Case Report

Low Grade Fibromyxoid Sarcoma of the Parotid in a 5-year-old Child

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Abstract

Low-grade fibromyxoid sarcoma (LGFMS) is a rare slow-growing malignant tumour with a deceptively benign histologic appearance. It typically involves young or middle-aged adults. These tumours usually occur in the proximal extremities and trunk. Sporadically they may be found in unusual locations leading to diagnostic dilemma. We report a 5year old boy that presented with painless right parotid swelling of 6 weeks duration. There was a 12-week presentation-to-diagnosis delay, due to the initial misdiagnosis as Burkitt's lymphoma that was entertained because of the region involved, rate of progression and endemicity of Burkitt's in our setting. Compromise of the aerodigestive tract was a major challenge, in addition to early recurrence and metastasis. We advocate for high index of suspicion, prompt tissue diagnosis and multidisciplinary intervention for better outcome.

Keywords: Fibromyxoid Sarcoma; Parotid Tumour; Multidisciplinary Team; Burkkitt's Lymphoma; Evan's Tumour; Child.

Introduction

Low-grade fibromyxoid sarcoma (LGFMS) is a rare tumor first described by Harry Evans in 1987 (Evan's tumour).[1-3] LGFMS has high metastasizing potential, despite the benign histologic appearance.[1,4] It typically involves young or middle-aged adults, although few pediatric cases have also been reported.[1,4] These tumours usually occur in the proximal extremities and trunk.[4] Sporadically they may be found in unusual locations leading to diagnostic dilemma. Primary LGFMS is an extremely rare tumour of the salivary glands and arises from undifferentiated pluripotentialmesenchymal cells.[4] We report a 5year old boy with LGFMS of the parotid gland masquerading as Burkitt's lymphoma.

Case Presentation

A 5-year-old boy referred by the pediatrician to the Ear Nose and Throat Clinic with a painless right parotid swelling of 6 weeks duration. The swelling started like the size of a peanut, it increased gradually in size from the onset. No preceding history of trauma, no fever, no swelling in other parts of the body. The patient was otherwise well and active. General physical examination was essentially normal and vital signs were stable. The swelling measured 15cm X 12cm, it was firm, non-tender, and with no differential warmth (Figure 1). No palpable cervical lymphadenopathy.

There was a moderately progressive increase in tumor size with intra-oral extension by the second

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week (Figure 1 and 2), plain X-ray showed destruction of the right ramus of the mandible; giving an impression of Burkitt's lymphoma but fine needle aspiration cytology (FNAC) was not suggestive. MRI demonstrated a dump bell shaped hyper-intense lesion in the parotid region with minimally enhancing area of mixed intensity (Figure 3). Oral extension of the tumour precluded feeding and constituted a threat to breathing (Figure 1). An incisional biopsy was taken, tracheostomy was done to prevent imminent airway obstruction, while a feeding gastrostomy was established at the same sitting. Histological diagnosis of LGFMS was made base on microscopic feature of a proliferating sarcomatous tumor disposed in whorled pattern in areas and myxoid in other areas. The tumor composed of alternating interwoven bundles of fibroblastic cells. The cells were fairly monomorphic with elongated nuclei and fairly abundant cytoplasm. Mitoses were inconspicuous and there was no area of necrosis. Histochemical stain confirmed the fibroblastic nature of the tumor (Figure 4). By this time the tumor had protruded out of the mouth with a huge extra-oral extension causing pressure necrosis of the lips. Tourniquet effect of the lips around the tumor also led to progressive necrosis and secondary infection of the tumor. Patient also developed lower motor neuron palsy of the cranial nerve VII. Subsequently, tumor was surgically excised followed by external beam radiotherapy; 60 Gy in 30 fractions of 2Gy per fraction using 60 mv photons. Patient became well and stable with good healing of the wound and subsequently had decannulation of tracheostomy tube. However, barely four months later he developed intraoral recurrence with features of intracranial extension and chest metastasis demonstrated on chest x-ray. Patient later succumbed after episodes of tonic-clonic convulsion and severe respiratory distress.

