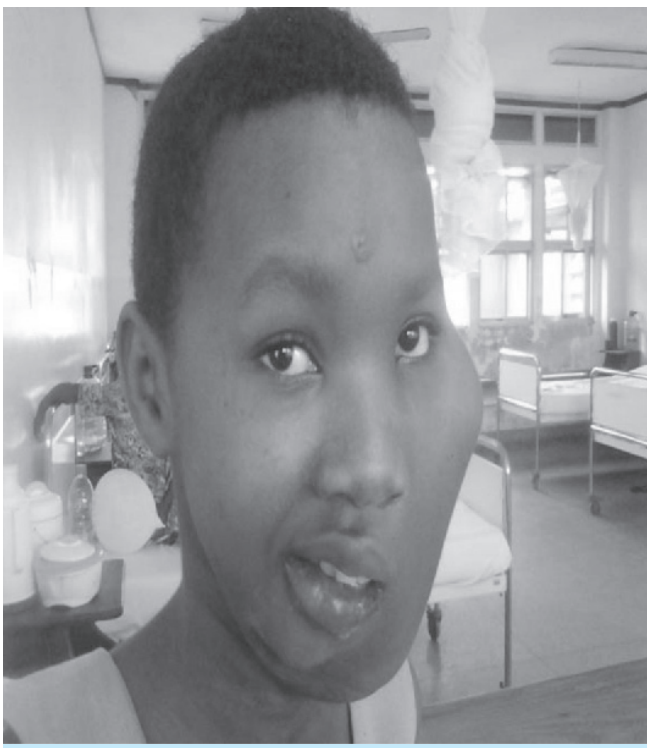

Fig-3. Patient after surgery

The sarcomatous lesion has also been called schwannomsarcoma, neurogenous sarcoma, neurofibrosarcoma and malignant neurilemmoma. The confusion having arisen due to uncertainties about its cell of origin. It is currently thought to arise from schwann cells on the basis of ultrastructural and histochemical studies.[5,6,18,19] MPNST is the coined term used by the WHO and corresponds to the malignant proliferation of any cell of the nerve sheath: Schwann cell, perineural fibroblast or endoneural fibroblast.[3,20,21] The tumour represents 10% of all soft tissue sarcomas and 8% to 16% in head and neck region.[20,22] Its development is thought to be a multi-step and multi gene process with an etiology of loss of chromosomal arm 17q sequence including complete inactivation of neurofibromatosis-1 gene.[5,7,23] This tumour occurs in the age group of 20 to 50 years with an equal male and female predilection.[5,6,9,24] Inspite of being uncommon, MPNST have been described in different locations of the body, occasionally associated or not with Neurofibromatosis type-I [(25),] most commonly affects the extremities, but within the head and neck their commonest site is within the superficial sensory nerves of the lateral neck. Cranial nerve involvement is uncommon, but malignant schwannomas of the III, V, VII, X, and XII cranial nerves have been described. Of these, the III cranial nerve is most frequently involved, however we only found a single report of a lingual nerve tumour and this was in a patient with von Reckling-hausen's disease.[26] Isolated cases in the orbit[27], neck[28], parapharyngealregion[6] have also been reported. On occasions the existence of the tumour is related to the presence of Neurofibromatosis or von Reckling-hausen's disease with described cases located in the bladder, thorax, orbit, mediastinum and prostate.[25] The tumour appears as a bosselated, sessile, circumscribed submucosal mass associated with pain or paresthesia or muscle weakness and atrophy.[6,7,23] This slow enlarging mass exhibits rapid growth[8] and two thirds of the lesions are more than 5cm at the time of diagnosis. Our patient illustrated the potential for these malignancies to present late due to their minimal propensity to produce symptoms. Although the tumouris malignant, its biological behavior (painless, slow enlarging mass and rare lymph node metastasis) necessitated the tumour to grow to a tremendous size without affecting patient's health or common

activities.

The definitive diagnosis of MPNST is histopathological. The tumor has two different cell patterns can usually be recognized on microscopy: Antoni type A: like pattern with spindle cells in a palisade formation, surrounded by an interstitial substance that forms verocay bodies.

Antoni type B: like pattern with irregular cells and a myxoid component.[10] Although Antoni A and B arrangements are commonly described in benign schwannoma[10] the long duration and slow growth of low grade MPNSTs can lead to similar histological characteristics. The presence of verocay bodies is pathognomic of neurilemomma[(10,29)]. Immuno-histochemistry plays an important part in the diagnosis and differential diagnosis of MPNSTs: excluding, fibrosarcoma, synovial sarcoma, fibrous histocytoma, adenoid cystic carcinoma, neurogenic sarcoma, and chondrosarcoma. Immuno-histochemically the tumour cells show immune-reactivity to the S-100 protein and vimentin, with focal positivity to CD68 and negativity to keratin.[30,31]

Radiological diagnosis of this tumor is very challenging. Radiographically thisintraosseoustumour of the jaws will show a complete destructive pattern with bony expansion, erosion, and tooth-mobility, widening of the mandibular canal[23,24] or mental foramen with or without irregular destruction of surrounding bone.[8] Our case showed complete bone destruction with mixed density which mimick radiographic appearance of osteosarcoma, ossifying fibroma and osteomyelitis. Therefore careful combined clinical, radiological and histological evaluation is essential for the diagnosis of this tumour.

The treatment of MPNSTs of jaws is wide surgical excision but local recurrence is common. Our case was treated by wide surgical excision but later was followed by local recurrence. Patient's survival is correlated to the size of lesion, adequacy of margins, association or not with neurofibromatosis-I.

CONCLUSION

Careful use of combined mode of diagnosis approach (clinical evaluation, histological evaluation and radiological evaluation) and close follow-up following treatment is essential for the management of this rare intraosseous malignant tumour.

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"You cannot escape the responsibility of tomorrow by evading it today."

Abraham Lincoln