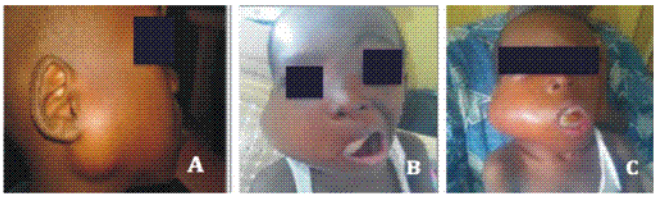

Figure 1: A- Swelling in parotid region at presentation B- Intraoral extension within 8weeks. C- Swelling fills the oral cavity within 12 weeks compromising feeding and breathing.

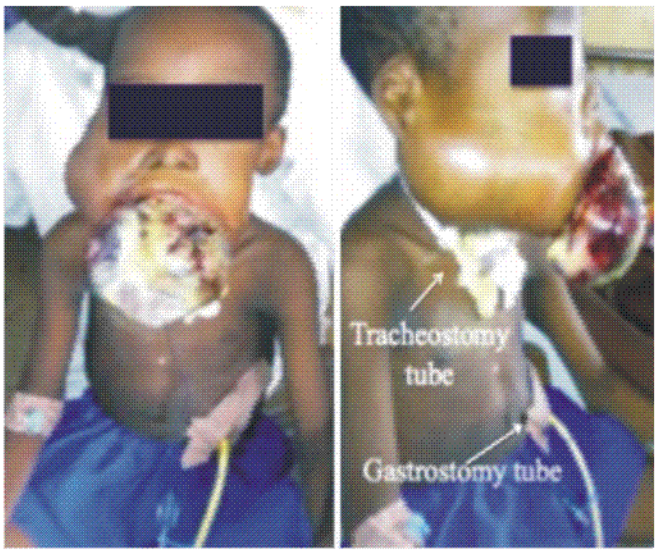

Figure 2: Patient with feeding gastrostomy and tracheostomy tube in situ.

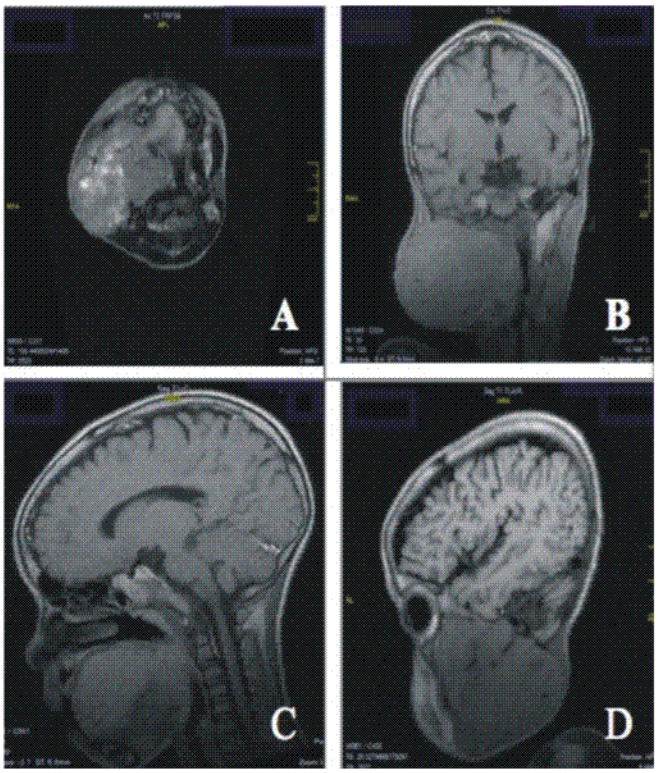

Figure 3: MRI showing: a dumbbell shaped expansile irregular mass in the parotid region (A&B). Appears extensive with extension into the oral cavity and upper margin of lesion in close proximity to the brain (C&D). Lesion is inhomogeneous hypo-intense on T1 Weighted image with foci of irregular more hypo-intensity. It is hyper-intensed on T2 Weighted image and enhances moderately with contrast.

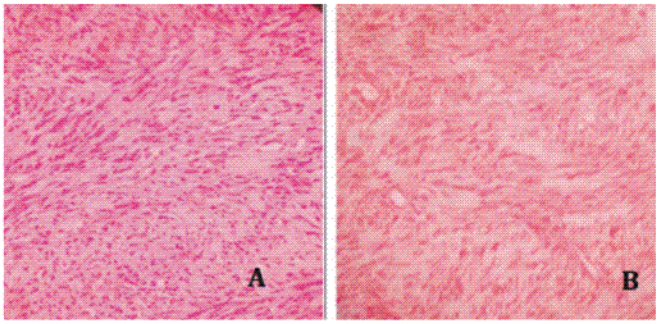

Figure 4: A- H/E x400 Section show: a proliferating sarcomatoustumor disposed in whorled pattern in areas and myxoid in other areas B- H1304-15(MT) X400 section show: Masson's trichrome (MT) stain. A tri-color staining protocol used in histology. Fibromyxoid area appear pale

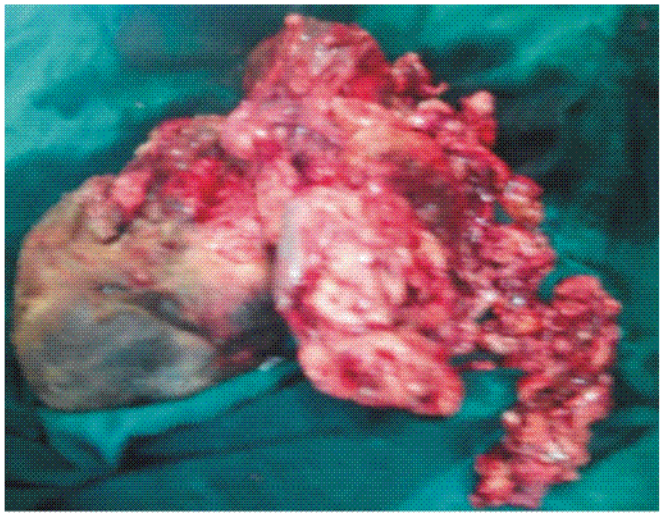

Figure 5: Tumour specimen

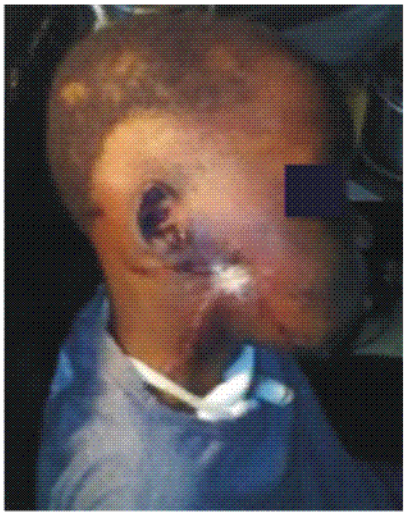

Figure 6:

Discussion

Low-grade fibromyxoid sarcoma (LGFMS) is a variant of fibrosarcoma first described as a pathologic entity by Harry Evans in 1987.[3] It belongs to the group of fibroblastic/ myofibroblastic soft tissue tumors. Two main subtypes have been recognized; classical LGFMS described by Evans[3] and LGFMS with giant collagen rosettes described by Lane et al.[5]The etiology and incidence of LGFMS is unknown and there are no known risk factor.[3,4]The male gender are more commonly affected than females.[4] The patient in this report is a 5-year-old boy with no family history of similar illness. Although LGFMS may occur at any age, it is predominantly seen in adults in their fourth decade of life.[5,6] A few pediatric cases have also been reported by earlier workers; with common primary locations in the paravertebral, thigh or intrathoracic region;[5-7] with the youngest documented case being a 4-year-old child.[7]

In our patient, the swelling was in the parotid region and was barely 6 weeks when he presented to the hospital. He was otherwise clinically stable and well preserved at this time (Fig. 1). This is relatively early going by the common attitude of late hospital presentation in Africa settings,[8] and may be explained by the unhidden nature of head and neck region and the progression of the swelling. In a series, pre-biopsy duration of over 5 years in 15% of patients was recorded,[4] and on rare occasions, acute respiratory distress and chest pain (in case of chest wall LGFMS) or seizure were factors that prompted early hospital presentation.[7,9] However, in this patient, the benefit of early presentation was hindered by a delay in arriving at a definitive diagnosis and surgical intervention. The tumor was locally advanced before intervention; both the superficial and deep lobe of the parotid was involved with erosion of the mandible and surrounding muscles. This is not uncommon, because bone destruction can occur secondary to pressure erosion;[4] particularly the destruction may be significant in thepaediatric age group. Such a finding does not necessarily indicate malignancy.[4]The tumour extended close to the orbit and skull base and filled the oral cavity and oropharynx precluding oral feeding and constituting a threat to breathing (Figure 2 and 3). A gastrostomy

had to be established for feeding and a preemptive tracheostomy was done to avert potential upper airway obstruction from the rapidly growing tumor. It is not clear whether this rapidity of tumour growth was precipitated by preceding events and procedures like FNAC, incision biopsy or trial chemotherapy carried out on the patient. However, further protrusion of the mass outside the mouth occurred thereafter and tourniquet effect of the orbicularis oris muscle around the protruding tumor mass caused ischemic necrosis and super-added infection rendering a large part of the fungating tissue non-representative of the original tissue after the final excision (Figure 5). These features of secondary inflammation and haemorrhage are not specific to LGFMS.[4] Lee et al presented a case of LGFMS with a fungating mass with internal haemorrhage and increased vascularity which was misdiagnosed initially as hemangioma on MRI rather than LGFMS.[4]

The cytologic features of LGFMS are not specific enough for a definitive diagnosis based on pre-operative FNAC alone;[10] nevertheless, correlating the cytologic and clinical findings can narrow the range of differential diagnoses which may include other myxoid lesions, like superficial or intramuscular myxoma and myxofibrosarcoma.[1,2,10] Cytogenetics is particularly useful in excluding rare cases of LGFMS from any myxoid pattern with unclear diagnosis.[10] In our case, FNAC was inconclusive; an incisional biopsy had to be taken before a diagnosis could be made and definitive treatment decision taken. Histopathology report in our center takes two weeks on the average and facility for cytogenetic study is also not available. The patient deteriorated clinically before histology report could be gotten; that was largely as a result of sepsis, poor feeding and contact bleeding from the macerating lips and tumor edges severe enough to warrant repeated blood transfusion.

Surgical excision was done using a modified Blair's incision and significant debulking was achieved. The superficial part of the dumbbell-shaped tumor was pushed towards the deep part and the entire mass was delivered through the oral cavity. Wound closure was done by multiple advancement flap. This was followed by radiotherapy. Although, earlier studies have shown that neither radiotherapy nor chemotherapy affects the clinical course of LGFMS, but could be applied for residual tumour or when resection margin is not free of tumour.[2,4] In all reported cases, surgery is the treatment of choice and success rate of 5year-survival after surgery is as high as 90% with a more favorable prognosis with smaller tumours.[4,6] The tumour in our case was huge coupled with the paediatric age of the patient and the delay in reaching a definitive diagnosis.

LGFMS are characteristically slow growing but may behave aggressively, with local recurrence and distant metastases which is common to the lungs.[4,6] Local recurrences of tumour within a few months to 15 years after primary surgery have been documented in the literature,[2,4] while distant metastasis has occurred from 0 to 45 years after surgery.[2,4] The index patient had two occasions of enucleation of recurrent tumours in the oral cavity barely twelve months after chemo-radiation. Those recurrent (LGFM) lesions present theclassical well-demarcated, yellow-whitish mass with focal glistening area,[4]which was easier to identify than the initial fungating mass. Distal metastasis was first noticed in the lungs and demonstrated on chest X-ray; he subsequently developed respiratory distress, tonic-clonic convulsion and other features of intracranial extension and finally succumbed to these complications.

One of the major challenges in the management of sarcomas especially that involving the head and neck is that en bloc resection is usually impossible in most of the cases. This often lead to early recurrence, and potentially life-threatening complications from damage to vital neurovascular structures.[1]More so, there is currently no data to support the use of any systemic or locoregional treatments in advanced, recurrent or metastatic disease.[2]

Conclusion

Fibromyxoid sarcoma of the parotid is rare and the clinical behavior may mimic Burkitt's lymphoma, which is relatively more common in African children. Compromise of the aerodigestive tract is a major challenge, while recurrence and metastasis could occur earlier than usual in the paediatric age group. We advocate for a high index of suspicion, promptttissue diagnosis and multidisciplinary

intervention for better outcome.

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